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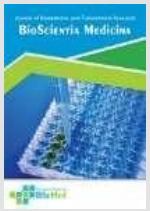

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The Clinical Spectrum and Management Patterns of Orbital Meningioma: A 6-Year Single-Institution Case Series from Indonesia

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A B S T R A C T

Background: Orbital meningiomas (OMs) are rare, complex tumors whose clinical course and management are debated. Data from Southeast Asian populations, particularly Indonesia, is scarce. This study aims to define the clinical spectrum, radiological features, and management trends of OMs at a single Indonesian tertiary referral center, with a focus on hormonal factors in a predominantly female cohort. **Methods:** A retrospective, descriptive case series was conducted on all patients diagnosed with orbital meningioma at Dr. M. Djamil General Hospital, Padang, Indonesia, from January 2018 to December 2023. Data on demographics, detailed clinical presentation (visual acuity, proptosis, extraocular movement), radiological findings (location, size, hyperostosis), and hormonal risk factors were collected. Management strategies, histopathological results (WHO grade), and follow-up outcomes were analyzed. **Results:** Twenty patients were included, with a significant female predominance (n=16, 80%; F:M ratio 4:1). The median age at diagnosis was 47 years (range 28-71). A strong association with hormonal factors was noted; 50% (8/16) of female patients had a history of exogenous hormonal contraceptive use. The most common presenting symptoms were progressive proptosis (n=15, 75%) and visual loss (n=12, 60%). The mean proptosis was 5.5 ± 2.1 mm. The most frequent tumor location was retrobulbar (intraconal) (n=11, 55%), followed by optic nerve sheath (n=5, 25%). A multidisciplinary, conservative-led approach was the primary management strategy, with 75% (n=15) of patients managed with active observation and neurosurgical consultation. Surgical intervention was performed in 25% (n=5) of cases, primarily for severe symptoms or aggressive radiological features. Histopathology (n=5) confirmed WHO Grade I in four cases (80%) and WHO Grade II (atypical) in one case (20%). **Conclusion:** Orbital meningiomas in this Indonesian cohort demonstrate a striking female predominance and a strong association with hormonal contraceptive use, suggesting a significant role for hormonal pathways in their pathophysiology. The management paradigm has shifted towards a multidisciplinary, observation-first approach, reserving surgical intervention for cases with documented progression, severe vision loss, or high-grade pathological features.

1. Introduction

Orbital meningiomas (OMs) are neoplasms arising from the arachnoid cap cells of the meninges, representing 3–9% of all orbital tumors.[1] These tumors pose a formidable challenge in ophthalmology and neurosurgery due to their location within the intricate and functionally dense orbital apex, their insidious clinical presentation, and their complex relationship with vital neurovascular structures, including the optic nerve. OMs are broadly classified into two distinct categories based on their origin. Primary OMs arise directly from intraorbital arachnoid cells, most commonly the optic nerve sheath (optic nerve sheath meningiomas, or ONSMs), accounting for approximately one-third of all primary optic nerve tumors.[2] Secondary OMs, which are more common,

originate intracranially and invade the orbit through contiguous structures, such as the sphenoid wing, optic canal, or superior orbital fissure.[3] Spheno-orbital meningiomas (SOMs) are the most frequent subtype of secondary OM, notorious for causing significant hyperostosis of the sphenoid bone, leading to progressive, disfiguring proptosis and compressive optic neuropathy.[4]

The clinical presentation of OMs is classically described as a triad of gradual, painless visual loss, progressive proptosis, and optic disc changes (either edema in early stages or atrophy in late stages).[5] However, the presentation can be highly variable, ranging from asymptomatic incidental findings to aggressive functional deficits, including diplopia from extraocular muscle restriction, afferent pupillary defects (RAPD), and complete blindness.

The pathophysiology of meningiomas, particularly their notable predilection for females (with reported female-to-male ratios as high as 10:1 in some series), remains a subject of intense investigation.[6] A substantial body of evidence points to a profound hormonal influence. The majority of meningiomas, especially WHO Grade I tumors, express progesterone receptors (PRs). This has led to the hypothesis that progesterone and exogenous progestins may act as key promoters of tumor growth. This hormonal link is further supported by reports of tumor enlargement during pregnancy or in patients receiving hormonal replacement therapy. In addition to hormonal factors, the inactivation of the neurofibromatosis type 2 (NF2) tumor suppressor gene on chromosome 22q12, which codes for the protein merlin, is the most common genetic alteration implicated in meningioma pathogenesis.[7]

The management of OMs has undergone a significant paradigm shift. Historically, aggressive surgical resection was the standard, often resulting in significant morbidity, including iatrogenic vision loss. The contemporary approach is far more nuanced, balancing the goals of tumor control, vision preservation, and quality of life.[8] This has led to a multidisciplinary strategy incorporating active observation ("wait-and-see") for slow-growing tumors, advanced surgical techniques for decompression or debulking, and the increasing use of stereotactic radiosurgery (SRS) or fractionated stereotactic radiotherapy (FSRT) as both a primary and adjuvant modality.[9]

