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Original Article

Clinicopathology Profile and Post–Microsurgical Outcome of Sphenoorbital Meningioma: Single Institution Experience

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Abstract

Introduction: Meningiomas are among the most common types of intracranial neoplasm. Sphenoorbita meningioma usually affect anatomical and functional around the orbits. This study aimed to analyse the epidemiological distribution and clinical data of patients with sphenoorbital meningioma who underwent tumor resection microsurgery with a focus on surgical outcomes.

Patients and Methods: This is a retrospective descriptive study conducted in Dr. Cipto Mangunkusumo Hospital as a national referral hospital in Indonesia between 2018 and 2023. Epidemiological data and clinical status were collected through medical record.

Results: There were 66 patients included in this study, majority of patients were women (93.9%), with a female–to–male ratio of 15.5:1. The mean age at tumor resection surgery was 44.68 ± 7.8 years old, with the majority of patients in the 41–50 age group (53.0%). Tumor resection in sphenoorbital meningioma presented fairly good outcomes depending on the degree of resection, with optimum improvement in structural symptoms, such as proptosis (100%), but debatable results in functional symptoms, such as visual acuity (6.1% improvement and 83.3% stabilization).

Discussion: Sphenoorbital meningioma is a common intracranial neoplasm that can cause visual disturbances, proptosis, and other neurological symptoms. Surgical resection is the definitive treatment and the degree of total resection is commonly measured using the Simpson grading system.

Conclusion: Understanding patient outcomes is crucial to improve surgical techniques and reduce postoperative complications. This study provides valuable epidemiological and clinical data as a basic knowledge for further research about surgical management in sphenoorbital meningioma patients.

Keywords: Sphenoorbital meningioma, microsurgery, CNS tumor, tumor resection

Introduction

Meningiomas are among the most common types of intracranial neoplasm. They originate from meningeal tissue, particularly the dural layer, and can form in various locations around the brain and spine. Sphenoorbital meningiomas arise around the sphenoid bone, orbit, and the optic canal. Symptoms of sphenoorbital meningioma are largely determined by tumor extension and compression of the surrounding organs. Owing to the location of the tumor around the eye, the most common triad of symptoms include visual disturbances, progressive proptosis, and visual field disturbances. Other rare symptoms include general neurological disturbances, such as headache, seizures, memory impairment, or cranial nerve palsy, especially the oculomotor nerve to the cochleovestibular nerve.[1,2]

Sphenoorbital meningiomas are benign tumors that are classified as WHO–I or WHO–II; however, they can be radiologically malignant because of their high degree of invasion, with potential extension to the fossa media, superior orbital fissure, anterior clinoid, m. temporalis, m. lateral pterygoid, and cavernous sinus. Tumor resection is a common definitive management method to completely remove the mass; however, total resection cannot always

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be performed because of the complex anatomical structure and the presence of important neurovascular structures in the vicinity. Such drawbacks increase the importance of understanding the outcomes of microsurgery in patients with sphenoorbital meningioma, especially in Dr. Cipto Mangunkusumo Hospital.[1,2] The objective of this study was to present the epidemiological distribution and clinical data, especially the surgical outcomes, of sphenoorbital meningioma treatment.

Patients and Methods

Study Method

This study was conducted as a retrospective descriptive study which explored epidemiological data and clinical status of patients with sphenoorbital meningioma. Every patient underwent tumor resection surgery as the definitive treatment, as the study continued to focus on clinical outcomes on patients after surgery.

Secondary data were obtained only from medical records, and observation was performed purely without direct intervention by the subjects. All patient diagnosed with sphenoorbital meningioma from clinical and radiological data were included in this study. However, if there is missing of variable data in medical record, subject were excluded. Consecutive sampling was used, as all patients who met the inclusion and exclusion criteria were selected as study subjects.

Patient Population

The study selected patients who underwent tumor resection for sphenoorbital meningioma at Dr. Cipto Mangunkusumo Hospital between 2018–2023. During this period, microsurgery was performed on 66 patients.

