European Journal of Ophthalmology / Vol. 19 no. 2, 2009 / pp. 292-294

Episcleral hemangioma as an isolated finding

HUDA AL-GHADEER[1], ALI A. AL-RAJHI[1], FENWICK C. RILEY[2]

[1]Anterior Segment Division
[2]Pathology and Screening Clinic, King Khaled Eye Specialist Hospital, Riyadh - Kingdom of Saudi Arabia

PURPOSE. *To report on a case of episcleral hemangioma in a child.*
METHODS. *A 3-year-old boy presented with a 4-week history of limbal mass in the left eye and had an excision of that mass.*
RESULTS. *There was an 8 x 7 mm oval lesion, 3–4 mm below the inferior limbus. It was reddish, sharply elevated with smooth surface, and firmly attached to the underlying sclera with two large blood vessels over it. Histopathologic examination demonstrated a cellular lesion containing vascular spaces of various sizes. The solid appearance indicated an increase of cells that appear to be consistent with endothelial cells which formed a nest surrounded by PAS-positive strands. Small to moderate size capillaries were evident throughout the solid portion of the lesion. These findings were consistent with episcleral hemangioma of capillary type.*
CONCLUSIONS. *Episcleral hemangioma is a rare tumor and should be included in the differential diagnosis of episcleral tumors. To our knowledge, this is the first reported case of episcleral hemangioma in a child as an isolated finding. (Eur J Ophthalmol 2009, 19: 292-4)*

KEY WORDS. *Capillary, Children, Episclera, Hemangioma, Vascular tumors*

Accepted: August 14, 2008

INTRODUCTION

Hemangiomas are the most common soft tissue tumors of infancy. The rate of incidence stands at 5–10% in infants (1, 2). They are defined as vascular tumors with a growth phase, marked by hypercellularity and involutional phase (1). They are more frequently found in girls than boys ranging from a 3:1 to 5:1 ratio, and are thought to be common in white patients than in black patients (1). We report on a case of capillary type episcleral hemangioma and its histopathologic findings.

Case report

A 3-year-old boy presented to the emergency room at King Khaled Eye Specialist Hospital with a 4-week history of painless swelling in the left eye, which had gradually increased in size and with no change in visual acuity (VA). The patient fell down from his bicycle 5 weeks prior to his complaint. He was a product of full-term twin pregnancy with no prenatal or postnatal complications. The family history was unremarkable.

Ocular examination showed apparently normal VA in both eyes. Intraocular pressure was within normal limits, right eye was normal. Left eye assessment revealed normal lids, full extraocular movement, and regular round reactive pupil. Both fundi were normal. Slit-lamp examination showed clear cornea and lens, deep and quiet anterior chamber. There was an 8 x 7 mm oval lesion, 3–4 mm below the inferior limbus. It was reddish, sharply elevated with smooth surface, and firmly attached to the underlying sclera with two large blood vessels over it (Fig. 1). Ultrasound biomicroscopy of the left eye revealed a 6 x 7 mm lesion with two small rounded cystic spaces surrounded by homogenous dense opacities. The sclera appeared to be thin in the area of the lesion with no evidence of ciliary body, scleral, or globe involvement. The conjunctiva was dissected from the tumor from the limbal side and the conjunctiva overlying the tumor was also dissected until the boundaries of the tumor were identified which were attached to the sclera. There were two large blood vessels coming inferiorly, one from the nasal and some telangiectatic blood vessels at the limbus, over conjunctiva. These were cauterized and the tumor was

1120-6721/292-03$25.00/0

Presented as a poster at the annual Saudi Ophthalmology 2004 conference (March 7–10); Riyadh, Kingdom of Saudi Arabia.

