

CASE REPORT

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Ancient schwannoma in phthisis bulbi: Case Report

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Abstract

Schwannoma is benign nerve sheath tumor of Schwann cells. A 41-year-old female patient presented with pain and a complaint of inability to see. A mass revealed in left eye. Mass was histopathologically evaluated as schwannoma. This lesion in a rare location is presented with the literature.

Keywords: Intraocular, intraorbital, schwannoma, phthisis bulbi, ancient.

Introduction

Schwannomas are benign, well- circumscribed nerve sheath tumor derived from Schwann cells, formerly known as neurilemmoma or neurinoma. The tumor usually presents with a slowly progressing mass effect. When the tumor is small, it is usually asymptomatic. Tumor includes Antoni A (hypercellular) areas, Antoni B (hypocellular) areas and Verocay bodies (palisaded nuclear aggregates) at microscopic examination. Tumor cells are positive with S100 antibody [1-3].

Case Report

A 41-year-old female presented to the ophthalmology clinic with the complaint of pain and loss of vision at the left eye. Ultrasound results were compatible with retinal detachment. Left eye was enucleated. Macroscopic examination showed a 1.5 cm diameter mass extending into the vitreous cavity. Histopathologic examination revealed a spindle cell tumor which includes focally ossification areas, palisaded cellular areas and myxoid hypocellular areas (Figure1, 2). Immunohistochemically, the tumoral cells showed positive immunostaining with S-100 (Figure 3). Our case was reported as ancient schwannoma which includes ossification areas.

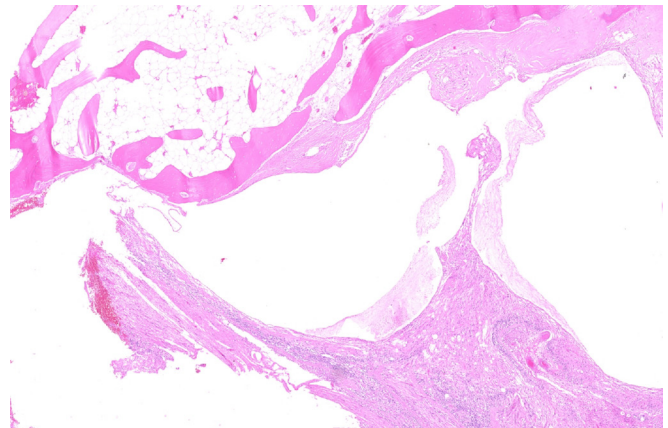


Figure 1. Cystic degeneration and ossification areas in the tumor. H&E x10

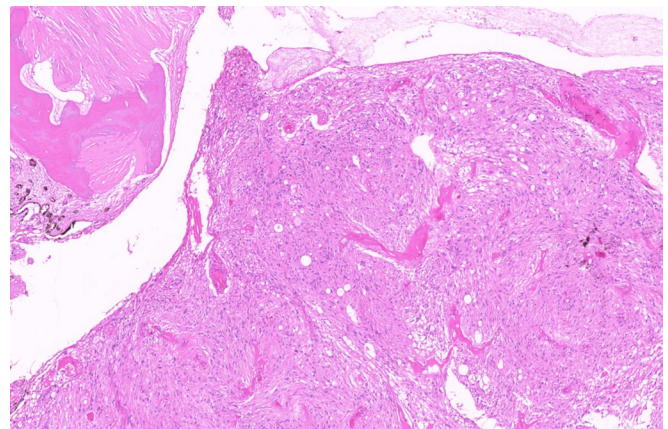


Figure 2. The focus of ossification in the vicinity of ciliary body (top left of the picture) and the classic schwannoma areas (in the bottom of the picture). H&E x25

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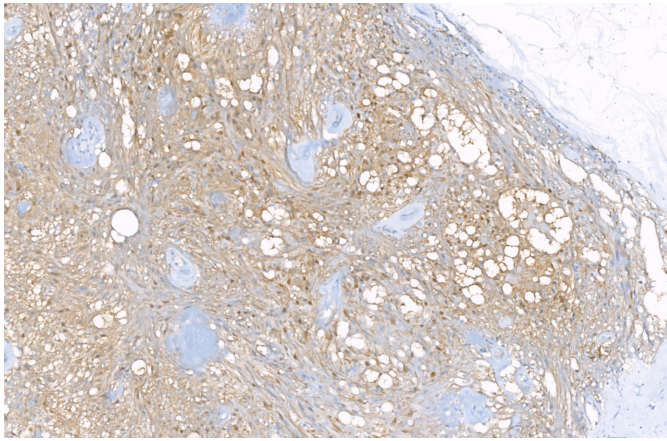


Figure 3. Tumor cells showed positive staining with S100 antibody. S100 x25

Discussion

Orbital schwannomas are very rare (%1 of all orbital neoplasms) lesions. Although intraorbital schwannomas are benign lesions, they can make blindness because of mass effect. And because of this effect surgery must be done as earliest as possible [3]. Schwannoma can be seen in every age, but more common in 20-60 year-old range. Our case was in the age range mentioned in the literature. Most cases are sporadic, the rest is associated with various syndromes (neurofibromatosis, schwannomatosis, carney complex etc). Our case was sporadic. Schwannomas are often seen in the head and neck region, and extremities. Deep located cases may be located in the retroperitoneum and posterior mediastinum [4]. Cystic, haemorrhagic, myxoid degeneration and calcifications may exist in schwannomas. In our case, tumor was defined as ancient schwannoma because of including cystic degeneration and ossification areas [4,5]. It is very rare in the eye as in our case. Most of schwannomas have “conventional” schwannoma features. In the literature, orbital schwannomas were found to be more oval or more spindle-shaped than other regions schwannomas, but in our case histopathological classical morphology (some of cells closely packed spindle cells with fusiform nuclei, some of cells haphazardly distributed cells with distinct cytoplasmic margins) was present. Although tumor is easy to diagnose histopathologically, positive immunohistochemical staining of tumor cells with S100 antibody should be shown. Complete surgical excision is the gold standard treatment. Local recurrences may be seen in incomplete surgery. Schwannomas are resistant to radiotherapy [5]. Rarely malign transformation can occur [4-6]. There was no evidence of malignancy (mitosis, necrosis, nuclear atypia or pleomorphism) in our case.

Phthisis bulbi is defined as atrophy with shrinkage of the globe and

disorganization of the intraocular structures. Most common cause of phthisis bulbi is trauma. Other causes are injuries (close globe, open globe, chemical, radiation), surgeries, inflammation (uveitis, etc.), infections (keratitis, endophthalmitis, panophthalmitis), drugs (cidofovir) and tumors. Among these etiologies in the etiology are most common tumors are retinoblastoma and choroidal melanoma. Other tumors very rarely cause phthisis bulbi [7,8]

Conclusion

Our case is evaluated to be presentable because it is extremely rare an ancient schwannoma that causes phthisis bulbi.

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Conflict of interest

The authors declare that they have no conflict of interest

Ethics committee approval

This work has been approved by the Institutional Review Board

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