While extensive literature exists on OMs from North American and European centers, there is a pronounced paucity of data from Southeast Asian populations. Genetic, environmental, and hormonal exposure patterns (such as contraceptive practices) can vary significantly across ethnicities, potentially influencing the clinical behavior and prevalence of these tumors. Reports from Indonesia, the world's fourth-most populous country, are particularly rare and are largely limited to individual case reports or small series that are not widely available in international literature.[10]

This study aims to characterize the comprehensive clinical spectrum, detailed radiological patterns, and definitive management strategies for orbital meningiomas at a single major Indonesian tertiary referral center. The novelty of this study lies in providing the first detailed clinicopathological and management series from this specific demographic, with a unique focus on analyzing potential hormonal risk factors in a cohort that was found to be predominantly female.

2. Methods

This study was a retrospective, descriptive case series conducted at the Department of Ophthalmology in collaboration with the Department of Neurosurgery and the Central Medical Records division at Dr. M. Djamil General Hospital, Padang, West Sumatra, Indonesia. Dr. M. Djamil General Hospital is the principal tertiary referral hospital for the province of West Sumatra and surrounding regions, serving a population of over five million. The study protocol adhered to the principles of the Declaration of Helsinki. Ethical approval was obtained from the Institutional Review Board (IRB) of the Faculty of Medicine, Universitas Andalas. Due to the

retrospective nature of the study, the requirement for individual patient consent was waived by the ethics committee. All patient data was anonymized prior to analysis to ensure confidentiality.

We performed a systematic search of the hospital's medical records database and the sub-specialty Oculoplastic and Oncology clinic database for all patients diagnosed with orbital meningioma between January 1st, 2018, and December 31st, 2023. The inclusion criterion was any patient with a definitive diagnosis of orbital meningioma confirmed by high-resolution computed tomography (CT) and/or magnetic resonance imaging (MRI) of the orbit and brain. The exclusion criteria were: (1) patients with incomplete or missing critical data in their medical records, (2) patients with radiological findings ambiguous or suggestive of other orbital pathologies (such as optic glioma, lymphoma, metastatic disease) without pathological confirmation, and (3) patients who were lost to follow-up immediately after the initial diagnosis.

A standardized data collection form was used to extract the following variables: (1) Demographics: Age at diagnosis, gender; (2) Risk Factor History: (i) Occupational History: Categorized as "Exposed" (such as history of work in industrial radiography, chemical manufacturing, or long-term agricultural pesticide use) or "Not Exposed" (such as homemaker, office worker, teacher); (ii) Hormonal History (for female patients): Parity, menopausal status, and history of exogenous hormonal use (type of contraceptive hormonal vs. non-hormonal, duration of use); (3) Clinical Presentation: (i) Symptomatology: Primary presenting complaint (proptosis, vision loss, diplopia, headache, pain) and duration of symptoms (in months) prior to diagnosis; (ii) Ophthalmic Examination: Best-corrected visual acuity (BCVA) converted to the logarithm of the minimal angle of resolution (LogMAR) for analysis, presence of a relative afferent pupillary defect (RAPD), and extraocular movement (EOM) limitations; (iii) Proptosis measurement: Hertel exophthalmometry reading (in millimeters) for the affected eye and the contralateral eye to determine the degree of proptosis; (4) Radiological findings: (i) imaging modality: CT, MRI, or both; (ii) Tumor Origin/Location: classified as primary optic nerve sheath meningioma (ONSM), secondary spheno-orbital meningioma (SOM), Retrobulbar (intraconal, non-ONSM), or Para/Suprasellar with orbital extension; (iii) Radiological Signs: Presence of characteristic features such as hyperostosis of the sphenoid bone, a "dural tail" sign, and optic canal involvement or stenosis; (iv) Tumor Metrics: Maximum tumor diameter (in millimeters); (5) Management: (i) Primary Strategy: Categorized as Active Observation with multidisciplinary consultation, Primary Surgical Intervention, or Primary Radiotherapy; (ii) Surgical Details: Type of procedure performed (lateral orbitotomy with debulking, exenteration, enucleation); (6) Histopathology: For all surgically excised specimens, the pathological diagnosis, WHO grade (I, II, or III), and histological subtype (meningothelial, fibrous, transitional, atypical) were recorded; (7) Follow-up: Median follow-up duration (in months), status of vision (improved, stable, worse), and evidence of tumor progression or recurrence on follow-up imaging.

All collected data were entered into a database using Microsoft Excel 2019 (Microsoft Corp., Redmond, WA) and subsequently analyzed using SPSS Statistics for Windows, Version 26.0 (IBM Corp., Armonk, NY). Descriptive statistics were used to summarize the data. Continuous variables (age, symptom duration, proptosis measurement, tumor diameter) were assessed for normality using the Shapiro-Wilk test. Normally distributed data were presented as mean ± standard deviation (SD), while non-normally distributed data were presented as median and interquartile range (IQR) or full range. Categorical variables (gender, symptom type, radiological location, management strategy, WHO grade) were presented as frequencies and percentages.

