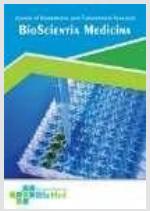
eISSN (Online): 2598-0580



Bioscientia Medicina: Journal of Biomedicine & Translational Research

Journal Homepage: www.bioscmed.com

Navigating a Rare Entity: Pancreatic Lipomatosis in a 34-Year-Old Female Without Classical Risk Factors – A Case Report

Ezra Gunadi[1]*, Ari Adrianto[2]

[1]Resident of General Surgery, Faculty of Medicine, Universitas Diponegoro/Dr. Kariadi General Hospital, Semarang, Indonesia

[2]Department of Digestive Surgery, Faculty of Medicine, Universitas Diponegoro/Dr. Kariadi General Hospital, Semarang, Indonesia

ARTICLE INFO

Keywords:
Fatty pancreas
Incidental finding
Non-alcoholic fatty pancreas disease
Pancreatic lipomatosis
Pancreatic steatosis

***Corresponding author:**
Ezra Gunadi

E-mail address:
egoenadi.dr@gmail.com

All authors have reviewed and approved the final version of the manuscript.

https://doi.org/10.37275/bsm.v9i8.1352

A B S T R A C T

Background: Pancreatic lipomatosis (PL), characterized by the replacement of pancreatic acinar tissue with mature adipose cells, is an uncommon condition with an etiology that is often not fully understood. It is frequently identified incidentally during imaging studies. The aim of this case report is to contribute to and advance the clinical understanding of this exceedingly rare and atypical presentation of pancreatic lipomatosis. **Case presentation:** This report details the case of a 34-year-old female who presented for evaluation of nonspecific, intermittent epigastric discomfort. Her medical history was devoid of significant illnesses, alcohol abuse, or known genetic conditions predisposing to pancreatic disorders. Comprehensive laboratory evaluations, encompassing serum amylase, lipase, liver function tests, lipid profile, and HbA1c, yielded results entirely within normal parameters. Abdominal computed tomography (CT) imaging revealed a striking diffuse, homogeneous fatty replacement of the entire pancreatic parenchyma, a hallmark of total pancreatic lipomatosis. Importantly, there was no evidence of pancreatic ductal dilatation, calcifications, or any discrete pancreatic masses. The patient's management was conservative, involving lifestyle counseling and scheduled periodic monitoring for the potential, though infrequent, development of complications such as exocrine pancreatic insufficiency or secondary diabetes mellitus. **Conclusion:** This case distinctly illustrates the occurrence of extensive pancreatic lipomatosis in a young, otherwise healthy female lacking classical risk factors. The pivotal role of cross-sectional imaging, specifically CT, in accurately diagnosing this benign condition is emphasized, which is crucial for averting misdiagnosis and precluding unnecessary invasive interventions. Enhanced awareness among clinicians is vital for appropriate patient counseling, the implementation of conservative management strategies, and diligent long-term monitoring for any potential metabolic or functional sequelae.

1. Introduction

Pancreatic lipomatosis (PL), a condition also recognized by terms such as fatty pancreas, pancreatic steatosis, or within certain contexts as non-alcoholic fatty pancreas disease (NAFPD), represents a distinct pathological state where the exocrine glandular tissue of the pancreas, primarily the acinar cells, undergoes progressive replacement by mature adipose tissue. While the endocrine components, the islets of Langerhans, and the pancreatic ductal system often maintain their structural integrity, particularly in earlier stages, extensive and complete fatty infiltration can, in some circumstances, lead to a compromise in overall pancreatic function. The reported incidence and prevalence of PL have demonstrated a discernible increase over the past few decades, a phenomenon largely ascribed to the significant advancements in the resolution capabilities and the increasingly widespread clinical application of sophisticated cross-sectional imaging technologies, including computed tomography (CT) and magnetic resonance imaging (MRI). Despite this increased detection, true diffuse or

total pancreatic lipomatosis, a state where the entirety of the pancreatic gland is almost completely supplanted by fat, continues to be a decidedly uncommon diagnostic finding. This is especially true when encountered in younger individuals who do not present with known predisposing conditions or identifiable risk factors.[1-3]

The etiology underlying the development of PL is understood to be multifactorial and, in many instances, remains incompletely elucidated. It has been observed in association with a diverse array of clinical conditions and factors. These include, but are not limited to, obesity, the cluster of metabolic abnormalities termed metabolic syndrome, type 2 diabetes mellitus, various forms of dyslipidemia, chronic and excessive alcohol consumption, certain viral infections (such as hepatitis B and C), exposure to particular medications (corticosteroids and tamoxifen being notable examples), states of malnutrition, iron overload disorders like hemochromatosis, and specific genetic syndromes, including Shwachman-Diamond syndrome and Johanson-Blizzard syndrome. Pancreatic lipomatosis is a particularly well-recognized and common manifestation in individuals with cystic fibrosis, resulting from the inspissation of secretions and chronic inflammation. In a subset of cases, PL may be classified as idiopathic, wherein no discernible underlying causative factor or associated condition can be identified despite thorough investigation.[4-6]

