

Haemangiopericytoma of the eyelid and orbit: report of 2 cases

M. B. Alhassan, F. Kyari and *A. Mohammed

Department of Clinical Ophthalmology, National Eye Centre, Kaduna and*Department of Pathology, Ahmadu Bello University Teaching Hospital, Zaria, Nigeria

Reprint requests to: Dr. Fatima Kyari, Department of Clinical Ophthalmology, National Eye Centre, P. M. B. 2267, Kaduna, Nigeria. E-mail: fatimayk@hotmail.com

Abstract

Haemangiopericytoma is a rare tumour of the pericytes. This is a report of two adult Nigerians presenting with large orbital masses confirmed at histology to be haemangiopericytoma of the eyelid and orbit respectively. Massive proptosis in both patients produced a clinical diagnostic dilemma, which was only resolved by histopathological studies.

Key words: Haemangiopericytoma, eyelid, orbit, tumour

Introduction

Haemangiopericytoma is an uncommon tumour and its occurrence in the ocular adnexae is even more so. It is more frequently encountered in the lower extremities and retroperitoneum, usually occurring in middle-aged adults, with a mean age of 42 years.¹ With the orbito-ocular tumour, patients present with progressive proptosis, palpable mass, diplopia, decrease in vision and varying amounts of pain. The onset and progression are usually more rapid than with cavernous haemangioma. The tumour arises from the pericyte, which occupies a position outside the vascular endothelial cell and in close apposition to it in-capillary and post-capillary venules.

We discuss the clinical pathological findings in two Nigerian patients with

haemangiopericytoma, one of the lid and the other of the orbit.

Case reports

Case 1

A 31-year old man presented with an 18-month history of gradually increasing swelling in the right upper eyelid. Systemic review and physical examination were unremarkable. View of the right globe was precluded by the huge mass (figure 1). The left eye, with a visual acuity (VA) of 6/6, was essentially normal. A firm, non-tender, huge mass was noted in the right upper eyelid, and attached to the overlying skin with no pulsations or bruits. The right orbital rim was normal. Because it was difficult to retract the eyelid, examination of the right eye was only possible post-operatively,

having a VA of 6/18 showing normal cornea, anterior and posterior segments.

Orbital X-rays showed normal orbital bones and outline. The mass was excised with inadvertent partial rupture of the capsule. Histology showed; partially encapsulated yellowish tumour measuring 4x3x3.5x2.5 centimetres, weighing 27 grams. Cut section showed white and yellow appearances. Light microscopy (figure 2) showed a tumour composed of sheets of ovoid to spindle cells arranged in indistinct lobules. There are thin vascular spaces with tumour cells outside the vessels and no areas of sclerosis.

Case 2

A 20-year old woman presented with gradual, painless and progressive protrusion of the left eye, displaced downwards and inwards, of 2 years duration. There was associated gradual visual loss with an initial brief period of binocular diplopia. Past ocular history, general systemic review and examination were unremarkable. Best-corrected visual acuities were OD 6/6 and OS light perception only. Ocular examination revealed an essentially normal right eye. The left eye had a massive proptosis with medial and inferior displacement of the

globe of 25 millimetres. There was exposure keratopathy, a formed anterior chamber and a relative afferent pupillary defect. The hazy cornea precluded meaningful visualization of the posterior segment. There was a palpable, firm, non-tender, non-pulsatile mass in the superior temporal quadrant of the left orbit, with no bruits. We made a clinical impression of a benign lacrimal gland tumour.

Plain orbital and skull X-rays showed enlargement of the left orbit with a dense soft tissue shadow supero-temporally and excavation of the left lacrimal fossa. The orbital bones did not have any defects or abnormal calcifications. A Computed tomographic (CT) scan was not available. The mass was excised en bloc. The patient was last seen 2 months post-operatively with OS VA of 5/60 and features of optic neuropathy and no obvious tumour recurrence. Histology showed; nodular tissue measuring 5x4.8x3.8 centimetres and weighing 31 grams. The cut surface revealed a solid tumour that is well encapsulated. Light microscopy (figure 3) showed an encapsulated vascular tumour of fairly regular spindle cells around thin vascular spaces some of which have a stag-horn pattern.