3. Results

Over the 6-year study period, a total of 20 patients met the inclusion criteria. A striking female

predominance was observed, with 16 female patients (80%) and 4 male patients (20%), yielding a female-to-male ratio of 4:1. The median age at diagnosis was 47 years (IQR 41.5–50.75; range 28–71 years). The peak incidence occurred in the 41–50 age group, accounting for 9 (45%) of all cases. A detailed breakdown of age distribution is presented in Table 1.

Table 1. Distribution of orbital meningioma cases by age and gender (n=20).

AGE GROUP (YEARS)	MALE (N=4)	FEMALE (N=16)	TOTAL (N=20)
< 30	0 (0%)	1 (6.3%)	1 (5%)
31–40	2 (50%)	2 (12.5%)	4 (20%)
41–50	1 (25%)	8 (50%)	9 (45%)
51–60	0 (0%)	4 (25%)	4 (20%)
61–70	1 (25%)	0 (0%)	1 (5We%)
> 70	0 (0%)	1 (6.3%)	1 (5%)
Total	4 (100%)	16 (100%)	20 (100%)

A significant history of exogenous hormonal use was identified. Of the 16 female patients, 8 (50%) had a history of using hormonal contraceptives (HC). Of these, 7 (87.5%) used progestin-only injectables (DMPA), and 1 (12.5%) used combined oral contraceptive pills. The mean duration of HC use was 7.2 ± 3.1 years. Two female patients (12.5%) used a non-hormonal intrauterine device (IUD). The remaining 6 (37.5%) had no history of contraceptive use. Among the female cohort, 12 (75%) were premenopausal at the time of diagnosis, and 4 (25%) were postmenopausal. Four patients (20%), all male, were classified as having potential occupational exposure (two in agricultural chemical handling, one in industrial radiography, and one in construction material manufacturing). The remaining 16 patients (80%), including all 16 female patients, were classified as "Not Exposed" (12 were housewives, 4 were office workers/teachers). Patients presented after a prolonged symptomatic period, with a median symptom duration of 18 months (IQR 9.5–24; range 3–60 months). The most common presenting complaint was progressive, painless proptosis, reported by 15 patients (75%). This was followed by subjective visual loss, reported by 12 patients (60%). Other complaints included diplopia (n=7, 35%), persistent headache

(n=5, 25%), and orbital pain (n=3, 15%). Detailed ophthalmic examination findings at presentation are summarized in Table 2. The mean proptosis in the affected eye was 5.5 ± 2.1 mm (range 2–10 mm) greater than the contralateral eye. A RAPD was present in 10 patients (50%), indicating significant optic nerve compromise. The mean presenting BCVA was 0.6 ± 0.4 LogMAR (approx. 20/80 Snellen), with 4 patients (20%) presenting with severe vision loss (worse than 1.0 LogMAR or 20/200).

Table 2. Summary of clinical presentation findings (n=20).

CLINICAL FINDING	FREQUENCY (N)	PERCENTAGE (%)
PRIMARY SYMPTOM		
Progressive Proptosis	15	75%
Visual Loss	12	60%
Diplopia / EOM Restriction	7	35%
Headache	5	25%
Orbital Pain	3	15%
OPHTHALMIC SIGN		
Measurable Proptosis (>2mm)	18	90%
Relative Afferent Pupillary Defect (RAPD)	10	50%
Extraocular Movement (EOM) Restriction	7	35%
Optic Disc Edema	6	30%
Optic Disc Atrophy	8	40%
Normal Optic Disc	6	30%

All 20 patients underwent high-resolution CT scans, and 12 (60%) also received an MRI with gadolinium contrast for better soft-tissue delineation. The mean maximum tumor diameter was 28 ± 11 mm (range 14–52 mm). The most frequent radiological location was retrobulbar (intraconal), identified in 11 cases (55%). These were tumors located posterior to the globe within the muscle cone, but did not demonstrate the classic "tram-track" sign of ONSM. Primary ONSM (with clear circumferential thickening of the optic nerve sheath) was identified in 5 cases (25%). Secondary spheno-orbital meningioma (SOM) with significant sphenoid wing involvement was seen in 3 cases (15%), and 1 case (5%) was a para/suprasellar tumor with secondary orbital apex extension.

Key radiological signs included the "dural tail" sign, which was identified in 14 cases (70%) on contrast-

enhanced MRI. Significant hyperostosis was a key feature in all 3 (100%) SOM cases and was also noted in 5 other cases (total 8/20, 40%). Optic canal stenosis was evident in 6 cases (30%), all of whom presented with visual loss. Radiological details are provided in Table 3.

Table 3. Radiological characteristics of orbital meningiomas (n=20).

