Rare disease

Intrasclera schwannoma

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Summary

Schwannomas located in the eye are extremely rare and mainly arise from ciliary nerves. Ocular schwannoma usually present in the choroid, ciliary body or rarely sclera as a benign solitary amelanotic lesions. A 41-year-old woman presented with a history of decreased vision in her right eye of 3 months duration. She had an amelanotic lesion in the nasal side of left fundus. After all work-ups, malignant melanoma was suspected and enucleation was done. In clinicopathologic examination, findings were in favour of intrascleral schwannoma. Despite new modern diagnostic modalities, there are still some fundus lesions that are easily mistaken for amelanotic melanoma. Diagnostic procedures such as fine needle aspiration should be considered for equivocal lesions in the fundus that are not definitely diagnosed before surgery.

BACKGROUND

Schwannomas arising from ciliary nerves are extremely rare and only very few cases have been reported in the literature. Schwannomas located in other parts of the body (specially vestibular system) are usually associated with neurofibromatosis.¹ Intraocular tumours are usually present in the choroid, ciliary body or rarely sclera as a benign solitary, amelanotic lesion.²⁻⁴ In this article, we will present a patient with an intraocular tumour which was proved to be intrasclera schwannoma.

CASE PRESENTATION

A 41-year-old woman presented to the ophthalmology clinic with chief complaint of decreased visual acuity of right eye of 3 months duration. She was in a good health condition and family history was unremarkable. Her vision was 20/20 in the right eye and 20/200 in the left eye. The intraocular pressure of both eyes were 14 mm Hg. Slit lamp examination of anterior chamber of both eyes were normal without any abnormality of cornea, anterior chamber, lens or iris. Fundus evaluation of right eye was normal

Figure 1 Fundus picture of the nasal side of the left eye showing the bulging mass under the retina with irregular surface and tortuous vessels.

Figure 2 Fluorescein angiography shows abnormal leaky tumoural vessels. 203×229 mm (96 × 96 DPI)

Figure 3 T1-weighted image enhanced with gadolinium revealed ring enhancement around the tumour (A) while T2 weighted image shows in homogeneity inside the tumour (B). 253×97 mm (180 × 180 DPI)

but in the left eye there was a 10 disc diameter mushroom shaped mass in the nasal side of the posterior pole which had irregular surface and tortuous vessels on the top. The tumour was extended from nasal retinal periphery up to 2 mm to the optic disc (figure 1). The abnormal vessels

were leaky in fluorescein angiography (figure 2). Ocular ultrasound showed a fusiform mass with high initial echogenicity, irregular internal echo and high echogenicity of the posterior part of the tumour. No acoustic hollowness was noted. The patient's orbital MRI showed a mass with

Figure 4 Gross section of the enucleated globe showing the intrascleral tumour without significant choroidal thickening and with overlying retinal detachment. 282×239 mm (180 × 180 DPI)

ring enhancement around the lesion at the nasal side of globe wall (figure 3).

Vision gradually decreased to finger counting due to retinal detachment. According to the clinical and laboratory data findings, amelanotic melanoma was proposed and enucleation was done for her while systemic work-ups were insignificant. The enucleated globe grossly showed a whitish oval shaped bulging visible on the sclera adjacent to the optic nerve corresponding to the expected tumoural process as well as overlying retinal detachment (figure 4).

INVESTIGATIONS

Histopathology findings

Globe was fixed in 10% buffered formaldehyde for 24 h and then pupilo-optic cut section was made passing through the tumour area. The tissue was processed and 5 micron cut sections prepared from paraffin blocks were stained with H&E. In the light microscopic examination, there was a fusiform tumour in the scleral tissue consisted of a prominent hypo cellular myxoid area (Antoni B) surrounded by areas of higher cellularity (Antoni A) consisted of spindle shaped cells forming preliminary neural structures (figure 5). Immunohistochemical study of the specimen showed positivity for S100 and vimentin, suggestive of neural origin of the tumour.

Systemic work-up

Systemic evaluation for distant spread of tumour was performed. Complete blood cell count, liver ultrasonography, liver function test and chest x-ray were all normal.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis includes choroidal melanoma, hemangioma of choroid, schwannoma or a metastatic lesion.

TREATMENT

According to the clinical and laboratory data findings, amelanotic melanoma was proposed and enucleation was done.

OUTCOME AND FOLLOW-UP

One month after enucleation, ocular prosthesis was fitted and she is healthy after 16 months follow-up.

DISCUSSION

Intrasclera schwannoma, a very rare benign tumour arises from peripheral nerve sheath of ciliary nerves piercing the sclera and may show itself outside the sclera or present as an intraocular tumour. In the latter case, differentiation with amelanotic melanoma or other choroidal tumours is difficult.⁵ Misdiagnosing schwannoma for amelanotic melanoma is reported by Matsuo *et al.*⁶ They have reported a 72-year-old woman who's left eye was enucleated with diagnosis of melanoma for which all the histopathologic studies were in favour of schwannoma. Lee *et al* have also reported a case of intrachoroidal schwannoma which was enucleated with diagnosis of amelanotic melanoma.⁵ Intrasclera schwannoma reports are actually very rare in the literature. Three cases were reported one of which was an 11-year-old girl with an intrascleral mass located in the

Figure 5 Histopathology shows the mass embedded in sclera with areas of hypo and hypercellularity. Inset: Higher magnification shows the benign looking cells without atypia. (H&E $\times 100$ & 400) 129×100 mm (300×300 DPI).

anterior part of globe in supra temporal area. Resection of tumour was done, saving the globe and, in histopathology it was diagnosed as schwannoma.⁷ Schwannoma can be diagnosed histopathologically with the existence of palisaded spindle cells (Antony A), in the mixoid areas of degeneration (Antoni B).

