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Head and Neck Sarcomas Characteristics at Dr. Hasan Sadikin General Hospital for 5 Years (2017-2021)

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ABSTRACT

Background: Sarcoma is a malignant neoplasm originating from mesodermal tissue with an incidence rate of 1% of neoplasms throughout the body. Head and neck sarcoma requires more complex management and until now, reviews of this disease are still very rare. This study aimed to report the incidence and characteristics of head and neck sarcoma during 2017-2021 at Dr. Hasan Sadikin General Hospital, Universitas Padjadjaran, Bandung. Methods: Research data was taken from patient medical records through SIRS at Dr. Hasan Sadikin General Hospital during the 2017-2021 period according to the ICD10 of head and neck malignancies and histopathological data. Results: During 2017-2021, 15 cases of head and neck sarcoma were found, consisting of 9 cases (60%) rhabdomyosarcoma, 2 cases (13.3%) fibrosarcoma, and 1 case each (6.7%) synovial sarcoma, undifferentiated pleomorphic sarcoma, angiosarcoma dermatofibrosarcoma protuberans. The neck and shoulder area are the most frequently affected site (40%), followed by the scalp (26.7%), maxilla (20%), and oral cavity (13.3%). The highest incidence rates were in female (60%) and children aged <17 years (46.7%). Conclusion: The incidence of head and neck sarcoma at Dr. Hasan Sadikin General Hospital Bandung is rare. The most frequent cases are rhabdomyosarcoma the most commonly affected areas are the neck and shoulders.

1. Introduction

Sarcomas are rare malignant neoplasms, which account for only 1% of all malignancies. 1-3 Sarcoma is a solid tumor originating from mesenchymal tissue originating from bone or soft tissue with an annual incidence of 5/100,000.1,4,5 The predilection can occur throughout the body, and a very rare predilection is the head and neck, which is 5-15% of all sarcomas and 1% of all head and neck malignancies. 3,4,6,7 According to the 2020 WHO classification, there are as many as 70 sub-classifications of sarcomas with their own characteristics. 5 According to a study by Bentz et al., the subtype that most often occurs in the head and neck respectively is pleomorphic sarcoma (or malignant fibrous histiocytoma-MFH), fibrosarcoma,

angiosarcoma, malignant peripheral nerve sheath tumor and non-classified/non-differentiated sarcoma.^{4,8} Meanwhile, Pandey et al. revealed that the most common head and neck sarcoma is rhabdomyosarcoma (RMS), followed by pleomorphic sarcoma, malignant schwannoma, angiosarcoma, liposarcoma, and neurofibrosarcoma.²

The main treatment for sarcoma is wide excision with adequate excision margins to prevent local recurrence. Administration of adjuvant radiation can improve local control. Even so, the management of head and neck sarcoma has its own challenges considering the many vital and important structures in that area, so often the treatment involves multidisciplinary¹. There has never been data on head

and neck sarcomas in Indonesia, especially at the Dr. Hasan Sadikin General Hospital. This study aimed to report the characteristics of head and neck sarcoma patients for 5 years (2017-2021) who were treated at Dr. Hasan Sadikin General Hospital, Bandung.

2. Methods

This study was a descriptive observational study and used secondary data obtained from medical record data at the medical records installation of Dr. Hasan Sadikin General Hospital, Bandung, Indonesia. A total of 15 cases of head and neck sarcoma were included in this study. The subjects of this study met the inclusion criteria. The inclusion criteria in this study were patients who were treated at Dr. Hasan Sadikin General Hospital Bandung, Indonesia, for the 2017-2021 period and were diagnosed with head and neck sarcoma, and the research subjects had complete medical record data. This study made observations on sociodemographic data, clinical data, and laboratory data. Data analysis was performed using SPSS software version 25. Univariate analysis performed to present data for each test variable by frequency distribution.

3. Results

During 2017-2021, only 15 cases of head and neck sarcoma were found seeking treatment at RSHS. Incidence in women more than men (3:2). Based on age at the time of diagnosis, the most were children aged <17 years (7 cases - 46.7%) followed by young adults 17-40 years (3 cases - 20%), old adults 41-60 years (2 cases - 13.3%) and elderly > 60 years (3 cases-20%). Based on the tissue origin, there were 9 cases of sarcoma originating from striated muscle (60%), 3 cases sarcoma originating fibroblasts/myofibroblasts (20%), 2 cases of sarcoma belonging to the category uncertain differentiation (13.3%) and 1 case of sarcoma originating from blood vessels (6.7%). Based on the histopathological results, there were 9 cases of rhabdomyosarcoma (60%), 2 cases of fibrosarcoma (13.3%), and 1 case each (6.7%) synovial sarcoma, undifferentiated pleomorphic sarcoma, angiosarcoma, and dermatofibrosarcoma protuberans. Based on the site of predilection, the most commonly affected areas were the neck and shoulders in 6 cases (40%), followed by 4 cases in the scalp (26.7%), 3 cases in the maxilla (20%), and 2 cases in the oral cavity (13.3%).

Table 1. Characteristics of head and neck sarcoma patients in 2017-2021 at Dr. Hasan Sadikin General Hospital, Bandung.