Figure 1: Right eyelid mass in Case 1



Figure 2: Sheets of ovoid to spindle cells arranged in indistinct lobules with thin vascular spaces (right - H & E x 25; left reticulin x 25)

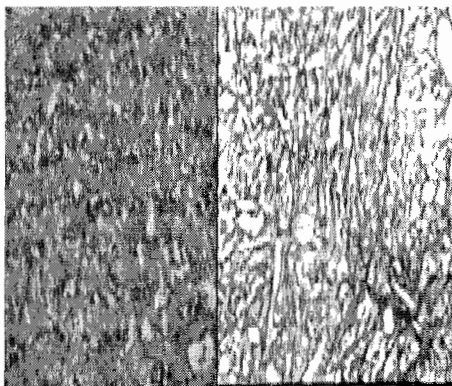
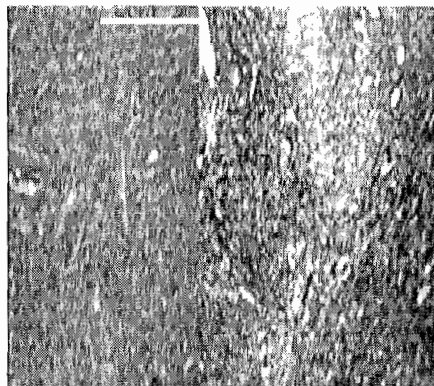


Figure 4: A vascular tumour of fairly regular spindle cells around thin vascular spaces some with stag-horn pattern (right - H & E x 25; left - reticulin x 25)



Discussion

Haemangiopericytoma is a rare tumour arising from the pericytes and occurs more frequently in the lower extremities and retroperitoneal areas.² Its occurrence in the nasal cavity has been reported.³ However, its occurrence in the eye and periorcular tissues is much rarer. Haemangiopericytoma has been reported to have occurred in the orbit,^{1,4,5} lacrimal sac,⁶⁻⁸ lacrimal gland,⁹ conjunctiva,^{10,11} superciliary,¹² and ciliary body.¹³ Hospital-based reviews of childhood malignancies of the eye and orbit¹⁴ and orbito-ocular tumours in Nigerians¹⁵ have not specifically reported the occurrence of haemangiopericytoma. More cases reported in the orbit seem to have a predilection for the superior orbit. Though our pre-operative imaging techniques were restricted due to cost and unavailability of a CT scan, the physical findings in the second patient were suggestive of a likely origin of the tumour in the lacrimal gland. However, not in all cases can only ocular examination be adequate in suggesting the tumour origin since orbital haemangiopericytoma has been reported to simulate an intraocular tumour.¹⁶

Kapoor and co-workers have reported haemangiopericytoma in the orbit of a 3-year old,¹⁷ but it commonly affects the middle-aged with no sex predilection. Here, our patients were aged 31 years and 20 years. The slow growing nature of the tumour is seen in case 2 that presented with a long history and a typically gradual, painless, progressive proptosis. The slow growth suggests a benign neoplasm. Haemangiopericytoma is usually well encapsulated but may be partially encapsulated, and may be aggressive and potentially malignant.¹⁸ The malignant types have been reported with systemic metastasis, especially after incomplete removal. Its biologic behaviour is uncertain. An effort has been made to determine the clinicopathologic characteristics that may correlate with the biologic behaviour.¹ The degree of cellularity, nuclear atypia, mitotic activity, tumour size and the presence of haemorrhage and necrosis were evaluated. The investigators did not find a basis for firm conclusion with recurrence and metastasis reported in all histological types.¹ The lack of unequivocal correlation between the histological features of haemangiopericytoma and its clinical behaviour is consistent with the unpredictable behaviour of this neoplasm. Thus, a total excision is advised in order to reduce the risk of recurrence. Likewise, long-term follow-up is very important in order to monitor and treat the patient for any recurrences, which can occur even later than 30 years after excision.¹⁹

Though the clinical and histopathologic picture of our patients seem that of the benign type, there is a strong need for us to have a long follow-up of these patients. This is usually a difficult venture in our practice as patients are soon lost to follow-up especially that it is the dramatic cosmetic effect of tumour excision is often the most appreciated aspect of treatment. The longer follow-up is that of 2 months in our second patient. We, however, strongly

believe that they will present again if there is a recurrence.

