Iris and iridociliary melanoma : concepts in diagnosis and management

Razzaq, L.

Chapter 4

Mesectodermal suprauveal iridociliary leiomyoma: Transscleral excision without postoperative iris defect

Lubna Razzaq[1], Ekaterina A Semenova[2], Marina Marinkovic[1], Rob JW de Keizer[1], Sjoerd G Van Duinen[3], Gregorius PM Luyten[1]

[1] Department of Ophthalmology, Leiden University Medical Center (LUMC), Leiden, the Netherlands.
[2] Moscow Helmholtz Research Institute of Eye Diseases, Moscow, Russia.
[3] Department of Pathology, Leiden University Medical Center (LUMC), Leiden, the Netherlands.

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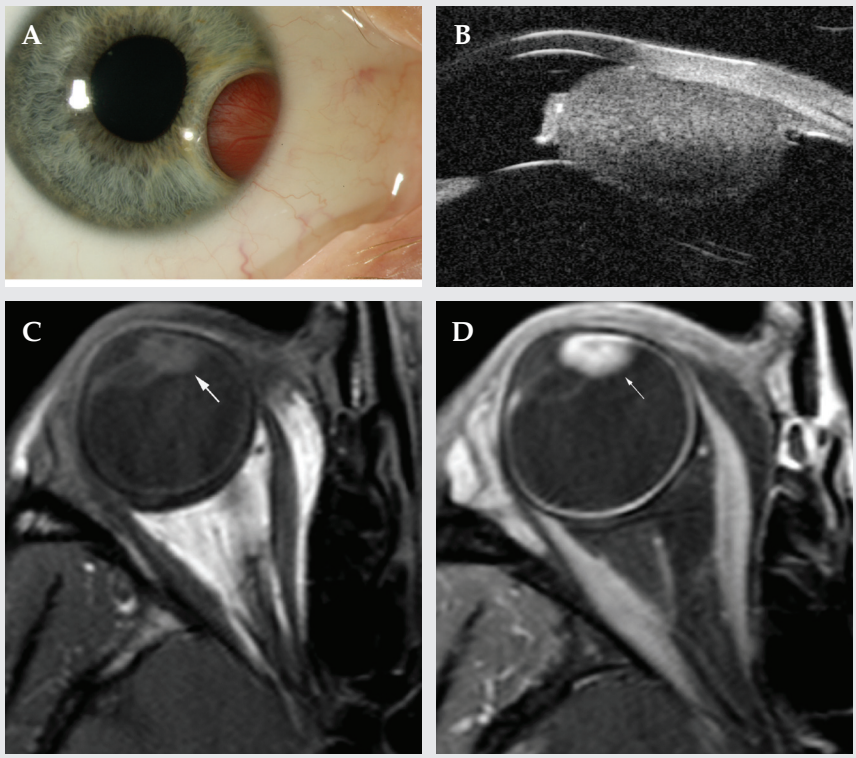
Mesectodermal leiomyoma of the ciliary body is a rare tumor originating from smooth muscle, having both muscular and neural differentiation [1]. The first case was reported in 1977 and so far 24 cases have been reported [1]. It should be considered in the differential diagnosis of an amelanotic melanoma, especially in young people. We here report a case of mesectodermal iridociliary leiomyoma in a young boy, showing the results of ultrasound biomicroscopy (UBM), 3-tesla MRI, histology and immunohistochemistry. The tumor was excised en-bloc by a transscleral approach and later treated by Ruthenium (Ru-106) plaque brachytherapy.

Report of the case

A 20-year old man was referred to our department because a slow-growing red mass, which had been visible in the right iris since three months, without any csymptoms. Visual acuity was 20/20 in both eyes. Slit-lamp examination revealed a vascularized mass extending from 3 - 5 o'clock position on the iris, pushing the iris root centrally, the lesion measured 4.5 x 2 mm (Figure 1A). Gonioscopy revealed the mass extending into the open anterior chamber angle. Transillumination showed enhanced transmission of light within the mass extending into the ciliary body. The intraocular pressure and fundus were normal. The left eye was unremarkable. UBM revealed a low reflective iridociliary mass measuring 9.7 x 6.8 mm with a prominence of 5.6 mm including the sclera (Figure1B). 3-Tesla MRI showed a well-defined mass located in the ciliary body and iris, which was hyperintense on T1 weighted image with marked enhancement after gadolinium (Figure 1C&D). Although the clinical diagnosis was a leiomyoma, in accordance with the parent's wish, a biopsy was performed, which established the smooth muscle nature of the tumor. The therapeutic options were discussed with the patient and his family and excision was chosen. Transscleral en-bloc excision of the tumor was performed by one of the authors (GPML), by peeling off the suprauveal tumor from the ciliary body under a large deep scleral flap, thus sparing the iris, ciliary body stroma and choroidal tissue [2]. Histopathology revealed bundles of elongated spindle cells with oval nuclei and eosinophilic cytoplasm composed of nonbranching fibrils. Immunohistochemistry showed positive results for α-SMA, Desmin, Caldesmon, CD-56 and neuron specific enolase (NSE) and results were negative for S-100 and HMB-45 (Figure 2A, B&C). Therefore, the diagnosis of an iridociliary mesectodermal leiomyoma was confirmed. The post-operative result was excellent with a normal appearance of the iris (Figure 2D). In order to avoid any recurrences as there were small remnants of the tumor left in situ, a Ru-106 CCB plaque, delivering 108 Gy at 2.4mm, was applied over the excised area.

