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Research Article

Intraorbital Meningiomas: A Histopahologic Study

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Abstract: Optic Nerve Sheath Meningiomas account for about 4% of all intraorbital masses. They include both primary and secondary orbital meningiomas. Primary meningiomas arise from the cells lining the intraorbital or intracanalicular segments of the optic nerve. Secondary orbital meningiomas arise from the sphenoid wing or suprasellar region and infiltrate the orbit. Most of the orbital meningiomas are secondary. We did a retrospective study of orbital meningiomas encountered at our Institute over a period of seven years from 2008 to 2014. Eight cases of orbital meningiomas were reported during the study period, of which seven (87.5%) were secondary in origin. Six cases (75.6%) occurred in adult female patients. A single case was reported in a nine year old male patient. Clinically most of the patients presented with proptosis. Bone invasion was seen in two cases on CT scan. Histologically, meningothelial type was the commonest. The current WHO criteria (2007) has been applied to these tumors and graded accordingly. The WHO criteria for grading atypical meningiomas and the prognosis of intraorbitalmeningiomas are discussed.
Keywords: Meningioma, Intraorbital, Secondary, Histology, WHO grading, Prognosis

INTRODUCTION

Meningiomas account for approximately 4% of all intraorbital tumors and fall into two broad categories. Primary lesions derive from the cells lining the intraorbitaland intracanalicular segments of the optic nerve. Secondary tumors arise intracranially and subsequently extend to the orbit [1].Optic Nerve sheath meningiomas can cause symptoms of optic nerve compression when small. Clinically, Optic canal tumors present with unilateral visual loss and closely mimic the clinical presentation of optic neuritis [2]. Primary intraorbital meningiomas arising from structures other than optic nerve sheath have been termed as primary intraorbital ectopic meningiomas [3]. Orbital meningiomas are more common in young adult females [4]. When onset is in childhood, the meningiomas tend to be much more aggressive and tend to have a much worse prognosis than when onset is at an older age [5]. Primary optic nerve sheath meningiomas more often grow as spindle shaped masses about the optic nerve immediately anterior to the foramen, though intracanalicular optic nerve meningiomas have been reported rarely [6]. Meningiomas are challenging lesions to manage because of their posterior location and slowly progressive visual loss [7]. The diagnosis of optic nerve sheath meningioma relies heavily on imaging findings. Orbital meningiomas have histological features similar to intracranial meningiomas [8]. The criteria for grading meningiomas varied significantly through the years and the 2007 World Health Organization (WHO) classification includes fifteen named entities [9]. Virchow was the first to describe the classical pathological features of meningioma, namely psammoma body [10]. The existence of atypical and malignant meningiomas was clearly recognized by Cushing and Eisenhardt in 1938 [11]. Optic nerve sheath meningiomas infiltrate through the dura into the orbital tissue or through the pia into the parenchyma of the optic nerve or into the retina or choroid [11].

MATERIALS AND METHODS

A retrospective study was conducted at Sarojini Devi Eye Hospital, Hyderabaad over a period of seven years from 2008 to 2014. A total of eight specimens of intraorbitalmeningiomas were received at the Department of Pathology during this period. The available clinical data such as patient's age, sex imaging and surgical findings were recorded. The records showed that the patients underwent routine ophthalmological examination, B scan Ultrasonography and CT scan of the Orbits.Complete laboratory work up was done in each case. The pathological data showed that in five casesincisional biopsy was performed and in three cases subtotal exenteration was done. The histopathology slides of all the cases were retrieved and

reviewed. Grading was done applying the WHO 2007 Classification scheme.

RESULTS

Our study group consisted of eight histologically proven cases of intra orbital meningiomas. There were five females and three males. The age of the oldest patient was 60 years and youngest patient was9 years. Most of the cases occurred in the third and fourth decades. Incisional biopsy was done in five cases. Grossly, the size ranged from 1.5 to 2.5cm. Subtotal exenteration was done in three cases, as one of the cases was clinically diagnosed as fungal mucormycosis and the other as a large lacrimal gland tumor. The third case was a recurrent meningioma which showed intracranial extension. Histologically, four cases showed features of meningothelialmeningioma. Among others, one case each was representated by transitional type, fibroblastic type andangiomatous type. There was a single case of atypical meningioma (Table. 1)