Immunohistochemical confirmation can be done by showing positivity for protein S-100 and vimentin.⁷ Our case was confirmed by histopathology as well as immunohistochemistry. Another interesting point is that the MRI showed ring enhancement which is less consistent with the diagnosis of melanoma or metastasis. Because of low index of suspicion, this type of tumour usually is misdiagnosed⁵⁻⁷ as in our case. While T1 and T2 weighted MRI images are routinely used to diagnose intraocular tumours, newer methods like magnetic resonance spectroscopy could be considered in atypical cases to provide in vivo metabolic information of the tumours; this procedure has been shown to be effective in differentiation between benign and malignant orbital lesions in 93.7% of cases.⁸ Although procedures like positron emission tomography fused with CT are capable to detect the physiologic activity of medium to large size malignant tumours, they have limited value in diagnosis of early stages.⁹ Exact confirmation of melanoma can be performed with fine needle aspiration biopsy (FNAB).¹⁰ In their study on 25 eyes with macular choroidal melanoma, Young *et al*, reported that FNAB is able to provide the exact diagnosis in 74% of cases.¹⁰ However, FNAB was not done for our patient because of some caveat about secondary iatrogenic spread of tumour and technical problems due to nasal location of the tumour and small orbit of the patient. But we strongly recommend FNAB when it is feasible in

suspicious cases of choroidal melanoma because of its accuracy for definite diagnosis. Plaque brachytherapy is a popular modality for treatment of choroidal melanoma; however, Papageorgiou *et al* have reported some factors such as young age, tumours larger than 5 mm and proximity to the fovea as some risk factors for prediction of poor response to brachytherapy.¹¹ Plaque brachytherapy was not a good choice for our patient because she was young and had a large tumour (measuring about 10 disc diameter). Partial resection of choroidal melanoma, which was invented and promoted by Peyman,¹² was not possible in this case due to posterior location and lack of experience.

Learning points

- Considering rare tumours in the case of atypical intraocular tumour and confirming the diagnosis may spare the eye from enucleation.
- Procedures such as fine needle aspiration or vitrectomy assisted biopsy (if fine needle aspiration did not yield enough cells) may help to make the definite diagnosis.¹³

Competing interests None.

Patient consent Obtained.

REFERENCES

1. Mautner VF, Schröder S, Pulst SM, *et al*. [Neurofibromatosis versus schwannomatosis]. *Fortschr Neurol Psychiatr* 1998;**66**:271–7.
2. Shields JA, Sanborn GE, Kurz GH, *et al*. Benign peripheral nerve tumor of the choroid: a clinicopathologic correlation and review of the literature. *Ophthalmology* 1981;**88**:1322–9.

3. **Smith PA**, Damato BE, Ko MK, *et al.* Anterior uveal neurilemmoma—a rare neoplasm simulating malignant melanoma. *Br J Ophthalmol* 1987;**71**:34–40.
4. **Cho YJ**, Won JB, Byeon SH, *et al.* A choroidal schwannoma confirmed by surgical excision. *Korean J Ophthalmol* 2009;**23**:49–52.
5. **Lee SH**, Hong JS, Choi JH, *et al.* Choroidal schwannoma. *Acta Ophthalmol Scand* 2005;**83**:754–6.
6. **Matsuo T**, Notohara K. Choroidal schwannoma: immunohistochemical and electron-microscopic study. *Ophthalmologica* 2000;**214**:156–60.
7. **Graham CM**, McCartney AC, Buckley RJ. Intrasccleral neurilemmoma. *Br J Ophthalmol* 1989;**73**:378–81.
8. **Roshdy N**, Shahin M, Kishk H, *et al.* MRI in diagnosis of orbital masses. *Curr Eye Res* 2010;**35**:986–91.
9. **Reddy S**, Kurli M, Tena LB, *et al.* PET/CT imaging: detection of choroidal melanoma. *Br J Ophthalmol* 2005;**89**:1265–9.
10. **Young TA**, Buegess BL, Rao NP. Transscleral fine-needle aspiration biopsy of macular choroidal melanoma. *Am J Ophthalmol* 2008;**145**:297–302.
11. **Papageorgiou KI**, Cohen VM, Bunce C, *et al.* Predicting local control of choroidal melanomas following 106Ru plaque brachytherapy. *Br J Ophthalmol* 2011;**95**:166–70.
12. **Peyman GA**, Gremillion CM. Eye wall resection in the management of uveal neoplasms. *Jpn J Ophthalmol* 1989;**33**:458–71.
13. **Eide N**, Walaas L. Fine-needle aspiration biopsy and other biopsies in suspected intraocular malignant disease: a review. *Acta Ophthalmol* 2009;**87**:588–601.

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Hosseini H, Eghtedari M, Roozitalab M, Ashraf M, Shahrzad S. Intrascclera schwannoma. *BMJ Case Reports* 2011;10.1136/bcr.07.2010.3158, date of publication

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