From a clinical perspective, the majority of individuals diagnosed with PL remain entirely asymptomatic. Consequently, the condition is often discovered incidentally when imaging studies are conducted for unrelated medical indications or symptoms. In those instances where symptoms do manifest, they are typically nonspecific in nature and may encompass complaints such as vague abdominal pain or discomfort, a sensation of bloating, or, in cases characterized by severe and extensive fatty infiltration, symptoms directly attributable to exocrine or endocrine pancreatic insufficiency. Exocrine pancreatic insufficiency (EPI) can develop and lead to clinical manifestations such as steatorrhea (the passage of fatty, bulky stools), maldigestion of nutrients, and unintentional weight loss. Endocrine dysfunction, on the other hand, may present as impaired glucose tolerance or progress to overt diabetes mellitus. Computed tomography imaging is widely regarded as the diagnostic gold standard for PL, characteristically demonstrating a diffuse reduction in the attenuation of the pancreatic parenchyma, with density values frequently approaching those observed in subcutaneous or retroperitoneal adipose tissue. Magnetic resonance imaging also provides excellent diagnostic accuracy in demonstrating fatty replacement, offering the advantages of superior soft tissue contrast and the absence of ionizing radiation.[7,8]

The particular novelty of the case presented in this report is centered on the observation of extensive, diffuse pancreatic lipomatosis in a young, 34-year-old female patient. She presented with only nonspecific epigastric discomfort and, quite significantly, had a complete absence of any identifiable classical risk factors typically associated with this condition, such as obesity, features of metabolic syndrome, pre-existing diabetes, a history of significant alcohol intake, or any known genetic disorders like cystic fibrosis. Such a comprehensive presentation of pancreatic lipomatosis in this specific demographic is notably unusual and certainly warrants detailed clinical attention and documentation.[9,10] Therefore, the primary aim of this case report is to contribute to and advance the clinical understanding of this exceedingly rare and atypical presentation of pancreatic lipomatosis. We intend to meticulously highlight the diagnostic pathway followed, with particular emphasis on the indispensable role of CT imaging in confirming the diagnosis.

2. Case Presentation

A 34-year-old Indonesian female patient presented to the outpatient digestive surgery clinic at Diponegoro National Hospital, Semarang, reporting a history of intermittent, nonspecific epigastric discomfort that

had been occurring for approximately six months prior to her consultation. The patient described the discomfort as a vague, dull ache, localized to the epigastrium. This sensation would typically arise two to three times per week, with each episode lasting for a duration of 30 to 60 minutes. She could not identify a consistent relationship between the onset of discomfort and her food intake, nor did she experience any radiation of the pain, particularly to the back. Associated symptoms such as nausea, vomiting, fever, jaundice, discernible changes in her bowel habits, or any significant unintentional weight loss were explicitly denied. The patient also reported that she had not found any specific factors that reliably relieved or aggravated her symptoms. She had previously tried over-the-counter antacid preparations, which provided only minimal and inconsistent relief from her epigastric discomfort.

Her past medical history was entirely unremarkable. She reported no history of chronic medical conditions, including diabetes mellitus, hypertension, dyslipidemia, previous episodes of pancreatitis, or any known liver diseases. There was no history of prior abdominal surgical procedures or significant abdominal trauma. Regarding her social history, she denied any consumption of alcohol, any history of smoking, or the use of illicit drugs. Her family history was negative for pancreatic diseases, cystic fibrosis, or any known hereditary forms of pancreatitis. At the time of presentation, she was not taking any regular prescribed or over-the-counter medications, specifically denying the use of corticosteroids or tamoxifen. She was employed as an office administrator, and her lifestyle was described as moderately active. Her Body Mass Index (BMI) was calculated to be 22.5 kg/m^2, a value falling comfortably within the normal range, thus excluding obesity as a contributing factor.

Upon physical examination, the patient was alert, fully oriented, and appeared to be in no acute distress. Her vital signs were stable and within normal limits: her blood pressure was recorded at 120/70 mmHg, heart rate was 76 beats per minute, respiratory rate was 16 breaths per minute, and her body temperature was 36.7°C. The abdominal examination revealed a soft, non-tender, and non-distended abdomen. There were no palpable abdominal masses, no evidence of hepatomegaly or splenomegaly, and no clinical signs suggestive of ascites. Bowel sounds were assessed as normoactive during auscultation. Specific clinical signs such as Murphy's sign (for cholecystitis) and Carnett's sign (to differentiate abdominal wall pain from visceral pain) were both negative. The remainder of her systemic physical examination was unremarkable.

A comprehensive panel of routine laboratory investigations was conducted to assess her general health and specifically to evaluate pancreatic and hepatobiliary function. The complete blood count, renal function tests (including urea and creatinine), and serum electrolyte levels were all found to be within their respective normal ranges. Liver function tests, which included measurements of alanine aminotransferase (ALT), aspartate aminotransferase (AST), alkaline phosphatase (ALP), gamma-glutamyl transferase (GGT), and total bilirubin, were all confirmed to be normal. Of particular significance to the pancreatic assessment, her serum pancreatic enzyme levels – specifically serum amylase and lipase – were well within the normal reference ranges (Amylase: 60 U/L, reference range 28-100 U/L; Lipase: 45 U/L, reference range 13-60 U/L). Her fasting blood glucose level was 88 mg/dL, and her hemoglobin A1c (HbA1c) level was 5.2%, effectively ruling out the presence of underlying diabetes mellitus or impaired glucose tolerance. The patient's lipid profile, encompassing total cholesterol, triglycerides, HDL-cholesterol, and LDL-cholesterol, was also found to be within acceptable and normal limits. Given the extensive nature of the fatty replacement of the pancreas that was subsequently identified on imaging, a screening test for common cystic fibrosis transmembrane conductance regulator (CFTR) gene mutations, particularly those prevalent in Asian populations, was considered. However, this was deferred because the patient exhibited no other clinical

stigmata typically associated with cystic fibrosis, such as a history of recurrent respiratory infections, chronic malabsorption dating back to a young age, or issues with infertility. Serum IgG4 levels were not initially measured, as there were no clinical or radiological features suggestive of autoimmune pancreatitis.