Surgical Technique

Adequate history–taking and physical examination were completed primarily to assess the clinical status and management strategy, especially for surgical considerations. Preoperative radiologic examination was performed using Computed Tomography (CT) of the brain to calculate the hyperostosis, and Magnetic Resonance Imaging (MRI) of the brain to evaluate the tumor mass and its proportion intradural and intraorbitally. CT of the orbit can also be performed to measure proptosis index in appropriate cases.

The patient was operated on in a supine position under general anaesthesia in supine position. Mayfield head clamp was used to fix the head in a suitable position according to the tumor location, and aseptic and antiseptic steps were performed. Dissection was performed to expose the surgical area until the neurosurgeon achieved optimum visual field of the tumor. Craniotomy using pterional approach or extended to cranio–orbito–zygomatic approach for tumor removal depend on the extension of the tumor. Occasionally, pathological bone was resected and drilled until the dura was exposed. The tumor was resected carefully considering the border of the healthy duramater. Tumor extended to retrobulbar also resected sometimes joined with ophthalmologist depend on the intraconal extension. The neoplasm was then sent for histopathological examination. The dural tissue was closed using durorrhaphy or duroplasty. The surgical incision was closed layer by layer, sometimes requiring reconstruction.

Follow–Up Examinations

Clinical examination after surgery either immediate or during control in outpatient clinic was performed. Computed tomography (CT) was done a day after surgical procedure, and magnetic resonance imaging (MRI) was done a few months after surgery. Proptosis index was calculated from post–surgical imaging.

Statistical Analysis

All measured data were statistically analysed. Important descriptive data were presented in the form of tables.

Results

Demographic Result

There were 62 female (93.9%) and 4 male (5.1%) with a female–to–male ratio of 15.5 : 1. The mean age at tumor resection surgery was 44.68 ± 7.8 years old with 33 years as the youngest and 82 years as the oldest. The majority of patients were in the 41–50 age group (35 patients, 53.0%), followed by the 31–40 age group (21 patients, 31.8%),–51–60 age group (8 patients, 12.1%), and older than 60 years (2 patients, 3.0%).

Signs and Symptoms

Visual disturbance and proptosis were the most prevalent symptom of sphenoorbital meningioma. Other symptoms such as cranial nerve palsy, most notably involving the oculomotor nerve. These findings underscore the diverse array of symptoms linked to sphenoorbital meningiomas, highlighting their potential to significantly affect a patient's quality of life. A comprehensive account of the symptoms that manifested prior to surgery in patients with sphenoorbital meningioma is presented in Table 1.

Clinical Features	No. of Cases	Percentage
Visual acuity disturbance		
Yes	50	75.8%
No	16	24.2%
Proptosis		
Right Eye	27	40.9%
Left Eye	25	37.9%
No	14	21.2%
Diplopia		
Yes	1	1.5%
No	65	98.5%
Cranial nerve palsy		
N. III	21	31.8%
N. IV	18	27.3%
N. V	9	13.6%
N. VI	4	6.1%
N. VIII	1	1.5%

Table 1. Clinical symptoms of sphenoorbiital meningioma patients

Histological Diagnosis	No. of Cases	Percentage
WHO Grade I		
Meningothelial	49	74.2%
Mixed	15	22.7%
WHO Grade II		
Atypical	2	3.1%

Table 2. Histopathological Examination of Patients with Sphenoorbital Meningioma

Postoperative Outcome	No. of Cases	Percentage
Visual Outcome		
Improvement	4	6.1%
Stabilization	55	83.3%
Worsening	7	10.6%
Proptosis		
Improvement	52	100% (of patients with preoperative proptosis)

Table 3. Postoperative Outcome of Patients with Sphenoorbital Meningioma

Tumor Pathology

There are three grades of meningioma according to CNS classification. However, only grades I and II were found in our institution. The predominant histological subtype was meningothelial meningioma, constituting the diagnosis in 49 patients (74.2%), whereas mixed meningioma accounted for 15 (22.7%). Atypical meningioma is a rare observation, identified in only two cases (3.1 %). These findings imply that sphenoorbital meningiomas primarily comprise low–grade tumors, considering the significance of treatment approaches and prognosis. Insights regarding the histopathological evaluation of patients with sphenoorbital meningioma are provided in Table 2.