RADIOLOGICAL FEATURE	FREQUENCY (N)	PERCENTAGE (%)
TUMOR LOCATION/ORIGIN		
Retrobulbar (Intraconal, non-ONSM)	11	55%
Optic Nerve Sheath (ONSM)	5	25%
Sphenoid Ridge (Secondary SOM)	3	15%
Para/Suprasellar (Secondary)	1	5%
KEY RADIOLOGICAL SIGNS		
Dural Tail Sign (on CECT, n=20)	14	70%
Calcification	9	45%
Bony Hyperostosis	8	40%
Optic Canal Stenosis/Involvement	6	30%

The primary management strategy for the majority of patients (n=15, 75%) was active observation with multidisciplinary consultation, primarily involving the neurosurgery department. This "wait-and-see" approach was typically chosen for patients with stable or mild vision loss, manageable proptosis, and no evidence of aggressive intracranial extension. Surgical intervention was the primary treatment for 5 patients (25%). The indications for surgery were severe and progressive vision loss (n=2), disfiguring proptosis with exposure keratopathy (n=2), and a blind, painful eye (n=1). The procedures performed were: (1) Lateral Orbitotomy (Krönlein) with Tumor Debulking (n=2, 10%): Performed for large, compressive retrobulbar tumors (one WHO Grade I Meningothelial, one WHO Grade I Fibrous); (2) Exenteration (n=2, 10%): Performed for massive, diffuse orbital involvement. One case was a blind eye with intractable pain (WHO Grade I Transitional), and the other was a radiologically aggressive tumor later confirmed as a WHO Grade II Atypical Meningioma; (3) Enucleation (n=1, 5%): Performed for a blind, painful globe secondary to a posterior ONSM causing central retinal artery occlusion (pathology confirmed WHO Grade I Meningothelial). No patients in this cohort received radiotherapy (SRS or FSRT) as a primary treatment, though 3 of the 15 observation-group patients were referred to a national radiotherapy center for consideration.

Histopathological confirmation was obtained for all 5 surgical cases. Four cases (80%) were classified as WHO Grade I (benign), with subtypes including

meningothelial (n=2), fibrous (n=1), and transitional (n=1). One case (20%) was confirmed as WHO Grade II (atypical), which correlated with the aggressive radiological presentation and the clinical decision for exenteration. No WHO Grade III (anaplastic) cases were identified.

The median follow-up duration for the entire cohort was 26 months (IQR 14–42 months). In the observation group (n=15), over a median follow-up of 24 months, visual acuity remained stable (within 0.1 LogMAR) in 12 (80%) patients. Two patients (13.3%) showed slow progression of visual loss, and one (6.7%) had documented radiological progression and was scheduled for radiosurgery. While in the surgical group (n=5), 2 patients who underwent orbitotomy and debulking reported improvement in proptosis and headache; vision stabilized in one and slightly improved in the other. Two patients who underwent exenteration and one patient who had enucleation were successfully fitted with orbital/ocular prostheses. Tumor recurrence was observed in 1 (20%) of the surgical patients: the patient with the WHO Grade II atypical meningioma experienced a local recurrence at 18 months post-exenteration and was referred for adjuvant radiotherapy.

4. Discussion

The findings of this six-year institutional case series, while based on a cohort of 20 patients, provide critical insights into the clinical, etiological, and management patterns of orbital meningioma (OM) within an Indonesian population. Our discussion focuses on a deep analysis of the data, organized into three key domains: (1) The powerful etiological evidence for a hormono-genetic driver, (2) The clinical-radiological correlation defining the disease's natural history and presentation, and (3) The profound implications of our institution's conservative, multidisciplinary management philosophy.[11]