Histopathology type	Network origin	Number of patients	Gender	Age	Predilection
Rhabdomyosarcoma	Skeletal muscle	9	Male	1 year	Neck
(60%)	(60%)		Male	2 years	Labium oris
			Female	6 years	Scalp
			Female	10 years	Maxilla
			Male	12 years	Maxilla
			Female	13 years	Maxilla
			Male	14 years	Shoulders
			Female	28 years	Shoulders
			Female	75 years	Scalp
Fibrosarcoma (13,3%)	Fibroblast/	2	Female	29 years	Shoulders
	myofibroblast		Male	70 years	Buccal
Dermatofibrosarcoma protuberans (6,7%)	(20%)	1	Female	73 years	Neck
Synovial sarcoma (6,7%)	Tumor of	1	Female	23 years	Neck
Undifferentiated	uncertain	1	Male	52 years	Scalp
Pleomorphic sarcoma	differentiation			-	
(6,7%)	(13,3%)				
Angiosarcoma (6.7%)	Vascular (6.7%)	1	Female	48 years	Scalp

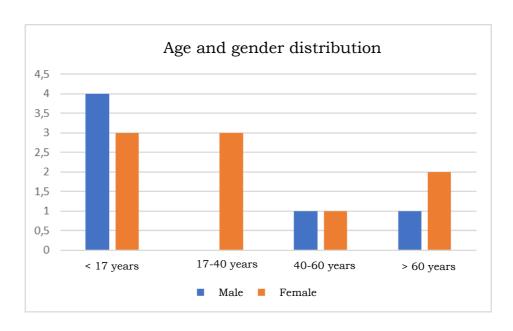


Figure 1. Age and gender distribution of head and neck sarcomas.

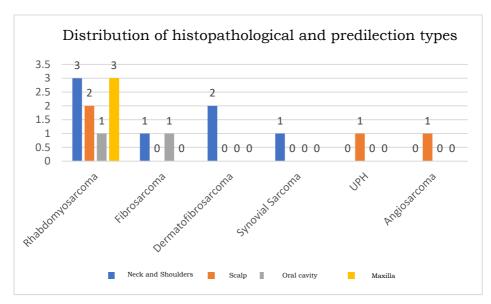


Figure 2. Distribution of histopathological types and predilection for head and neck sarcoma.

4. Discussion

In this study, it was found that the incidence of head and neck sarcomas in females was higher than in males (3:2). This is inconsistent with studies in Australia, Europe, and India, which state that there are more males than females.^{1,2} Based on the type of histopathology, rhabdomyosarcoma is the most common with the majority of children, namely 7 cases between the ages of 1 year to 14 years. From this

population, the sex of the majority is male. According to research in Australia and India, the most common type of head and neck sarcoma in children is rhabdomyosarcoma. 1–3 Likewise, research on M.D. Anderson Cancer Health Center University of Texas in 1970-1989 stated that the most common head and neck sarcomas were rhabdomyosarcomas, with the majority being children. Nonetheless, the highest predilection in this study was the pharynx and orbit,

followed by the oral cavity, paranasal sinuses, neck, and ears. This is different from this study, where the most common predilection sites for rhabdomyosarcoma are the maxilla, neck, and shoulders, followed by the oral cavity and scalp.

Fibrosarcoma is the second most common head and neck sarcoma in this study. There are studies that say that adult fibrosarcoma ranks first most often, but there are also those that rank second. 1,8,9 Most head and neck fibrosarcomas appear at the age of 40-50 years, with an age range between 14-72 years. 3,10 There are also studies that say the age of predilection for fibrosarcoma is the age of 25-79 years, with the most frequent age being between the ages of 30-60 years. 11 In this study, there were 2 cases of fibrosarcoma, where 1 case was 28 years old, and the other case was 75 years old. Although rare, fibrosarcoma can also occur in old age.

Some studies say that the most common head and neck sarcomas are malignant fibrous histiocytoma (MFH), which, according to WHO classification in 2021, is included in undifferentiated pleomorphic sarcoma (UPH).1,2,6,9 In this study, only 1 UPH case was found aged 52 years. The age of occurrence of UPH is in accordance with the literature, where the highest incidence of UPH in the head and neck is between the ages of 46 -> 66 years, although it can also occur at a young age. 10 Dermatofibrosarcoma protuberans (DFTSP) only found 1 case aged 73 years. Head and neck DFTSP often occurs in young adults (16-45 years), even up to the age of 65 years. 10 Likewise, cases of synovial sarcoma and angiosarcoma were only 1 case each. According to the literature, synovial sarcoma can be found at the age of 6-25 years, while angiosarcoma can be found at the age of 46 -> 66 years10. In this study, the age of synovial sarcoma patients was 23 years, and 48 years of age for angiosarcoma patients. This is in accordance with the existing literature.

Rhabdomyosarcoma, fibrosarcoma, synovial sarcoma, angiosarcoma, and UPH are included in the high-grade sarcoma. Meanwhile, dermatofibrosarcoma protuberans are included in the

low-grade sarcoma.^{3,6} In this study, only 1 case (6.7%) included a low-grade sarcoma, while 93.3% were highgrade. Tejani et al., in their research, revealed that 77% of head and neck sarcomas were of high grade⁷, while other studies mention 56% high grading and 44% low grading.⁸ Overall, the most predilection sites for head and neck sarcoma in this study were the neck and shoulders, followed by the scalp, maxilla, and oral cavity. This is in accordance with the research of Bentz et al., who conducted a retrospective study by taking data from Memorial Sloan-Kettering Cancer Center 1973-1999 stated that the most common anatomical location was the neck, followed by the face, scalp, oral cavity and nasopharynx.⁸

5. Conclusion

Head and neck sarcoma is rare. Based on data obtained at Dr. Hasan Sadikin General Hospital Bandung, the most common type is rhabdomyosarcoma, followed by fibrosarcoma, and the most commonly affected predilection sites are the neck and shoulders.

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