Owing to the persistent nature of her nonspecific epigastric discomfort, an abdominal computed tomography (CT) scan, performed with the administration of intravenous contrast material, was ordered to thoroughly evaluate the upper abdominal organs. The CT scan yielded striking and definitive findings pertaining to the pancreas. The entirety of the pancreatic gland, extending from its head through the body to the tail, demonstrated a diffuse and remarkably homogeneous replacement of its normal glandular tissue by tissue exhibiting fat attenuation characteristics. The mean attenuation values within the pancreatic parenchyma ranged from -80 to -110 Hounsfield Units (HU), consistent with mature adipose tissue. Despite this extensive fatty infiltration, the overall size and contour of the pancreas appeared to be maintained, possibly even slightly bulky due to the diffuse fat deposition, but critically, there was no evidence of any discrete pancreatic mass, cystic lesion, or surrounding inflammatory stranding. The main pancreatic duct was visualized and found to be of normal caliber, not dilated, and there were no visible pancreatic calcifications. The common bile duct was also observed to be normal in caliber. Other visualized abdominal organs, including the liver, spleen, kidneys, adrenal glands, and the accessible portions of the bowel, appeared unremarkable. No evidence of retroperitoneal lymphadenopathy was noted. These comprehensive imaging findings were considered highly characteristic and diagnostic of diffuse, total pancreatic lipomatosis (Figure 1 and 2). A summary of the patient's key clinical findings is presented in Table 1.

Table 1. Summary of patient's clinical findings.

Feature	Finding
Demographics	
Age	34 years
Gender	Female
Presenting complaint	
Chief complaint	Intermittent, nonspecific epigastric discomfort
Duration of complaint	Approximately 6 months
Relevant history	
Past medical history	Unremarkable; no chronic illnesses, pancreatitis, or liver disease
Social history	No alcohol consumption, no smoking
Family history	Negative for pancreatic diseases, cystic fibrosis
Medications	None regularly
Body mass index (BMI)	22.5 kg/m² (Normal range)
Physical examination	
General	No acute distress, vital signs stable
Abdomen	Soft, non-tender, no palpable masses, no hepatosplenomegaly
Laboratory investigations	
Serum amylase	60 U/L (Normal)
Serum lipase	45 U/L (Normal)
Liver function tests	Normal
Fasting blood glucose	88 mg/dL (Normal)
HbA1c	5.2% (Normal)
Lipid profile	Normal
Imaging findings (CT)	
Pancreatic parenchyma	Diffuse, homogeneous fat attenuation (-80 to -110 HU) replacing normal glandular tissue
Pancreatic size/Contour	Maintained, slightly bulky; no discrete mass or cyst
Pancreatic duct	Not dilated
Calcifications/Inflammation	No
Final diagnosis	Idiopathic Diffuse Total Pancreatic Lipomatosis

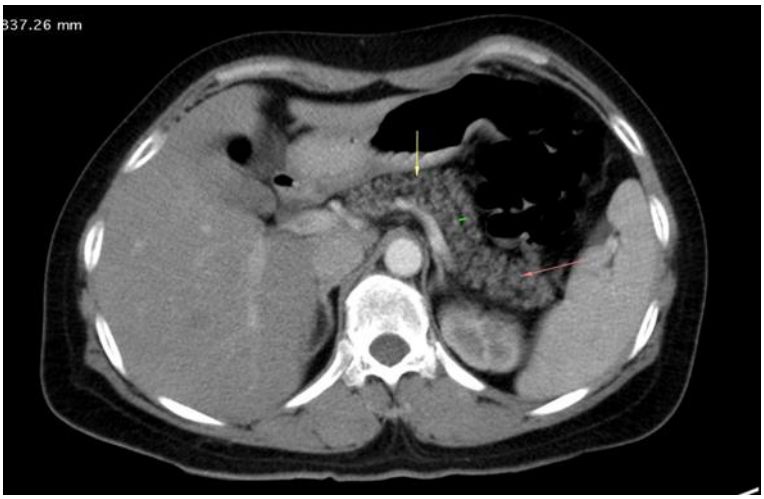


Figure 1. Axial non-contrast CT image through the mid-abdomen demonstrating near-complete replacement of the pancreatic parenchyma by adipose tissue (yellow arrow points to the head, green arrow to the body, red arrow to the tail region, illustrating the diffuse nature). The attenuation values were consistent with mature fat.