Surgical Outcome

The majority of patients (83.3%) reported stabilization of their visual symptoms, however there is a few patients experienced improvement. It is noteworthy that all patients who presented with preoperative proptosis showed improvement in their condition. These observations suggest that surgical intervention can effectively ameliorate proptosis, although its effect on the visual symptoms may be limited. A comprehensive summary of the outcomes after surgery for sphenoorbital meningioma is presented in Table 3.

Discussion

Sphenoorbital Meningioma

Meningiomas are tumors that form from the tissue of the brain membranes (meninges). In 1922, Cushing differentiated between two types of meningiomas: en masse meningioma and en plaque meningioma. Meningioma en masse generally forms a lobulated mass that appears fatter, whereas en plaque meningiomas have a more flattened and elongated shape. Sphenoorbital meningioma is a primary tumor of the en plaque type located in the facies cerebralis alae majoris and minoris of the sphenoid bone that can invade the surrounding bones and surrounding anatomical structures.[3] The most common symptoms of sphenoorbital meningioma are a triad of decreased vision, proptosis, and visual field disturbances.[2] Visual impairment and visual field disturbance are common because of interference with the n. opticus. This causes the complaints to occur unilaterally. Color blindness may also be present in a minority of cases. Proptosis caused by sphenoorbital meningioma is progressive, unilateral, and nonpulsatile. Proptosis in many cases causes aesthetic facial deformities.[4] Other cranial nerves can also be compressed, especially the n. III to VIII. Compression of the n. oculomotor, n. trochlearis and n. abducens responsible for eyeball movement can cause ophtalmoplegia and diplopia. Tumor suppression

of the trigeminal, facial, and vestibulocochlear nerves is less common than that of other cranial nerves. General neurologic disturbances may also present in the form of headache and epileptic seizures.[3]

Meningioma is one of the most common types of intracranial tumor, accounting for approximately 20% of all intracranial tumors in men and 38% of all intracranial tumors in women. The distribution occurs with a female–to–male ratio of 2:1.5, and the average age of patients with sphenoorbital meningioma is 51 ± 6 years.[4] These statistics are in line with our study, with an even higher ratio in women. As many as 4–18% of all meningioma cases are sphenoorbital meningiomas, making it the third most common meningioma after parasagittal meningioma and free confluent meningioma.[5–7]

Meningiomas are sporadic tumors. Molecularly, meningiomas are generated by excessive clonal development of single cells that is commonly associated with the deletion or inactivation of one or more focal chromosomes, leading to genetic instability. One of the most common genes known to play a role in some cases is NF2 on Chromosome 22. Genes that may play a role in the rarer prevalence include NF1, PTCH, CREBBP, VHL, PTEN, and CDKN2A. The complexity of the genetic aberrations that occur may increase as meningioma grade increases. Various risk factors can increase the likelihood of meningiomas. Some of these include hormonal factors, exposure to ionizing radiation or radiotherapy, obesity, alcohol consumption, comorbidities such as breast cancer, head trauma, and family predisposition to cancer, particularly history of meningioma. Hormonal factors that influence the incidence of meningioma are related to the estrogen, progesterone, and androgen receptors found in some meningiomas. This explains the higher number of cases in women than in men, as well as the significant correlation with breast cancer. This strong hormonal influence also makes the risk of meningioma increase at the peak of productive age in women, with the ratio of women to men reaching 3.15:1. The use of exogenous hormones such as oral contraceptives or hormone replacement therapy has also been found to increase the risk of meningioma. Exposure to ionizing radiation may increase the incidence of meningioma by 6–10 fold. Some evidence come from high–dose exposures such as atomic bomb survivors, but on the other hand, there is also evidence from low–dose exposures such as radiation therapy for children with ringworm infection of the scalp. A study showed that oral X–rays may increase the risk of meningioma (OR 2.06, 95% CI 1.03–4.17) although the association between dose and response was not significant (p=0.33). Diet and allergy patterns as risk factors for meningioma require further investigation. There have not been many studies that describe a clear correlation between head trauma and meningioma, but one study showed an increased incidence of meningioma after head trauma after 1 year (SIR 1.2, 95% CI 0.8–1.7). The risk of meningioma increases up to 2–fold in blood–related first–degree relatives of meningioma patients (SIR 2.2, 95% CI 1.4–3.1), with the risk decreasing with age.[8,9]

