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Case Report Open Access

Intraocular Schwannoma in an Eviscerated Eyeball

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Abstract

Schwannomas rarely present as intraocular tumors. The diagnosis is usually established histopathologically after enucleation or surgical excision for a suspected uveal melanoma. We present an interesting case of an intraocular schwannoma that presented as a cystic mass in an eviscerated eyeball.

Keywords: Intraocular schwannoma; Cystic schwannoma

Introduction

Schwannomas are benign, slow growing peripheral nerve sheath tumors. They comprise 1% of all orbital tumors [1]. Intraocular schwannomas are known to arise from the ciliary body and choroid and are extremely rare with only isolated case reports in the English literature [2-7]. In the combined studies of Ferry, and Shields and Zimmerman, they represented only 2 of 141 cases of pseudomelanomas [8,9]. We report a unique case of an intraocular cystic schwannoma presenting in an eviscerated eye.

Case Report

A 65 year old male, presented with a large painless mass filling the entire right orbit of 5 years duration (Figure 1a). The patient had a prior history of minor blunt ocular trauma 6 years back following which he developed a painful blind eye. Subsequently, 6 months later he underwent an evisceration without implant at a local ophthalmic centre. No records of any preoperative investigations and histopathological analysis of the ocular contents were available.

The right eye had no perception of light. The cornea was not present. A yellowish-white structure with overlying hypertrophied conjunctiva was visible. We presumed this to be distended sclera surrounding an intraocular mass. The underlying mass was non tender, soft to cystic on palpation with no nodularity. No pulsations were elicited and no change in size was observed upon the valsalva maneuver. The surrounding adnexal structures appeared to be normal. The left eye was within normal limits.

Ultrasonography revealed a large cystic structure filling the orbit with mild to moderate internal reflectivity. Computed tomography revealed a well defined, homogenous non-enhancing cystic mass contained within a distended sclera, filling a widened orbital cavity (Figure 1b). The optic nerve was seen at the orbital apex with associated widening of the optic canal. In a few sections, stretched extraocular muscles were identified. No extrascleral or intracranial extension of the mass was noted. No other orbital structures could be identified separately.

Fine needle aspiration cytology from the mass revealed altered blood. The patient was taken up for mass excision under general anesthesia. Around 27 ml of altered blood was aspirated from the mass.

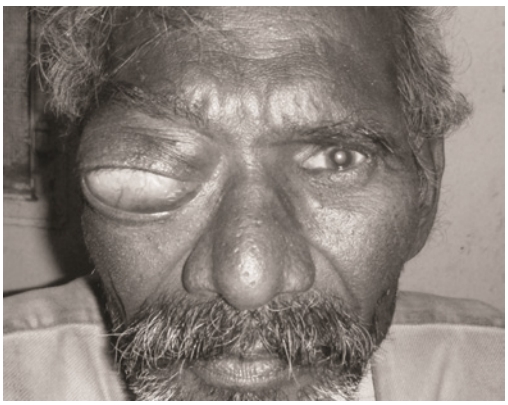

Figure 1a: A large cystic mass filling the right orbit.

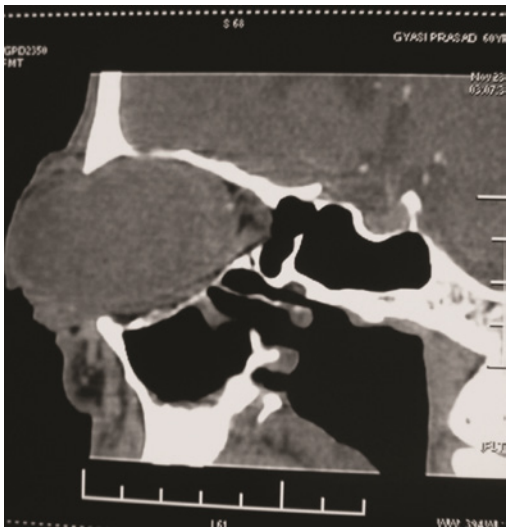

Figure 1b: Computed tomography showing a homogenous cystic mass contained within a distended sclera, filling up the entire orbit.

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Intraoperatively, the conjunctiva was separated from the underlying sclera. Stretched out and thickened extraocular muscles were identified, hooked and cut. Optic nerve was cut at the apex and the distended eyeball was removed. Hemostasis was achieved and the orbital cavity was packed with gauze. A second stage dermis fat graft was planned for rehabilitation.

Cut section revealed a yellowish necrotic mass with multiple blood clots filling the outer coat of the eye ball (Figure 2a). No other ocular structures could be identified. On histopathology, the presence of sclera and extraocular muscles was confirmed. On light microscopy a tumor composed of spindle shaped cells with large areas of hemorrhage and cystic degeneration were seen inside the eyeball (Figure 2b). On higher magnification, the spindle shaped cells had indefinite cytoplasmic outlines and wavy nuclei. Antoni A areas with pallisading nuclei were also seen (Figure 2c). Immunohistochemistry revealed positivity for S-100 and vimentin and was negative for CD-34, Smooth muscle actin and HMB-45. This was compatible with a diagnosis of schwannoma.