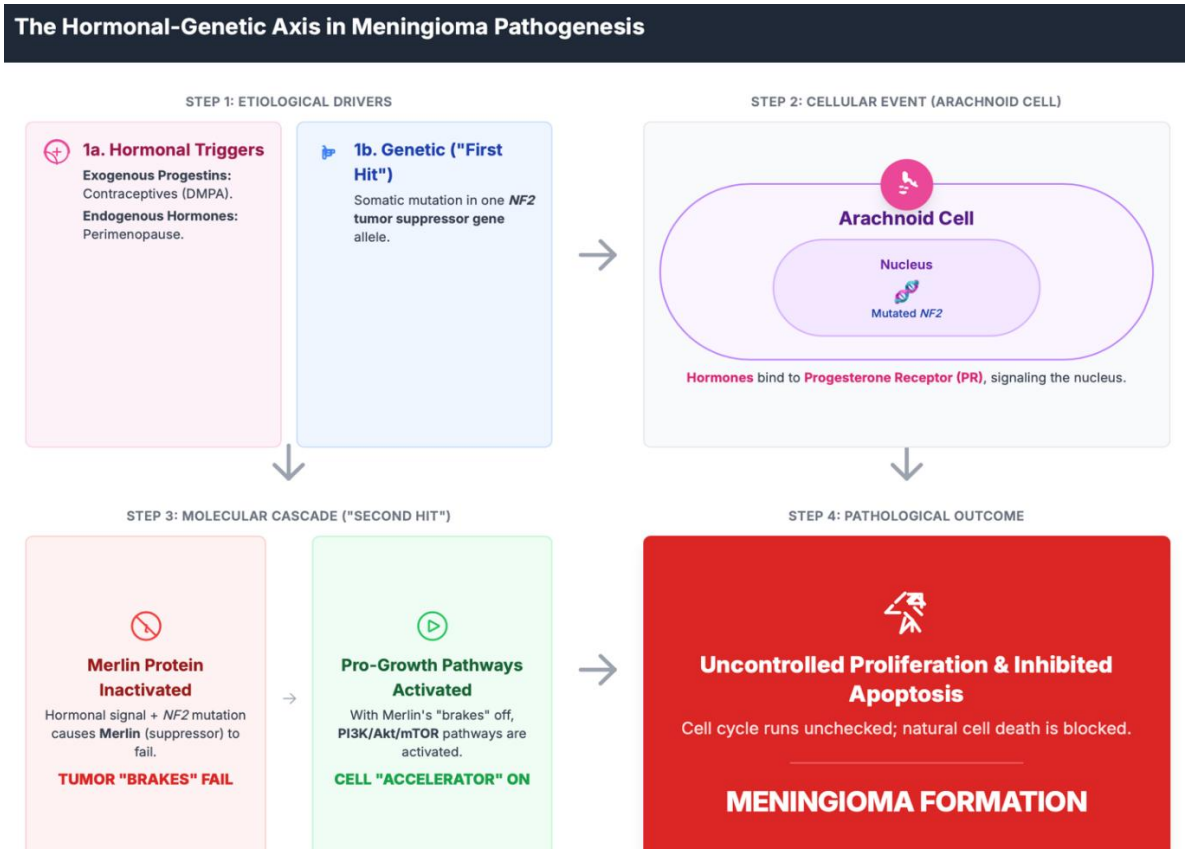

Figure 1. The hormonal-genetic axis in meningioma pathogenesis.

The most significant finding of our study is the profound female predominance, with women accounting for 80% of cases, yielding a 4:1 female-to-male ratio.[12] This ratio is more pronounced than the 2:1 or 3:1 ratios commonly reported in many Western cohorts, suggesting a potentially amplified etiological driver within our specific population. This demographic skew, when combined with our finding that 50% of all female patients (40% of the total cohort) had a history of exogenous hormonal contraceptive use, strongly implicates hormonal signaling in meningioma pathogenesis.

Meningiomas are, by definition, hormonally responsive tumors. The link is most irrefutably established with progesterone.[13] A vast body of literature confirms that between 70% and 100% of benign (WHO Grade I) meningiomas express high levels of progesterone receptors (PRs). In contrast, estrogen receptors (ERs) are expressed far less frequently, typically in only 10-30% of cases, and their role in tumorigenesis remains ambiguous. Androgen receptors (ARs) are found in an even smaller subset. Our findings compel us to focus on the progesterone pathway as the primary engine of tumor growth in this cohort.

The molecular pathophysiology is complex but centers on the interaction between PR signaling and the NF2 tumor suppressor gene.[14] The binding of progesterone (or a synthetic progestin) to its receptor—particularly the PR-B isoform, which is more associated with proliferation—initiates a cascade. The activated receptor dimerizes, translocates to the nucleus, and binds to Progesterone Response Elements (PREs) on target genes. This activation promotes tumorigenesis by upregulating key cell-cycle proteins, such as Cyclin D1 and c-Myc, while simultaneously inhibiting apoptosis through factors like Bcl-2.

However, this hormonal signaling does not occur in a vacuum. It interacts directly with the NF2 gene, the most commonly mutated gene in sporadic meningiomas. The NF2 product, merlin, is a critical "stop" signal in the cell. Located at the cell membrane, merlin integrates extracellular cues and inhibits multiple pro-growth pathways, most notably the PI3K/Akt/mTOR and Ras/MAPK cascades. In a healthy arachnoid cell, Merlin is active and keeps proliferation in check. In meningioma, this "merlin-off" switch is constitutively active.

The synergy between progesterone and NF2 dysfunction is the key. Progesterone signaling has been shown to dysregulate merlin function, either by directly downregulating NF2 gene expression or by promoting the phosphorylation (and thus, inactivation) of the merlin protein.[15] This creates a "two-hit" model perfectly suited to our cohort: an individual may harbor a "first hit" (a somatic mutation in one *NF2* allele) in a meningeal cell rest. This cell lies dormant until a "second hit" occurs. In our patients, this second hit appears to be a potent, non-genetic promoter: a sustained supraphysiological level of progestins. This hormonal surge effectively inactivates the remaining merlin function, unleashing the PI3K/Akt/mTOR pathway and driving relentless, pathological cell growth.

Our data provides powerful clinical support for this model. The finding that 50% of our female patients used hormonal contraception is critical, but the type of contraception is even more illuminating. The majority of these patients used depot medroxyprogesterone acetate (DMPA), a potent, long-acting synthetic progestin. This is not a subtle physiological signal; it is a powerful, iatrogenic pharmacological accelerant. This aligns perfectly with a landmark 2019 study by Supartoto et al., also in an Indonesian cohort, which demonstrated a direct relationship between exogenous progesterone use and lower NF2 mRNA expression, providing the molecular "smoking gun." This is further corroborated by large-scale European epidemiological data. A 2021 Danish nationwide cohort study, for example, found a more than 7-fold increased risk of intracranial meningioma associated with the long-term use of high-dose progestin-only therapies, confirming their role as potent tumor promoters.