Many classification systems have been established for the treatment of sphenoorbital meningioma. These systems are based on morphology, epicentral region of the sphenoid, and specific tumor extension. Based on their shape, sphenoorbital meningiomas can be divided into intraosseous and intradural types, with the latter further divided into the en plaque and globoid types. Based on the epicentral region of the sphenoid, tumors can be divided into three groups according to their location at the epicenter of the facies cerebralis alae majoris: medial, middle, and lateral. Tumor extension can also differentiate the types of meningiomas which include extension to the temporal fossa, infratemporal fossa, orbita, superior orbital fissure, anterior clinoid processus, canalis opticus, and cavernous sinus.[10,11]

The diagnosis of sphenoorbital meningioma is based on a combination of clinical examinations, radiology, and pathology. Typically, radiologic examination is the initial diagnostic test, followed by clinical suspicion. Computed tomography (CT) and magnetic resonance imaging (MRI) are preferred at the head–level. CT scan revealed the bony features and degree of invasion of the meningioma in the harder structures, whereas MRI is more commonly used to evaluate soft tissues, including the intradural portion of the meningioma, and its effect on the brain parenchyma, such as mass effect or edema. Histological examination is commonly applied after tumor resection to establish the diagnosis of meningioma as generally the radiological picture is sufficient to explain the presence of sphenoorbital meningioma.[3] Some of the differential diagnoses of sphenoorbital meningioma include fibrous dysplasia, osteoma, osteoblatoma, osteoblastic metastasis, Paget's disease, internal frontalis hyperostosis, and erithroid hyperplasia.[12]

The management of sphenoorbital meningiomas is individual specific. The European Association of Neuro–Oncology (EANO) has established a clear management algorithm for sphenoorbital meningioma. Once a diagnosis is made, a clinical examination is the first consideration. Mild clinical symptoms allow for routine follow–up every 3–6 months clinically and radiologically, whereas severe symptoms require immediate assessment of age and general condition to determine definitive therapy. Routine follow–up results in mildly symptomatic patients, that is, worsening both clinically and radiographically, may

lead to the same pathway as in severely symptomatic patients. Younger age and better general condition may result in management with a more optimal prognosis. The definitive management options include tumor resection through direct microsurgery or stereotactic radiosurgery which is generally performed when the meningioma size is not too large, or radiotherapy which is generally performed on larger tumors.[13]

Tumor Resection Microsurgery

One of the definitive treatments that can be applied in cases of sphenoorbital meningioma is microsurgical resection of the tumor. Similar to surgery in general tumor cases, complete resection is the target of surgery to maximize the improvement of clinical symptoms and decrease the risk of disease recurrence. Decreasing the recurrence rate is particularly important, as sphenoorbital meningioma is a highly recurrent intracranial tumor, with a recurrence rate of 35–50%. However, complete resection is not always achievable due to various surgical complications.[14,15]

The degree of total resection of meningioma is commonly measured using Simpson's grading system. Grades I–III indicate macroscopically complete resection with removal of the associated dura mater–bone, only the associated dura mater, and no removal of the dura mater–bone, respectively. Grade IV describes subtotal tumor resection, while grade V signifies decompression without or with biopsy.[15] A study showed a general distribution of total resection (Simpson grade I–II) in 48%, subtotal resection (Simpson grade III) in 28%, and partial resection (Simpson grade IV) in 24%.[14]