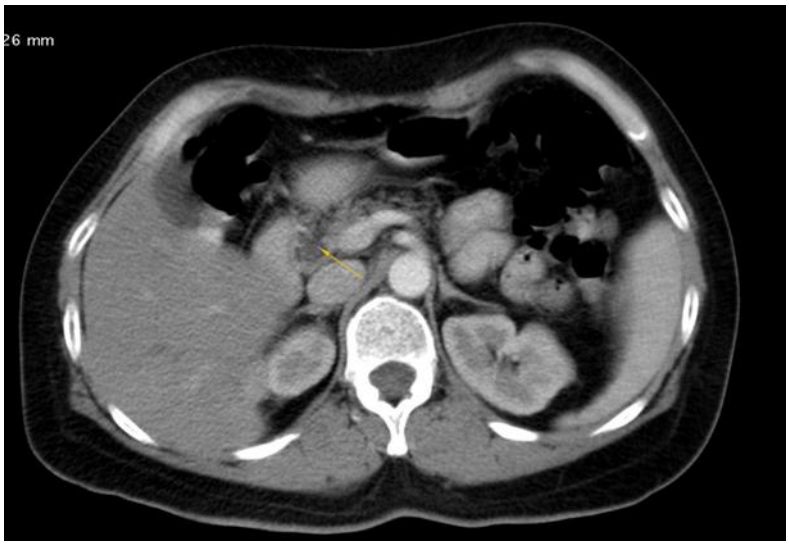


Figure 2. Axial contrast-enhanced CT image at a similar level showing homogeneous fat-density replacement of the entire gland (orange arrow indicating the extent of fatty change in the pancreatic body/tail). Note the absence of ductal dilatation or enhancing solid masses. The surrounding vessels and structures are clearly delineated.

Based on the conclusive CT findings and the overall clinical picture, a definitive diagnosis of idiopathic diffuse total pancreatic lipomatosis was established. The patient's reported mild epigastric discomfort was considered, after careful evaluation, to be potentially related to the altered pancreatic morphology or, perhaps more plausibly, a coexisting functional gastrointestinal symptom, particularly given its nonspecific nature and the typically asymptomatic clinical course observed in many individuals with pancreatic lipomatosis.

The management plan formulated for the patient was conservative, aligning with best practice for such benign incidental findings. She received

comprehensive education regarding the nature of pancreatic lipomatosis, specifically emphasizing its generally benign long-term course and the low likelihood that her current mild symptoms were indicative of severe pancreatic dysfunction, a conclusion supported by her consistently normal pancreatic enzyme levels and the absence of any malabsorptive symptoms. She was counseled on the importance of maintaining a healthy lifestyle, which included adherence to a balanced diet and engagement in regular physical activity, primarily to prevent the future development of metabolic risk factors that could potentially exacerbate fatty infiltration in various organs or lead to other unrelated health issues. No specific pharmacological treatment was initiated for the pancreatic lipomatosis itself. For her presenting complaint of epigastric discomfort, a short trial course of a proton pump inhibitor was prescribed for symptomatic relief, which did result in some subjective improvement in her symptoms.

A schedule for periodic follow-up was established to monitor for any new symptoms that might suggest the development of exocrine pancreatic insufficiency (such as steatorrhea or unexplained weight loss) or endocrine dysfunction (such as symptoms indicative of diabetes mellitus). This follow-up plan included clinical reviews at 6-month and 12-month intervals post-diagnosis. At both of these follow-up appointments, the patient reported that her intermittent epigastric discomfort remained infrequent and had not worsened in character or intensity. She specifically denied the onset of any new gastrointestinal or systemic symptoms. Repeat laboratory testing for pancreatic enzymes (amylase and lipase) and HbA1c, conducted at the 12-month follow-up visit, remained within normal limits. The ongoing plan involves continued annual clinical review, with further investigations to be guided by any new symptoms or clinical concerns that may arise. The details of her treatment and follow-up are summarized in Table 2.

Table 2. Procedure of treatment and follow-up.

Aspect	Details
Initial management	
Primary approach	Conservative management
Patient education	Explanation of Pancreatic Lipomatosis, benign nature, monitoring for rare complications
Lifestyle advice	Balanced diet, regular physical activity
Symptomatic treatment	Short course of proton pump inhibitor for epigastric discomfort (with some improvement)
Pharmacological for PL	None initiated
Follow-up schedule	
Short-term	Clinical review at 6 months and 12 months post-diagnosis
Long-term	Annual clinical review planned
Follow-up assessments	
Symptom review	Assessment for new or worsening symptoms (epigastric pain, steatorrhea, weight loss, diabetic symptoms)
Laboratory (12-month)	Repeat serum amylase, lipase, HbA1c
Imaging	No routine repeat imaging planned unless clinically indicated
Patient outcome at follow-up (12 months)	
Epigastric discomfort	Infrequent, unchanged character, not worsened
New symptoms	None reported (no steatorrhea, weight loss, or symptoms of diabetes)
Laboratory results	Amylase, lipase, HbA1c remained within normal limits
Overall status	Stable, well, continuing conservative management