This hormonal hypothesis is further triangulated and reinforced by our occupational data. A full 80% of our cohort (and 100% of the female patients) were homemakers. This is a crucial finding of exclusion. It effectively rules out the only definitively proven environmental risk factor for meningioma: ionizing radiation. Furthermore, it minimizes the potential confounding role of occupational chemical exposures (solvents, pesticides, formaldehyde) that have been weakly and inconsistently associated with meningiomas in other studies. The absence of these environmental factors in our cohort strongly shifts the etiological focus away from external carcinogens and reinforces the hypothesis of an endogenous or iatrogenic (hormonal) driver.[16]

Finally, the peak age of incidence in our cohort (41-50 years) is not coincidental. This bracket corresponds precisely to the perimenopausal transition. This life stage is characterized by significant hormonal chaos, including anovulatory cycles that lead to periods of unopposed estrogen and, more importantly, erratic and dysregulated progesterone secretion. This endogenous hormonal volatility, combined with the iatrogenic progestin exposure, creates a "perfect storm" of proliferative signaling, providing the ideal fertile ground for the growth of pre-existing, subclinical meningeal cell rests.

The clinical and radiological findings in our series paint a clear, cohesive picture of a slow-growing, insidious disease that is often diagnosed late, only after significant and often irreversible functional damage has occurred. The median symptom duration of 18 months highlights this diagnostic delay. Orbital meningiomas are classic "silent" tumors. The orbit, a 30cc space, can accommodate a slow-growing mass for months or even years. The globe is gradually pushed forward (proptosis) with such indolence that the brain's plasticity often adapts, preventing diplopia until late in the disease. Patients and their families may not notice the slow, asymmetric facial changes.[17]

This "accommodation" phase inevitably gives way to compressive optic neuropathy (CON). Our data clearly illustrates the devastating timeline of CON. The initial insult is axoplasmic stasis from compression and, critically, ischemia from compromised pial blood vessels. This manifests as optic disc edema, which was present in 30% of our cohort—likely representing the "earlier" presenters. As compression persists, axons begin to die. This is the inflection point where an objective, afferent neurological deficit appears. Our finding that 50% of patients already had a Relative Afferent Pupillary Defect (RAPD) at their first consultation is a stark indicator of late presentation. A RAPD signals significant, asymmetric axonal loss. The end-stage of this process is optic disc atrophy, found in 40% of our patients. Atrophy represents widespread, irreversible axonal death. The triad of cardinal symptoms—proptosis (75%) and visual loss (60%)—is therefore a late finding, not an early one.

Our radiological data correlates perfectly with this clinical picture. The 30% rate of optic canal stenosis is a critical prognostic finding. This bony "choke point" at the orbital apex is where the optic nerve is most vulnerable. It is no surprise that this 30% stenosis rate correlates directly with the 60% rate of visual loss and 50% rate of RAPD. The compression at this unyielding anatomical bottleneck is the primary mechanism of blindness. The distribution of tumor locations in our series is also illuminating. The high rate (55%) of "retrobulbar (intraconal)" tumors is a noteworthy finding. This classification is radiologically distinct from the classic pathognomonic "tram-track" sign of an optic nerve sheath meningioma (ONSM), which constituted 25% of our cohort. The ONSM involves a circumferential, sheath-like encasement of the nerve. It is also distinct from the classic spheno-orbital meningioma (SOM), which represented 15% of our cases. SOMs are typically *en plaque* (carpet-like) tumors along the sphenoid wing, whose primary orbital manifestation is not the tumor mass itself, but the severe, reactive bony hyperostosis it induces. Our finding that 40% of the total cohort had hyperostosis is a testament to the prevalence of this sphenoid wing involvement.[18]

The large "retrobulbar" category (55%) suggests that a significant proportion of primary OMs in our

population arises from arachnoid cells of the orbital soft tissue or from the optic nerve sheath in a more globular, eccentric, or diffuse fashion, rather than the classic circumferential pattern. This distinction is not merely academic; it is profoundly surgical. A globular, displacing tumor is potentially resectable with nerve-sparing, whereas a truly encasing ONSM is not.[19] The high prevalence of the "dural tail" sign (70%)—representing reactive, enhancing dura mater rather than frank tumor—and the presence of calcification (45%) are classic hallmarks that underscore the high diagnostic utility of contrast-enhanced CT and MRI.