The mainstay of treatment of haemangiopericytoma has been complete surgical excision. Exenteration is not ordinarily indicated. Radiotherapy has been effective in controlling recurrences with long-term disease-free period after treatment, while the role of chemotherapy is yet to be ascertained.

Other tumours that mimic haemangiopericytoma histologically include spindle-cell lipoma, neurilemma, angiolipoma and liposarcoma. Immunohistochemistry is useful in their histological classification.

References

1. Croxatto J. O, Font R. L. Haemangiopericytoma of the orbit: a clinicopathological study of 30 cases. *J Hum Pathol* 1982; 13: 210-218.
2. Ezinger F. M, Smith B. H. Haemangiopericytoma: an analysis of 106 cases. *Lum Pathol* 1976; 7: 61-82.
3. Alli A. F, Singh S. P. Haemangiopericytoma in the nasal cavity in Ibadan, Nigeria. *J Laryngol Otol* 1972; 86:405-410.
4. Jakobiec F. A, Howard G. M, Jones L. S. et al. Haemangiopericytoma of the orbit. *Am J Ophthalmol* 1974; 78: 816-834.
5. Sullivan J. J, Wright J. E, Wulc A. E. et al. Haemangiopericytoma of the orbit. *Aust N Z J Ophthalmol* 1992; 20: 329.
6. Rubin P. A, Shore J. W, Jakobiec F. A. et al. Haemangiopericytoma of the lacrimal sac. *Ophthalmic Surg* 1992; 23: 562-563.
7. Roth S. I, August C. Z, Lissner G. S. et al. Haemangiopericytoma of the lacrimal sac. *Ophthalmology* 1991; 98: 925-927.
8. Carnevali L, Trimarchi F, Rosso R. et al. Haemangiopericytoma of the eyelid. *Am J Ophthalmol* 1991; 111: 100-102.

9. Redmond R. M, Mannor G. E, Garner A. et al. Lacrimal gland haemangiopericytoma. *Am J Ophthalmol* 1995; 119: 99-100.
10. Grossniklaus H. E, Green W. R, Wolff S. M. et al. Haemangiopericytoma of the conjunctiva: two cases. *Ophthalmology* 1980; 93: 265-267.
11. Sathya S, Sampath R, Bonthek R. E. et al. Conjunctival haemangiopericytoma. *Br J Ophthalmol* 1994; 78: 497-499.
12. Brown H. H, Brodsky M. C, Hembre K. et al. Supraciliary haemangiopericytoma. *Ophthalmology* 1991; 98: 378-382.
13. Gieser S. C, Hufmayer T, Jaros P. A. et al. Haemangiopericytoma of the ciliary body. *Arch Ophthalmol* 1988; 106: 1269-1272.
14. Abiose A, Adido J, Agarwal S. C. Childhood malignancies of the eye and orbit in northern Nigeria. *Cancer* 1985; 55: 3889-3893.
15. Ogunbiyi O, Williams A. O. Ocular and lacrimal tumors in Nigeria. *J Cancer* 1972; 30:580-587.
16. Aminzadeh M, Shields J. A, Shields M. P. et al. Haemangiopericytoma of the orbit. *Br J Ophthalmol* 1999; 83: 756-757.
17. Kapoor S, Kapoor M. S, Boushra M. et al. Orbital haemangiopericytoma in a 3-year-old child. *Indian J Ophthalmol* 1978; 26: 40-42.
18. Henderson W. F. et al. Primary orbital haemangiopericytoma: an aggressive and potentially malignant neoplasm. *Arch Ophthalmol* 1978; 96: 666-671.
19. Rice G. D, Kersten R. et al. Bilateral orbital haemangiopericytoma. *Arch Ophthalmol* 1982; 100: 552-556.
20. Setzkorn B, Kai Lee D, Jiff N. et al. Haemangiopericytoma of the orbit: a case report. *Am J Ophthalmol* 1991; 111: 100-102.

- al. Haemangiopericytoma of the orbit treated with conservative surgery and radiotherapy. *Arch Ophthalmol* 1987; 105: 1103–1105.
21. Borg M. F, Benjamin C. S. A 20-year review of haemangiopericytoma in Auckland, New Zealand. *Clin Oncol* 1994; 6: 371-376.