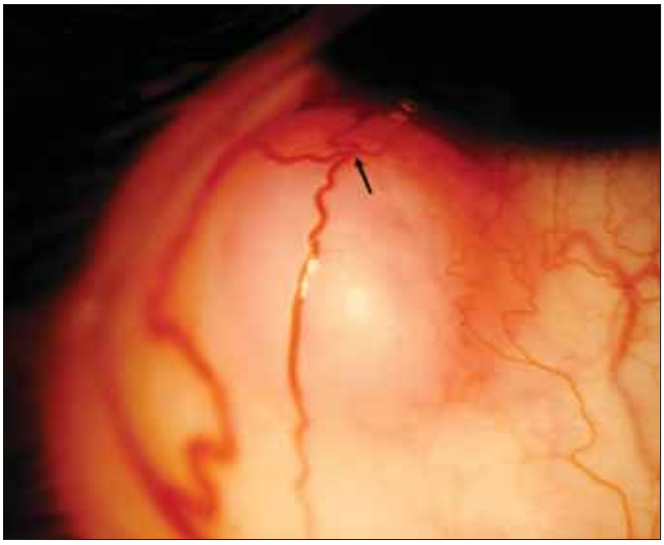

Fig. 1 - *Feeding blood vessel over the lesion as indicated by the arrow.*

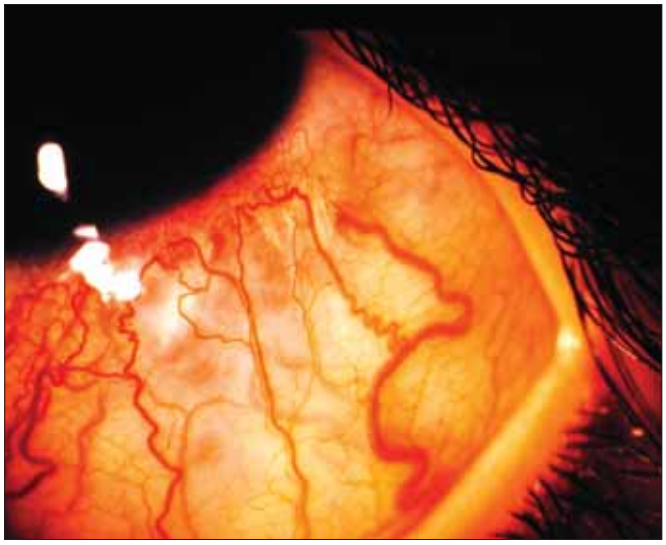

Fig. 2 - *After excision.*

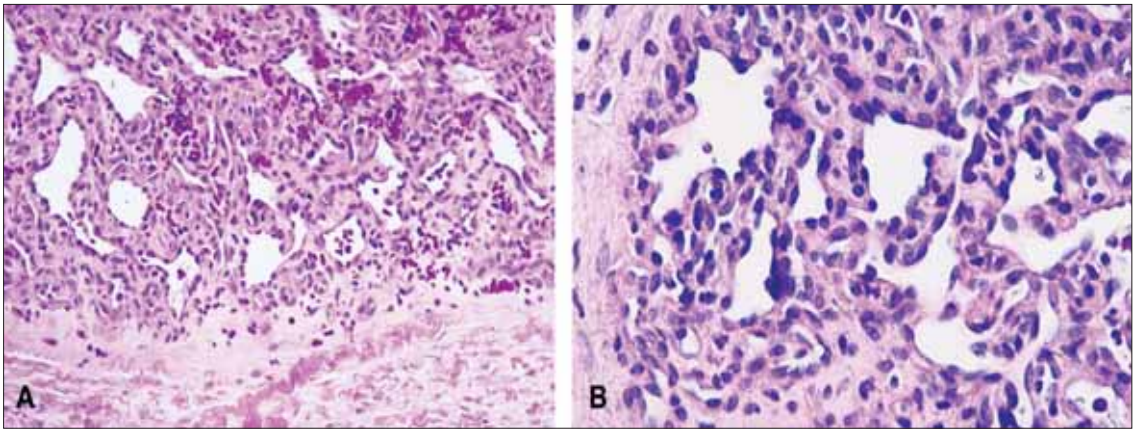

Fig. 3 - (A) *Peripheral areas with capillary proliferation and capsule (H-E x 100).* **(B)** *Capillary proliferation (H-E x 200).*

dissected from the underlying sclera. It was found to be firmly attached to the sclera. The tumor, in our case, was excised completely (Fig. 2) and obtained for histopathologic study.