Perhaps the most telling finding regarding clinical practice at our tertiary center is the management strategy. A remarkable 75% of patients (15 of 20) were managed with active observation and multidisciplinary consultation, primarily with neurosurgery. This finding does not represent therapeutic nihilism; rather, it reflects a mature, evidence-based paradigm shift away from the historical dogma of mandatory, aggressive surgical extirpation. The rationale for this conservative approach is twofold. First, we now understand the natural history of most OMs. As our histopathological data confirms, the vast majority (80% of our surgical cohort, and likely a similar proportion of the observed cohort) are WHO Grade I. These tumors grow exceptionally slowly—often at rates of 1-2 mm per year—and may stabilize for decades. Second, the morbidity of surgical intervention in the orbit is exceptionally high. The optic nerve and its delicate pial vascular supply are exquisitely sensitive. Attempts at "gross total resection" of an encasing ONSM, an ambition of a bygone surgical era, almost invariably result in stripping this blood supply and causing complete, permanent iatrogenic blindness.[20]

Therefore, the modern management philosophy, as clearly practiced in our center, is one of functional preservation. Observation is the preferred course for patients with good or stable vision, regardless of tumor size. Surgical intervention, which comprised 25% (n=5) of our series, is now reserved for specific, function-threatening indications: (1) progressive, severe vision loss directly attributable to compression, (2) disfiguring proptosis leading to exposure keratopathy, globe subluxation, or severe cosmetic deformity, (3) intractable orbital pain, or (4) radiological or clinical suspicion of a high-grade tumor.

Our surgical data directly reflects this philosophy. The interventions were not elective but necessary. They included lateral orbitotomies for decompression and debulking (n=2), enucleation for a blind, painful eye (n=1), and exenteration (n=2). The histopathological data from these cases provides the final piece of the puzzle. Our one case of a WHO Grade II (atypical) meningioma—representing 20% of the surgical cohort—was found in a patient who required exenteration. This is a vital clinical-pathological correlation. Atypical meningiomas, defined by increased mitotic activity (4-19 mitoses per 10 high-power fields) or specific histological features (sheet-like growth, high cellularity), have a much higher rate of recurrence and aggressive behavior. This single case, which correlated with a more aggressive clinical course (rapid progression, pain), highlights the absolute necessity of obtaining tissue for diagnosis in radiologically aggressive or rapidly progressing tumors. It provides the crucial counter-argument to a purely observational approach.[17,18]

Finally, the "Observation + Neurosurgery Consult" (75%) category is itself illuminating. It highlights that OM is no longer—and should never have been—considered a purely ophthalmological disease. It is a "borderland" tumor that respects no specialty. The high rate of consultation underscores the complex, intracranial nature of these lesions. Even primary OMs can and do extend through the optic canal to involve the chiasm. The neurosurgery consultation is essential for evaluating intracranial extension, managing the spheno-orbital and skull base components, and planning potential combined-approach surgeries (e.g., frontotemporal craniotomy with orbital decompression). This team-based approach, integrating Oculoplastic/Orbital Surgery, Neurosurgery, Neuro-ophthalmology, and Radiology,

is the unequivocal standard of care.

A notable finding in our series is the absence of primary radiotherapy as a treatment modality, with radiosurgery only being a consideration upon referral. This likely reflects resource availability and established treatment protocols within our specific regional center. This, however, represents a significant "treatment gap" when compared to global standards. Stereotactic radiosurgery (SRS) and, more importantly, fractionated stereotactic radiotherapy (FSRT) have become mainstays of OM management. FSRT, in particular (50.4 Gy in 28 fractions), is now the primary treatment of choice in many world-class centers for ONSM with progressive vision loss. Its mechanism is not cytotoxic but radiostatic; it halts tumor growth by inducing DNA damage and obliterating the tumor's microvascular supply. By fractionating the dose, the optic nerve is allowed to repair between sessions, leading to high rates (80-90%) of long-term tumor control while preserving or even *improving* vision in a significant subset of patients. The development of a dedicated orbital radiosurgery program represents the clearest and most critical area for future advancement in our center's management algorithm.[19,20]

This study, while providing novel data, has limitations inherent to its design. The retrospective nature can lead to incomplete data and selection bias. Our sample size (n=20) is small, which limits the statistical power and generalizability of our findings, although it is a respectable cohort for such a rare tumor at a single institution. The lack of histopathology for 75% of patients in the observation group means their diagnoses are based on radiological criteria alone, which, while highly accurate, is not definitive. Follow-up was variable, and a longer-term analysis is needed to truly understand the natural history and recurrence rates in our observation-heavy cohort.

5. Conclusion

This 6-year case series from Indonesia provides a unique and valuable snapshot of orbital meningioma in a Southeast Asian population. Our findings confirm its status as a disease predominantly affecting middle-aged females, and we provide strong clinical evidence supporting the critical role of hormonal pathophysiology, particularly the association with exogenous progestin-based contraceptives. The clinical presentation in our cohort is often delayed, with significant functional compromise already present at diagnosis. Radiologically, a large proportion of tumors are retrobulbar and intraconal, in addition to the classic ONSM and SOM subtypes. Most importantly, our study demonstrates a clear management paradigm favoring a multidisciplinary, conservative, "wait-and-see" approach, with 75% of patients managed by observation and neurosurgical consultation. Surgical intervention is reserved for severe functional decline or aggressive pathological features. This study lays the groundwork for future, larger, multi-center prospective studies in Indonesia to further elucidate the unique etiological factors and optimize management for this complex disease.