3. Discussion

Pancreatic lipomatosis (PL) encompasses a spectrum of conditions involving the infiltration or replacement of pancreatic exocrine tissue with mature adipose cells. This case of a 34-year-old female, presenting with extensive, near-total pancreatic lipomatosis diagnosed via characteristic CT findings, yet lacking traditional risk factors like obesity, metabolic syndrome, or cystic fibrosis, and exhibiting only mild, nonspecific epigastric discomfort with preserved pancreatic exocrine and endocrine function, offers a valuable opportunity to delve into the nuanced aspects of this condition. The etiology of pancreatic lipomatosis is diverse and, in many instances such as the present case, remains idiopathic despite thorough clinical evaluation. The absence of common culprits—obesity (BMI 22.5 kg/m²), type 2 diabetes (normal HbA1c), dyslipidemia (normal lipid profile), chronic alcohol use, medication-induced steatosis, or genetic syndromes like cystic fibrosis —compels a deeper consideration of less common or unknown mechanisms. While non-alcoholic fatty pancreas disease (NAFPD) is increasingly recognized as a component of the metabolic syndrome, mirroring non-alcoholic fatty liver disease (NAFLD), this patient did not fit that paradigm. NAFPD is strongly linked to insulin resistance, visceral adiposity, and ectopic fat deposition. The pathophysiology involves an imbalance between fatty acid uptake, synthesis, and oxidation within pancreatic cells, potentially leading to lipotoxicity, oxidative stress, and low-grade inflammation, which can further drive acinar cell damage and replacement by fat. Studies have demonstrated that pancreatic fat content correlates with waist circumference, BMI, serum triglycerides, and insulin resistance markers. In patients with metabolic syndrome, inflammatory cytokines and adipokines released from visceral adipose tissue might also contribute to pancreatic steatosis. The stark contrast in our patient—a young, lean individual with no biochemical markers of metabolic dysregulation—suggests her PL likely stems from a pathway distinct from the common NAFPD associated with metabolic syndrome. This highlights that while metabolic factors are a major driver for PL in the general population, they are not a universal prerequisite.[11,12]

One longstanding theory posits that PL can result from progressive acinar cell atrophy secondary to various forms of pancreatic injury. This could include chronic obstruction of the pancreatic ducts (not evident in this case), recurrent subclinical pancreatitis (no history or enzymatic evidence), viral infections, or exposure to toxins. When acinar cells are lost, the structural framework of the pancreas may be repopulated by adipose tissue, possibly derived from local mesenchymal stem cells or transdifferentiation of other cell types. Siegler's original description emphasized the near absence of exocrine tissue with concurrent replacement by normal adipose tissue, alongside preservation of ducts and islets, which aligns with the imaging appearance in our patient. The trigger for such extensive acinar cell loss in an otherwise healthy young individual remains speculative. Perhaps a subclinical viral infection in the past, or an unknown congenital predisposition leading to heightened acinar cell fragility or accelerated apoptosis, could be involved. Given the patient's young age and the completeness of the fatty replacement, congenital or developmental anomalies warrant consideration. While complete pancreatic agenesis is a distinct and severe condition, more subtle developmental issues affecting acinar cell proliferation or survival, or an inherent predisposition of pancreatic stromal cells to differentiate into adipocytes, could theoretically lead to such a picture over years. Conditions like Johanson-Blizzard syndrome or Shwachman-Diamond syndrome, which feature PL, are associated with specific genetic mutations and other systemic manifestations not present here. However, this does not exclude the possibility of a novel or very rare genetic predisposition specific to pancreatic adipose regulation. The preservation of pancreatic ducts and islets, as inferred from the imaging and normal endocrine function, suggests that the insult or developmental issue was relatively specific to the acinar cell compartment or that the

supporting stromal and ductal structures were more resilient. When all known causes are excluded, a diagnosis of idiopathic PL is made. This case likely falls into this category. Idiopathic PL may represent a heterogeneous group of disorders with different underlying, currently unrecognized, pathomechanisms. The diffuse and homogeneous nature of the fatty replacement in this patient might suggest a globally acting factor or a fundamental alteration in pancreatic tissue homeostasis rather than a focal process. Research into the cellular origins of adipocytes in the pancreas—whether from periacinar stellate cells, resident fibroblasts, or circulating progenitors—is ongoing and might shed light on such idiopathic cases.[13,14]

The near-total replacement of functional exocrine parenchyma by adipose tissue, as seen in this patient, naturally raises questions about how such extensive morphological change can occur and why, in this instance, it did not lead to overt exocrine or endocrine insufficiency. Adipose tissue accumulation in the pancreas can occur through several mechanisms. Acinar Cell Death and Replacement. This is the most widely accepted model for extensive PL. Loss of acinar cells through apoptosis or necrosis (due to various insults discussed above) creates space that is subsequently filled by proliferating adipocytes. The source of these adipocytes could be local mesenchymal precursor cells within the pancreatic stroma that differentiate into fat cells. Pancreatic stellate cells, which are key players in pancreatic fibrosis, have also been proposed as potential precursors that can transdifferentiate into adipocyte-like cells under certain stimuli. In NAFPD associated with metabolic syndrome, increased circulating fatty acids and de novo lipogenesis within remaining acinar cells or infiltrating adipocytes contribute to fat accumulation. This pathway seems less relevant for our non-metabolically compromised patient.[15,16]