Table 1: Histological studies

Histological type	No.(%)	WHO Grade
Meningothelial Meningioma	4 (50%)	I
Transitional Meningioma	1 (12.5%)	I
Fibroblastic Meningioma	1 (12.5%)	I
Angiomatous Meningioma	1 (12.5%)	I
Atypical Meningioma	1 (12.5%)	II

Microscopically, Meningothelial meningiomas showed lobules of meningothelial cells arranged in syncitial pattern without definite cell borders (Fig.1). The nuclei were vesicular with intranuclear inclusions in some of them. Mitotic activity was low. Fibroblastic meningioma showed bundles of fibroblast like cells with areas showing collagen. Whorl formation was seen focally. Transitional meningioma showed a combination of features of meningothelial and fibroblastic meningioma (Fig. 2). Angiomatous meningioma showed many blood vessels of different sizes and shapes with endothelial lining (Fig. 3). Atypical meningioma showed increased cellularity, small cell change, prominent nucleoli, sheet like growth pattern, areas of necrosis and extension into extraocular muscles (Fig.4). Atypical meningioma was graded as WHO Grade II. Rest of the cases were graded as WHO Grade I tumors. Hyperostosis due to bone invasion was seen in two cases.

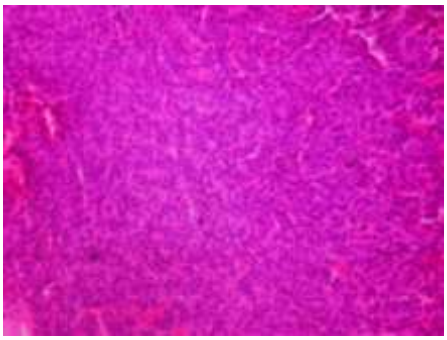

Fig.1: Meningothelial meningioma showing lobules of meningothelial cells in syncytial pattern

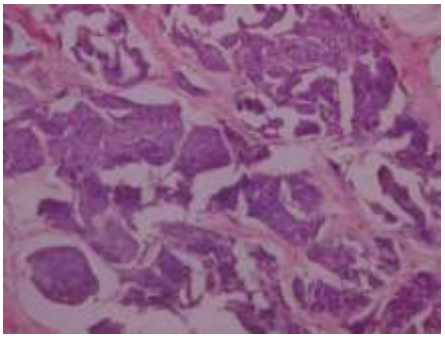

Fig. 2: Transitional meningioma showing a combination of meningothelial and fibroblastic pattern

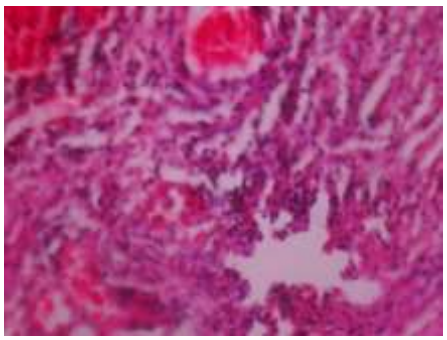

Fig. 3: Angiomatous meningioma showing blood vessels lined by endothelial cells

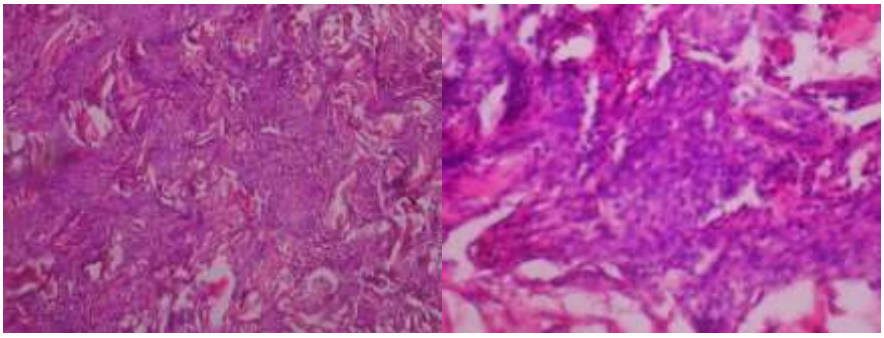

Fig. 4: Atypical meningioma showing sheeting pattern and small cells with prominent nucleoli

DISCUSSION

Optic Nerve sheath meningioma is a term applied to primary or secondary meningiomas of the optic nerve [8]. Primary orbital meningiomas arise from the optic nerve sheath from the cells lining the intraorbital or intracanalicular segments of the optic nerve sheath and may extend through the optic nerve intracranially [1] or may infiltrate the sphenoid wing and produce hyperostosis [12]. Secondary orbital meningiomas arise from the inner and outer aspects of the sphenoid wing or suprasellar region and infiltrate the orbit [12]. Secondary orbital meningiomas are more common than primary [8, 12]. In our study also seven (90%) out of eight were secondary meningiomas.