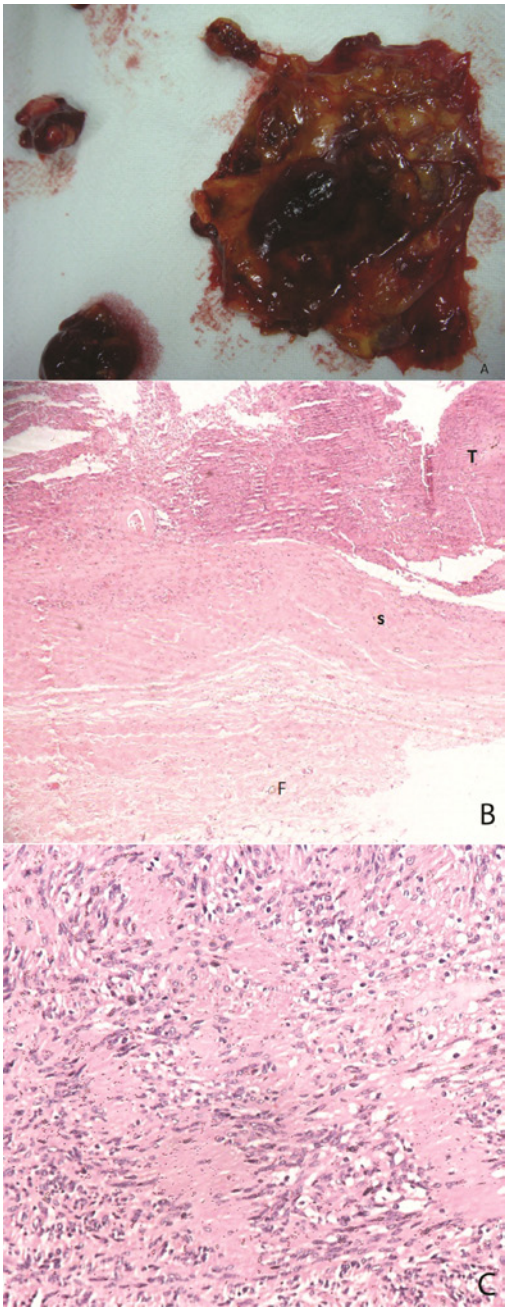

Figure 2a: Yellowish necrotic mass with multiple blood clots filling the sclera cavity. **b:** Microphotograph (4X) showing a spindle cell tumor (T) inside the sclera (S). Orbital fibrofatty tissue is seen outside (F) **c:** On higher magnification (20X) characteristic spindle shaped cells forming Verrucay bodies were seen.

Discussion

Schwannomas arising from the uveal tissue are rarely encountered as intraocular tumors [2]. They appear as solitary, amelanotic solid lesions usually of the ciliary body or choroid. Ocular findings, ultrasonography and imaging is usually not helpful in differentiating a schwannoma from other benign uveal tumors or an amelanotic melanoma [4]. Diagnosis is usually established histopathologically after enucleation for a suspected melanoma [2-4]. There are however, few case reports where in view of atypical findings of malignant melanoma or suspected benign etiology, successful local resection of uveal schwannomas has been done [5,6]. Though, the diagnosis essentially remains histopathological.

Schwannomas can be solid, multicystic or even purely cystic. Long standing schwannomas may develop degenerative changes, necrosis or hemorrhage within the tumor. In a series reported from our centre, cystic schwannomas constitute 41% of all orbital schwannomas. Purely cystic schwannomas are even rarer [10].

Our case here presents a unique situation of a rare intraocular tumor in an eviscerated eye. Moreover, a completely cystic schwannoma at this location has never been reported before. The intraocular location of the tumor was confirmed by the presence of sclera with surrounding extraocular muscles and the optic nerve noted intraoperatively and also by the histopathological sections. The microscopic and immunohistochemical findings were typical for a schwannoma. Presence of altered blood in the tumor and predominance of Antoni B areas on histopathology explains the cystic nature of the tumor.

We believe that our patient had an intraocular schwannoma with secondary glaucoma when he was eviscerated earlier for a presumed post traumatic painful blind eye. Five years later he presented with a recurrence inside the empty scleral cavity, perhaps from a remnant uveal /tumor tissue. The recurrent tumor was slow growing in view of the benign etiology, leading to distention of the surrounding sclera and orbital expansion. This led to compression and atrophy of the surrounding orbital structures. Moreover, cystic degeneration developed within the tumor. No extraocular extension was found. A ciliary body schwannoma recurring 15 years after an iridocyclectomy has been reported earlier [7].

Hence, intraocular schwannomas are still a diagnostic dilemma and a radical surgery is usually required in view of their rare incidence and inability to differentiate them from malignant melanomas. However, the possibility of a schwannoma should be considered in slow growing intraocular tumors with features atypical for a melanoma. In our case whether the schwannoma was present prior to evisceration or developed from a uveal remnant after the surgery is controversial. An enucleation was done aiming at complete removal of the intraocular mass.

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