The pancreas has a substantial exocrine reserve. Symptoms of maldigestion typically do not manifest until over 90% of acinar function is lost. In our patient, despite the imaging appearance of near-total fatty replacement, it is possible that microscopic foci of functional acinar cells, undetectable by CT, were still present and sufficient to maintain adequate digestion, evidenced by normal amylase/lipase and absence of malabsorptive symptoms. Alternatively, the process of fatty replacement might have been so gradual that any decline in function was too slow to become clinically apparent or for compensatory mechanisms (like hyperphagia, not reported here) to become overwhelmed. The normal amylase and lipase levels are particularly interesting; while these enzymes are produced by acinar cells, their serum levels primarily reflect cell damage or ductal obstruction leading to leakage into the bloodstream, rather than total functional mass. Thus, normal levels in this context indicate the absence of ongoing acute injury or significant obstruction, but not necessarily robust functional reserve. A more sensitive test of exocrine function, like a fecal elastase-1 measurement or a secretin-stimulated pancreatic function test, was not performed but might have revealed subclinical insufficiency. The islets of Langerhans, responsible for hormone production including insulin and glucagon, are often relatively spared in PL, particularly in cases not associated with severe, long-standing type 2 diabetes or cystic fibrosis. This anatomical observation likely explains the patient's normal HbA1c and fasting glucose. The embryological origin and structural organization of islets differ from acinar tissue, potentially rendering them more resistant to the processes driving fatty replacement of the exocrine component. However, some studies suggest that severe pancreatic steatosis, even NAFPD, can be associated with beta-cell dysfunction and an increased risk of type 2 diabetes, possibly due to local lipotoxicity affecting the islets or altered paracrine signaling within the fatty pancreas. Continued monitoring of glycemic status is therefore prudent.[17,18]

The patient presented with nonspecific, intermittent epigastric discomfort. While it is tempting to attribute any abdominal symptom to a striking radiological finding, the relationship between PL and such symptoms is not always clear-cut. Most cases of

PL, even extensive ones, are asymptomatic. When pain does occur, it's often in the context of associated pancreatitis, ductal obstruction, or very rarely, mass effect from extreme pancreatic enlargement (lipomatous pseudohypertrophy). None of these features were present here. It's conceivable that altered pancreatic texture or subtle changes in local mechanics due to massive fatty infiltration could cause vague discomfort, but this is speculative. Given the high prevalence of functional gastrointestinal disorders (FGIDs) like functional dyspepsia in the general population, it is highly plausible that the patient's epigastric discomfort was an independent issue, and the PL was a truly incidental finding. The partial improvement with a proton pump inhibitor might support a component of acid-related dyspepsia. The diagnostic workup, particularly the CT scan, was crucial not only in identifying PL but also in ruling out other, more sinister causes of epigastric pain, such as peptic ulcer disease (though CT is not primary for this), gallstones (not specifically mentioned but usually visible), chronic pancreatitis (no calcifications or duct changes), or pancreatic neoplasms (no mass).This case underscores the clinical challenge: when an unusual radiological finding is made in a patient with common, nonspecific symptoms, determining causality versus coincidence requires careful clinical judgment and exclusion of other more likely causes for the symptoms.[19,20]

CT imaging was central to the diagnosis in this case, demonstrating diffuse, homogeneous fat attenuation throughout the pancreatic parenchyma. The hallmark is the low attenuation values, typically ranging from -30 to -120 Hounsfield Units (HU), similar to subcutaneous or retroperitoneal fat. This allows clear differentiation from normal pancreatic parenchyma (typically +30 to +50 HU) and from fluid-filled cystic lesions or solid tumors. The homogeneity of the fat replacement, as seen here, is characteristic of benign PL. The preservation of the overall pancreatic shape, albeit sometimes with slight bulkiness, is also typical. The absence of ductal dilation, calcifications, inflammatory stranding, or enhancing solid components further supports the benign nature. While non-contrast CT is often sufficient to identify fat, contrast-enhanced phases are valuable for excluding enhancing masses and assessing vascular anatomy. Sometimes, residual strands of normal pancreatic tissue or compressed ductal structures within the fatty pancreas can enhance, creating a pseudomass appearance. Careful correlation with non-contrast images and awareness of this pitfall are important. In this case, the fatty replacement was so complete that such issues did not arise. MRI with fat-suppression sequences (Dixon, STIR) is equally, if not more, sensitive for detecting and quantifying pancreatic fat. T1-weighted images show high signal intensity from fat, which then suppresses on fat-saturated sequences. MRI offers superior soft tissue contrast, can better delineate small non-fatty lesions if present, and allows for MRCP to non-invasively visualize the pancreatic duct. However, CT is often more readily available, faster, and less expensive, making it a common first-line advanced imaging modality. Transabdominal ultrasound typically shows a hyperechoic (bright) pancreas in PL, as fat is more echogenic than normal pancreatic tissue. However, this finding is nonspecific and can be seen in chronic pancreatitis or even with aging. Visualization can be limited by bowel gas or body habitus. Endoscopic ultrasound (EUS) provides higher resolution images and can be useful for evaluating focal lesions or when other imaging is equivocal, but it is invasive and not necessary when CT findings are classic for diffuse PL, as in this case. The CT findings in this patient were so unequivocal for total pancreatic lipomatosis that further imaging or invasive procedures like EUS-FNA were deemed unnecessary, correctly avoiding potential complications and costs.

When encountering near-total fatty replacement of the pancreas, several differential diagnoses must be considered, although the classic CT appearance often makes the distinction straightforward. Congenital absence (agenesis) or underdevelopment (hypoplasia) of parts or all of the pancreas. Dorsal pancreatic agenesis is the most common form, where the body