Since many reports and series fail to distinguish between the primary and secondary tumors, determining the incidence of either with accuracy is difficult [8]. Orbital meningiomas tend to affect females more commonly. [1]. Our study also showed a female preponderance, six out of eight cases occurred in females (75.6%), correlating with other studies. However literature shows that there is wide variation in the male to female ratio [8]. All the patients in our study were adults except one case which occurred in a nine year old male child. Meningiomas are relatively uncommon in children [12]. Gliomas are more common in children and evoke a surrounding dural reaction that mimics meningioma. Clinically most of our cases presented with proptosis. Proptosis is commonly observed in secondary intraorbitalmeningiomas. Primary optic nerve sheath meningiomas present with slowly progressive optic atrophy characterized by slowly progressive painless visual loss [7]. CT scan was performed in all our cases prior to biopsy. Orbital CT and MRI are excellent modalities in the visualization of meningiomas [7]. Meningiomas of the orbit have pathological characteristics similar to intracranial meningiomas. Fifteen subtypes are identified by the revised WHO classification [12].

Table 2: WHO Classification of Meningiomas [9]

Type of Meningioma	WHO Grade
Benign	
Meningothelial	I
Fibrous (Fibroblastic)	I
Transitional	I
Psammomatous	I
Angiomatous	I
Microcystic	I
Secretory	I
Lymphoplasmacytic-rich	I
Metaplastic	I
Aggressive	
Atypical	II
Clear cell	II
Chordoid	II
Rhabdoid	III
Papillary	III
Anaplastic	III

The WHO grading system was developed through clinicopathologic correlations using intracranial meningiomas [1]. Applying the WHO criteria to our series, it was observed that 90% were WHO grade I, correlating with other studies [1]. Consistent with the previous reports, psammomatous variant, frequently seenintracranially, was not encountered in our series. Three of the following features places the tumor in atypical category i.e., increased mitotic activity, increased cellularity, small cells, prominent nucleoli, sheet like growth pattern and areas of necrosis. We encountered only one case of atypical meningioma, which comes under WHO Grade II. We did not encounter any Grade III meningiomas. Bone invasion was seen in two of our cases. Secondary orbital meningiomas usually show bone involvement demonstrating as hyperostosis on CT scan. Meningiomas are slow growing progressive tumors. All our cases were referred to Oncology Institute after diagnosis for further management. With the advent and characterization of new treatment modalities, numerous management options are available for the treatment of orbital meningiomas. Earlier, resection was the primary treatment modality for secondary orbital meningiomas. Due to the technical difficulties and possibility towards post operative blindness, radiation therapy has replaced surgery [8]. Recurrences are thought to be due to residual tumor in the operative bed [12]. In our study recurrence was seen in two cases. Observation is a reasonable option in primary optic nerve sheath meningiomas, however radiation slows the progression and surgery may be done to relieve eye pain.

CONCLUSION

Meningiomas are rare intraorbital tumors. Most of them are secondary arising from the sphenoid wing or suprasellar region and infiltrate the orbit. Orbital meningiomas are commonly seen in adult female patients with rare occurance in children and usually present clinically with proptosis. CT scan is a useful modality in the diagnosis of intraorbital meningioma. Biopsy is done for confirmation. Histologically, meningothelial pattern is the commonest. However psammoma bodies are seen less frequently compared to intracranial meningiomas. Meningiomas are slow growing tumors.

The tumors are graded based on the current World Health Organization criteria (2007) [9]. WHO Grade I tumors are commonly encountered. Atypical meningioma is graded as WHO Grade II tumor. There are numerous management options in the treatment of orbital meningiomas. Given its technical difficulties and trend towards post operative blindness, surgery has largely been replaced by radiation therapy, which is now accepted as the appropriate vision preserving therapy for the management of secondary intraorbital meningiomas. Observation may be a reasonable option in primary optic nerve sheath meningiomas.

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