Histopathologic findings demonstrated a cellular lesion containing vascular spaces of various sizes which was surrounded on three sides by a dense fibrous pseudocapsule. No epithelium was associated with the lesion. The solid appearance indicated an increase of cells that appear to be consistent with endothelial cells which formed a nest surrounded by PAS-positive strands (Fig. 3A). Small to moderate size capillaries were evident throughout the solid portion of the lesion (Fig. 3B). In some areas, these vascular spaces become a bit larger and appeared to have a venous architecture. In half of the specimen, a large vessel seems to be extending through the deeper portion of the lesion. These lesions were lined with flattened endothelial cells which appeared to be associated with neural cells. They were consistent with a large venule. Lymphocytes were scattered all through the lesions. Immunohistochemistry was done for von Willebrand factor (factor VIII), CD 34, and smooth muscle actin. Both von Willebrand and CD 34 were positive in the areas surrounding the vascular spaces, suggesting the origin is vascular endothelium.

DISCUSSION

Episcleral tumors are common but tumors of the sclera are rare. They may originate from the episcleral, vascular, or nervous tissues which pass through the extension of the lesions of conjunctiva or inner eye or may be a manifestation of systemic neoplastic process (3).

In early stages of the vascular tumors, they can be seen as a dilatation of some of the vessels of the deep episcleral network. In some congenital varieties, these anomalies can be extensive. They regress with age as can be seen in the cases of skin hemangioma (4). However, angiomas arising within the episclera resemble an amelanotic melanoma which, sometimes, cannot be distinguished except by biopsy. The new vessels which are formed in the tumor itself appear to radiate from the mass rather than evade it, as in the inflammatory conditions (4). Hence, it is suggested that trauma may be responsible for their clinical appearance or increase in growth (4).

Episcleral hemangioma is regarded as one of the ocular manifestations of Sturge-Weber syndrome. These lesions are often not clinically apparent in early childhood but become noticeable upon slit-lamp examination in the second half of the first decade. There were no signs of this syndrome in our patient.

Takahashi reported two cases of simple scleral hemangioma, one occurring in a 56-year-old woman, and the other in a 19-year-old man. The latter was associated with an incomplete form of Sturge-Weber syndrome. Slit lamp microscopy had revealed that these cases seem to be telangiectasias of capillaries with microaneurysms along the limbus (5). Currently, they are thought to result from an arrest in the development of the mesenchymal primordial in the earliest stage of vascular system formation (6).

Chang et al reported a case of diffuse neonatal hemangioma with eyelid, conjunctival, and iris hemangioma. The findings of histopathology disclosed capillaries of varying size which was similar to the findings in our case (7).

Absence of typical symptoms and signs of inflammatory conditions such as nodular episcleritis led to the decision to excise the tumor rather than attempt topical therapy. The histopathologic and immunohistochemical results support our diagnosis.

There was no recurrence of this lesion in our patient 3 years after presentation.

In conclusion, episcleral hemangioma, a rare tumor, should be included in the differential diagnosis of episcleral tumors. We are unaware of prior reports of episcleral hemangioma in a child as an isolated finding.

ACKNOWLEDGEMENTS

The authors thank Dr. Abdullah Al-Kandari for assistance with immunohistochemistry.

None of the authors has any proprietary or financial interest or conflicts with respect to any of the material presented in the article.

Reprint requests to:
Huda Al-Ghadeer, MD
Member, Anterior Segment Division
King Khaled Eye Specialist Hospital
PO Box 7191
Riyadh 11462, Kingdom of Saudi Arabia
hudagh2001@yahoo.com

REFERENCES

1. Drolet BA, Esterly NB, Frieden IJ. Hemangiomas in children. N Engl J Med 1999; 341: 173-83.
2. Sundine MJ, Wirth GA. Hemangiomas: an overview. Clin Pediatr 2007; 46: 206-21.
3. Watson PG, Hazleman BL. Other Affections of the Sclera. The Sclera and Systemic Disorders. London: W.B. Saunders, 1976; 347-81.
4. Watson P. Diseases of the sclera and episclera. In: Tasman W, Jaeger E, eds. Duane's Ophthalmology, Vol. 4. Philadelphia: Lippincott Williams & Wilkins, 1995; 1-45.
5. Takahashi H. Two cases of simple scleral hemangioma. Nippon Ganka Kiyo 1966; 17: 671-5.
6. Kiratli H, Bilgic S, Caglar M, Soylemezoglu F. Intramuscular hemangiomas of extraocular muscles. Ophthalmology 2003; 110: 564-8.
7. Chang CW, Rao NA, Stout JT. Histopathology of the eye in diffuse neonatal hemangiomatosis. Am J Ophthalmol 1998; 125: 868-70.