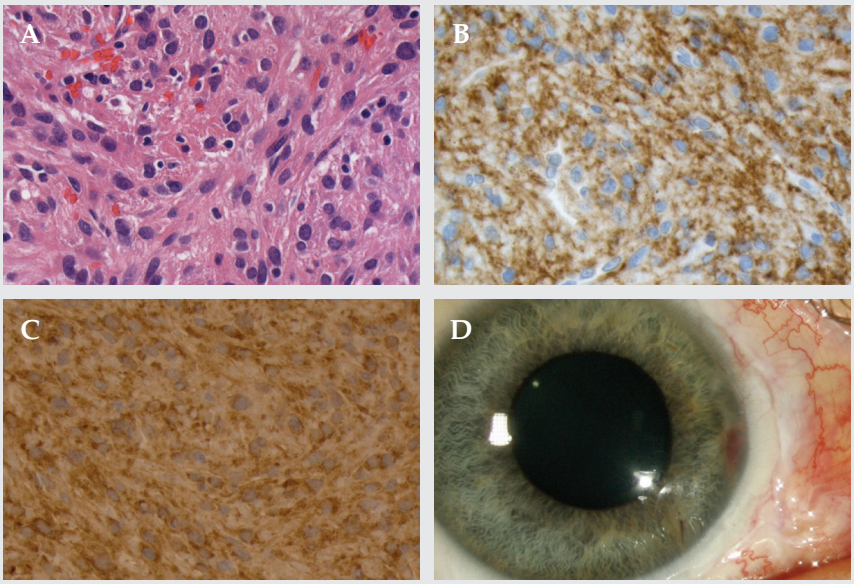
Figure 1 **A**: Slit-lamp photograph showing a vascular lesion in the nasal iris of right eye pushing the normal iris tissue centrally **B**: Ultra-sound biomicroscopy showing a low reflective solid mass arising from the ciliary body and involving the iris **C**: Axial MR-image shows a well defined mass (arrow) is seen in the right eye, located in the ciliary body and iris. The lesion is hyper intense to the vitreous on T1 weighted image with fat suppression **D**: A T1 weighted image shows same lesion with marked enhancement after gadolinium administration



Comment

Mesectodermal leiomyoma of the ciliary body originates from the mesectodermal smooth muscle cells derived embryologically from the neural crest. The Differential diagnosis includes amelanotic melanoma, metastasis, schwannoma

Figure 2 **A**: Photomicrograph of the tumor removed in this case showed elongated spindle cells with oval nuclei. (Hematoxylin and eosin, original magnification x 40) **B**: Tumor cells showing immunoreactivity for CD56 antigen (original magnification x 40) **C**: Photomicrograph of tumor cells showing positive staining with Neuron-specific enolase (NSE, original magnification x 40) **D**: Slit-lamp photograph of the same eye 20 days after the excision of tumor without any iris defect



and adenocarcinoma of ciliary body. It was impossible to distinguish intraocular leiomyoma from amelanotic melanoma without histochemistry [3]. The correlation of marked enhancement of leiomyoma demonstrated after gadolinium administration on T1 weighted images as compared to moderate enhancement in melanoma can be a diagnostic clue for leiomyoma, as suggested by our case also [2,4,5]. To date, immunohistochemistry could find only 3 cases positive for CD 56 and caldesmon while only 2 were positive for NSE [1,6]. Our case also showed positive results for all these markers. Although mesectodermal leiomyoma is a benign lesion, it can grow and cause complications such as lens subluxation and secondary glaucoma, and it can extend extrasclerally. For these reasons and for diagnostic confirmation, surgery is necessary. Transscleral

resection as performed in our case allowed the excision of the tumor without any postoperative iris defect, avoiding potential photophobia, astigmatism or other aberrations or decreased vision, and leads to good cosmetic results.

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