6. References

1. Dallan I, Serioli S, de Notaris M, Pasquini E, Zoli M, Mazzatenta D, et al. Transorbital management of selected spheno-orbital meningiomas: Insights from a multicentric retrospective Italian study of 52 cases. Neurosurg Rev. 2025; 48(1): 696.
2. Drosos E, Hammerbeck-Ward C, King AT, Pathmanaban ON, Attaulah S, Cannon PS, et al. Multidisciplinary management of spheno-orbital meningiomas. J Neurol Surg B Skull Base. 2025; 86(S 01): S1–576.
3. Ishaque AH, Cusimano MD. Spheno-orbital meningiomas: Case series and surgical outcomes following frontotemporal-orbitozygomatic craniotomy. J Neurol Surg B Skull Base. 2025; 86(S 01): S1–576.
4. Düzkalir AH, Samanci Y, Askeroglu O, Peker S. Optimizing outcome in radiosurgery for spheno-orbital meningioma: the critical role of

planning quality. Illustrative case. J Neurosurg Case Lessons. 2025; 9(15).
5. Steinmetz M, Lebeau J, Bouchain O, Moïse M, Bianchi E, Andris C, et al. Paradoxical evolution of spheno-orbital meningioma after cessation of progestin treatment. Surg Neurol Int. 2025; 16: 151.
6. Westarp E, Sharma N, Thieringer FM, Roethlisberger M. Spheno-orbital meningiomas: Advances in reconstruction workflow and cosmetic/functional outcomes in the digital era. Oral Maxillofac Surg Clin North Am. 2025; 37(3): 517–27.
7. Porto E, Carone G, Fiore G, Del Bene M, Galbiati T, Barbotti A, et al. Spheno-orbital meningiomas: predictors of recurrence and novel strategies for surgical management. J Neurosurg. 2025; 143(2): 365–74.
8. Costanzo R, D'Andrea M, Manfrellotti R, Nielsen JK, Tafuto R, Tomassini A, et al. Endoscopic transorbital approach and transcranial approach in spheno-orbital meningiomas: a comparative qualitative and preliminary quantitative anatomical study. J Clin Med. 2025; 14(19): 6744.
9. Rosito D, Plazola S. Management of orbital meningiomas and associated proptosis. J Neurol Surg B Skull Base. 2016; 77(S 01).
10. Patrick S, Tat YB, Amin MZM, Addenan M, Ab.Ghani S, Hanafi H. Atypical orbital primary optic nerve sheath meningioma with severe disfiguring proptosis: an alternative surgical approach. Med J Indones. 2023.
11. Wilson DS, Mohammad S, Hussain A. A novel report of suspected prostate adenocarcinoma to orbital roof meningioma metastasis. Ophthal Plast Reconstr Surg. 2023; 39(5): e166–8.
12. Vishwanath P, Backiavathy V, Subramanian K. Atypical presentation of sphenoid wing meningioma mimicking idiopathic orbital inflammation. Indian J Ophthalmol Case Rep. 2024; 4(4): 911–3.
13. Satrio J, Nugroho T. Characteristic of patients with orbital meningioma in Dr. Kariadi General Hospital Semarang. Ophthalmologica Indonesiana. 2024; 49(S2).
14. Tan JL, Dhepnorrarat C, Wong D, De Sousa J-L. Transorbital and endonasal resection of a rare orbital ectopic atypical meningioma. BMJ Case Rep. 2024; 17(3): e257490.
15. Murayi R, El-Abtah M, Xiao T, Recinos PF, Kshettry VR. Orbital reconstruction and volume in the correction of proptosis after resection of spheno-orbital meningiomas. J Neurosurg. 2024; 140(5): 1305–11.
16. Janah R, Rujito L, Wahyono DJ. Differences in Merlin and p53 expression as a predisposing factor in orbital meningioma. J Neurosci Rural Pract. 2024; 15(477): 477–83.
17. Li X, Chen Z, Mao G, Han Y. Curative effect of microscopic resection through the orbital-pteral approach for cranial orbital communicating meningioma invading the supraorbital fissure pathway. Asian J Surg. 2025; 48(6): 4062–3.
18. Lucarelli K, Kim W, Samarage HM, Rootman D. Endoscopic transorbital resection of spheno-orbital meningiomas for orbital decompression. J Neurol Surg B Skull Base. 2025; 86(S 01): S1–576.
19. Baig A, Ong A, Doyle J, Sipkova Z. Meningioma masquerading as orbital inflammation. EYE. 2025; 39(Suppl 1): 87.
20. Korkar GH, Alkarras MT, Rizk MM, El Saied BH, Mousa AM. Orbital reconstruction after spheno-orbital meningiomas excision with fascia Lata and fat graft. Ain Shams Med J. 2025; 76(2): 495–506.