Tumor resection in patients with sphenoorbital meningioma presents fairly good outcomes depending on the degree of resection, either it was gross total or subtotal resection or from the Simpson's grading system. In our cases, we found optimum improvement in structural symptoms such as proptosis, but debatable results in functional symptoms such as visual acuity. A previous study was conducted at our hospital to evaluate the health–related quality of life (HRQoL) of patients with sphenoorbital meningioma after surgery using the European Organization for Research and Treatment of Cancer Quality of Life Questionnaire (EORTC QLQ C–30). The results showed significant improvements in global health status/QoL, physical function, role function, emotional function, cognitive function, social function, and fatigue. The differences between the pre– and postsurgical EORTC scores were statistically significant ($p<0.05$). The findings of this study suggest that surgery for sphenoorbital meningioma can significantly improve HRQoL. An improvement in the global health status/QoL score indicated that patients experienced an overall improvement in their physical, emotional, and social well–being. Improvement in functional scales, such as physical role, emotional, cognitive, and social functioning, suggests that patients were able to perform their daily activities with less difficulty and had a better quality of life. Improvement in the fatigue score indicates that patients experienced less fatigue after surgery.[16] The patient's prognosis and quality of life are profoundly contingent upon their visual acuity, establishing it as a pivotal clinical parameter for individuals with sphenoorbital meningiomas. Surgical intervention, even in cases presenting with minimal visual impairment or hyperostosis, has the propensity to prevent the emergence of visual deficits. To enhance visual acuity, it is paramount to optimize both surgical intervention and postoperative surveillance. Because of the proclivity of sphenoorbital meningiomas to infiltrate the bony structures proximate to the cranial nerve foramina, early surgical intervention holds promise in averting extensive hyperostosis, constriction of the foramina, and the subsequent development of cranial nerve deficits. A study by Agosti et. al.[17] indicated that the timing of surgery plays a prognostic role in achieving favourable visual results. Significantly, involvement of the optic canal and intraorbital region has been identified as a predictive factor for postoperative visual deficits. The study showed promising results of clinical improvement after surgery with a suitable technique, reaching as high as 79.4% rate of better visual acuity and 71.5% rate of proptosis improvement. Nevertheless, it is crucial to acknowledge that the surgical procedure itself carries the inherent risk of incurring new visual and cranial nerve deficits. In scenarios involving geriatric patients, those afflicted with severe comorbidities, or those with extensive disease progression culminating in complete blindness, the potential benefits of surgery may not invariably outweigh the associated risks of complications. Nonetheless, it is generally postulated that the likelihood of experiencing novel complications diminishes when patients undergo early surgical intervention during the incipient stages of their disease progression, as cranial nerves are rendered less vulnerable when the degree of compression is relatively mild.[17,18]

The challenge to complete resection of meningiomas lies not only in the high degree of tumor invasion, but also in the hyperostotic bone it creates. Meningiomas and hyperostotic bones can extend into the orbit, cavernous sinus, and infratemporal region. Although the soft tissue component of the tumor is usually smaller, meningioma cells that often trigger recurrence can be located in the Haversian canal within the hyperostotic bone, making it necessary to remove it. In some cases, hyperostotic bone can cause visual disturbances, especially when

hyperostosis occurs in the anterior clinoid, foramen opticus, and canal opticus. Generally, patients with sphenoorbital meningiomas who undergo subtotal resection will be followed by postoperative radiotherapy.[14,19]

Conclusion

In summary, our study offers significant insights into sphenoorbital meningiomas. The most common symptom experienced by patients is visual disturbance, which affects approximately 75% of the individuals. It is essential to conduct preoperative radiological evaluations, including CT and MRI, to accurately determine the extent and location of the tumor.

Our results suggest that surgical resection of sphenoorbital meningiomas can be challenging, particularly in cases where the tumor is located in close proximity to critical structures. However, with careful surgical planning and execution, complete resection of the tumor can be achieved in most cases, resulting in favourable long–term outcomes. Our study underscores the importance of a multidisciplinary approach for the management of sphenoorbital meningiomas involving close collaboration between neurosurgeons, ophthalmologists, and radiologists.

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