and tail are absent, and the head (derived from the ventral bud) may be hypertrophied. Complete agenesis is extremely rare and usually incompatible with life without immediate and intensive pancreatic enzyme and hormone replacement. Imaging in agenesis shows a void in the pancreatic bed, not a pancreas replaced by fat. The key differentiating feature is the presence of a structurally intact pancreatic outline filled with fat and identifiable, albeit potentially atrophic, ductal structures in PL, versus complete absence of tissue and ducts in agenesis. Our patient had a pancreas of relatively normal size and shape, just composed of fat. End-stage chronic pancreatitis can lead to profound acinar loss and parenchymal atrophy, with some secondary fatty infiltration. However, chronic pancreatitis is almost invariably accompanied by other characteristic features: a shrunken, fibrotic gland; irregular ductal dilations and strictures; and often intraductal or parenchymal calcifications. None of these were seen in our patient. Her history was also negative for recurrent pancreatitis. As mentioned, CF is a well-known cause of extensive PL, often leading to complete fatty replacement by adulthood. However, CF is a multisystem disorder, typically presenting with chronic respiratory symptoms, malabsorption from a young age, failure to thrive, and male infertility. Our patient had no such history or clinical features. While a CFTR mutation screen was deferred, the clinical picture was strongly against CF. These are rare genetic syndromes characterized by exocrine pancreatic insufficiency (often with lipomatosis), skeletal abnormalities, and hematological dysfunction (SDS) or distinct craniofacial features and hypothyroidism (JBS). Again, the systemic features of these syndromes were absent. A benign tumor composed entirely of mature adipocytes. Pancreatic lipomas are rare and typically appear as well-encapsulated, focal fatty masses. Diffuse involvement mimicking total PL is not characteristic of a lipoma. An extremely rare malignant mesenchymal tumor. Pancreatic liposarcomas usually present as large, infiltrative, heterogeneous masses, often with areas of non-fatty soft tissue density, necrosis, or calcification. The uniform, benign appearance of the fatty tissue in our patient was not suggestive of malignancy. Rare germ cell tumors that can contain various tissue types, including fat. However, they are typically complex, heterogeneous masses with cystic and solid components, and often calcifications.

The CT findings in this case—diffuse, homogeneous fatty replacement of an otherwise morphologically normal-sized pancreas with no discrete masses, ductal abnormalities, or calcifications —were classic for benign pancreatic lipomatosis and did not strongly support these alternatives. Given the benign nature of idiopathic PL and the patient's minimal symptoms with preserved pancreatic function, a conservative management strategy was appropriately adopted. There is currently no specific medical or surgical treatment to reverse or remove the fatty infiltration in PL. Management is therefore focused on addressing any underlying causes (if identified and treatable), managing complications (EPI, diabetes), and providing reassurance. The trial of a proton pump inhibitor for her nonspecific epigastric discomfort was a reasonable step, addressing a common potential cause of such symptoms. Advising a healthy lifestyle is generally good practice, although its direct impact on established, extensive idiopathic PL is unknown. The key element of management is long-term monitoring for the potential, albeit low-probability in this specific context. Annual clinical review for symptoms like steatorrhea, weight loss, or new digestive complaints. Fecal elastase-1 could be considered if symptoms arise. Annual monitoring of HbA1c or fasting glucose, especially as intra-pancreatic fat can be associated with beta-cell dysfunction over time. The prognosis for asymptomatic or mildly symptomatic idiopathic PL with preserved function is generally excellent. The condition itself is not life-threatening. The main long-term considerations are the development of functional insufficiencies, which appear to be rare in idiopathic cases without other co-morbidities. There is some debate in the literature about whether NAFPD/PL increases the risk of pancreatic cancer, but the evidence is not definitive and often confounded by

shared risk factors like obesity and diabetes, which were absent here. For idiopathic PL in an otherwise healthy individual, the cancer risk is likely not significantly elevated above baseline. The decision to manage conservatively and avoid further invasive investigations or interventions was well-justified by the classic benign imaging features and the lack of concerning clinical or laboratory findings. This approach aligns with current recommendations and prioritizes patient well-being by avoiding the risks and costs of unnecessary procedures. This case of total pancreatic lipomatosis in a young, healthy female without any discernible risk factors contributes to the spectrum of PL presentations. It highlights that while PL is often linked to metabolic disorders or specific syndromes, it can also occur as an extensive, idiopathic finding. The generally benign nature of this condition, when presenting in isolation, supports a conservative management approach centered on education, reassurance, and long-term monitoring for functional changes. The striking imaging findings, juxtaposed with minimal clinical impact in this patient, underscore the pancreas's considerable functional reserve and the often-incidental nature of even profound morphological alterations.

4. Conclusion

This case report has meticulously detailed an unusual and noteworthy presentation of diffuse, total pancreatic lipomatosis in a young, 34-year-old female patient who notably lacked any of the classical risk factors commonly associated with this condition. The diagnosis was unequivocally established through characteristic abdominal CT findings, which revealed a striking and near-complete fatty replacement of the pancreatic parenchyma, while the gland's overall morphology and the integrity of its ductal system remained preserved. A critical aspect of this case is that despite the profound anatomical alteration visualized on imaging, the patient exhibited normal pancreatic exocrine and endocrine function at the time of her diagnosis and throughout her initial follow-up period. The presentation and subsequent management of this patient underscore several pivotal clinical learning points. Firstly, pancreatic lipomatosis, even in its most extensive forms, can manifest in young individuals who do not fit the typical patient profile associated with metabolic disorders or other known predisposing conditions. Secondly, computed tomography is a crucial and often definitive imaging modality for the accurate diagnosis of this condition. Accurate recognition of this benign entity is paramount to avoid misdiagnosis and prevent unnecessary invasive procedures. Finally, a conservative management strategy, encompassing patient education, lifestyle advice, and periodic long-term monitoring for potential functional changes, is the most appropriate course of action in such cases. Heightened awareness among clinicians regarding the varied presentations of pancreatic lipomatosis is essential for optimal patient care and for advancing our understanding of this intriguing condition, particularly in its idiopathic forms.

5. References

1. Beran A, Alchirazi KA, Ramai D, Kamal F, Saleem N, Fogel EL, et al. Pancreatic steatosis is a risk factor for post-ERCP pancreatitis: a systematic review and meta-analysis. Gastrointest Endosc. 2025; 101(5): S430–1.
2. Andert A, Ulmer TF, Schöning W, Kroy D, Hein M, Alizai PH, et al. Grade of donor liver microvesicular steatosis does not affect the postoperative outcome after liver transplantation. Hepatobiliary Pancreat Dis Int. 2017; 16(6): 617–23.
3. Moosburner S, Gassner JMGV, Nösser M, Pohl J, Wyrwal D, Claussen F, et al. Prevalence of steatosis hepatis in the eurotransplant region: Impact on graft acceptance rates. HPB Surg. 2018; 2018: 1–9.
4. Starodubova AV, Kosyura SD, Livantsova EN, Varaeva YR, Krasilova AA. Diagnosing pancreatic steatosis in obese patients. Bul of the Club of Pancr. 2019; 45(4): 30–3.

5. Gubergrits NB, Byelyayeva NV, Mozhyna TL, Monogarova NE, Borodiy KN, Yuryeva AV. Pancreatic steatosis: a giant step forward or recognition of ignorance. Bul of the Club of Pancr. 2020; 49(4): 21–41.
6. Jeon H, Jung MK, Lee JM. Nonalcoholic fatty pancreas disease. Korean J Pancreas Biliary Tract. 2023; 28(4): 95–102.
7. Munika E, Gunawan S, Wuyung PE, Soetikno V. Beneficial effects of 6-gingerol on fatty pancreas disease in high-fat high-fructose diet-induced metabolic syndrome rat model. Pancreas. 2024; 53(2): e193–8.
8. Kumar R, Bhargava A, Jaiswal G. A case report on total pancreatic lipomatosis: an unusual entity. Int J Health Sci (Qassim). 2017; 11(4): 71–3.
9. Schawkat K, Eshmuminov D, Lenggenhager D, Endhardt K, Vrugt B, Boss A, et al. Preoperative evaluation of pancreatic fibrosis and lipomatosis: Correlation of magnetic resonance findings with histology using magnetization transfer imaging and multigradient echo magnetic resonance imaging. Invest Radiol. 2018; 53(12): 720–7.
10. Gansner JM, Furutani E, Campagna DR, Fleming MD, Shimamura A. Pancreatic lipomatosis in Diamond–Blackfan anemia: The importance of genetic testing in bone marrow failure disorders. Am J Hematol. 2018; 93(9): 1194–5.
11. Kulalı F, Emir SN, Semiz-Oysu A, Özel Y, Bükte Y. The role of magnetic resonance imaging for evaluation of pancreatic lipomatosis after bariatric surgery. Haseki tıp bül. 2019; 57(3): 304–9.
12. Fahr L, Sunami Y, Maeritz N, Steiger K, Grünewald TGP, Gericke M, et al. Expression of the EWSR1-FLI1 fusion oncogene in pancreas cells drives pancreatic atrophy and lipomatosis. Pancreatology. 2020; 20(8): 1673–81.
13. assiri S, Tomaszewski M, Ou G. A276 total pancreatic lipomatosis and exocrine pancreatic insufficiency in a patient with celiac disease: an unusual entity. J Can Assoc Gastroenterol. 2020; 3(Suppl_1): 153–5.
14. Vuurberg NE, Bakker I, van den Boom AL, de Haas RJ, Duiker EW, van den Heuvel MC, et al. Surgery for ampullary cancer in a patient with pancreatic lipomatosis caused by cystic fibrosis. Case Rep Gastroenterol. 2020; 14(3): 695–701.
15. Charles Bronson S, Suresh E, Stephen Abraham Suresh Kumar S, Mythili C, Shanmugam A. A novel synergistic association of variants in PTRH2 and KIF1A relates to a syndrome of hereditary axonopathy, outer hair cell dysfunction, intellectual disability, pancreatic lipomatosis, diabetes, cerebellar atrophy, and vertebral artery hypoplasia. Cureus. 2021; 13(2): e13174.
16. Satwik N, Koppad SN, Nadipally SR. A case report of complete pancreatic lipomatosis. Int Surg J. 2021; 8(6): 1921.
17. Meas V, Mam K, San C, Dara V, Lem D. Solid pseudopapillary tumor in a patient with pancreatic lipomatosis: a case report. HPB (Oxford). 2022; 24: S274–5.
18. Mahrous A-R, Al-Alfy MN, Abdel-Hassib M, Amin MA. Non-alcoholic Pancreatic Lipomatosis among the Egyptian Type 2 Diabetic Patients. Al-Azhar Int Med J. 2024; 5(5).
19. Rana SS, Ancil S, Jayanna SH, Kang M, Gupta R. Clinical features and outcomes of total pancreatic lipomatosis with chronic pancreatitis: a case series. Ann Gastroenterol. 2025; 38(1): 100–4.
20. Singla SP, Giri S, Das S, Panigrahi SC, Mallick B, Sahu MK. Total pancreatic lipomatosis presenting as pancreatic exocrine insufficiency. Indian J Gastroenterol. 2025.