Unusual Presentation of a Capillary Haemangioma: Case Report

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Introduction: Capillary haemangiomas are benign endothelial cell neoplasms and are the most common tumours of the orbit and periorbital area in childhood[1,2]. They are believed to be due to harmatomatous proliferation of vascular endothelial cells[2]. They may present as small isolated lesions of minimal clinical significance, large disfiguring masses that may cause visual impairment, and/or systemic complications or could be part of a clinical syndrome[1,3]. Thirty percent present in the first few weeks of life. The tumour usually undergoes an initial rapid growth phase over the first 3-6 months, then a phase of stabilization after the age of 1 year, and then the phase of spontaneous regression (30-50% by 3years and 60-75% by 4-7 years)[1]. We report an unusual presentation of capillary haemangioma in an adult.

Case Presentation: A 36 year old man who presented four years ago with a two year history of a right eye swelling. It progressively increased in size; it was first noticed in the medial aspect of the upper and lower lid with eventual supero-temporal dystopia. There was no history of change in size with Valsalva maneuver nor swelling in the other eye or any other part of his body. There was progressive visual deterioration. There was redness, tearing, discharge, and excessive itching. However, there was no eye pain, head ache, dizziness, seizures or loss of consciousness. There was no history suggestive of any form of bleeding disorder, wheezing, breathlessness, easy fatigability, fainting attacks. Thirteen years prior to presentation, he had a similar swelling of about three years duration in the same eye and underwent a surgical excision in a private facility in Kaduna, with a histological diagnosis of a Capillary Haemangioma. There is a positive family history of similar lid swelling in patient's father, paternal uncle and grandmother with spontaneous regression. However details of the onset and other characteristics of these swellings in the family members could not be obtained. General examination revealed essentially normal findings. He had a visual acuity of "Counting Fingers" at 1 meter and 6/5 on the right and left respectively. There was a 34cm non axial proptosis in the right eye with supero-temporal displacement of the globe. Superiorly, there was a mass measuring about 2x2cm; and inferiorly there was a 6x2x1cm mass (Figure 1). They were both cystic, non-tender and mobile with normal overlying skin. The conjunctiva was chemosed, and cornea was hazy due to stromal oedema. Further view was precluded. The left eye appeared normal. A clinical diagnosis of Recurrent Orbital Capillary Haemangioma was made. Differentials diagnoses entertained include: orbital carvenous venous haemangioma, orbital venous varix, pleomorphic adenoma.

All haematological investigations done were normal. Orbital Computer Tomography scan showed a well circumscribed, Isodense right orbital cavity mass with soft tissue swelling anteriorly (Figure 2).

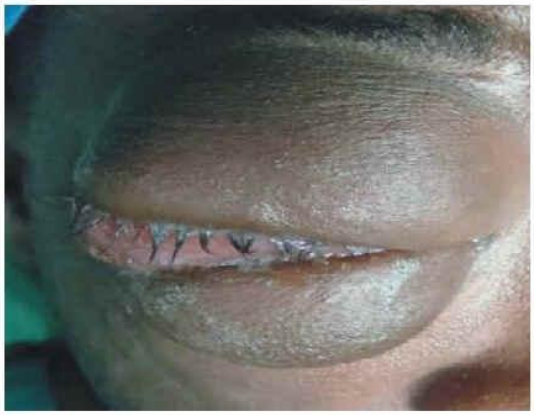

Fig. 1: Preoperative appearance of the right orbital mass

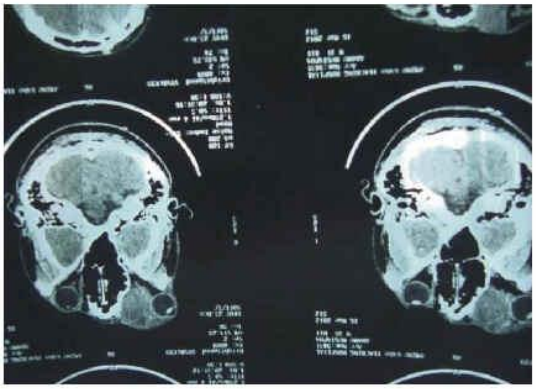

Fig. 2: An axial computed tomographic scan showing a well circumscribed iso-dense lesion with lateral displacement of the right globe. The globe was intact. There was no bony erosion

He was worked up for Orbitotomy under general anaesthesia, the tumour was removed en block (Figure 3) via a medial approach and haemostasis secured by packing. Biopsy was taken for histology. He had an uneventful post-operative period, immediate post-operative visual acuity improved to "Counting Fingers" at 2metres, and there was resolution of swelling and chemosis. There was Keratoconus, central corneal opacity, and a glimpse of a pink disc was seen but it was difficult to make out the fundus details. He was discharged 11 days after surgery and had four follow up visits.

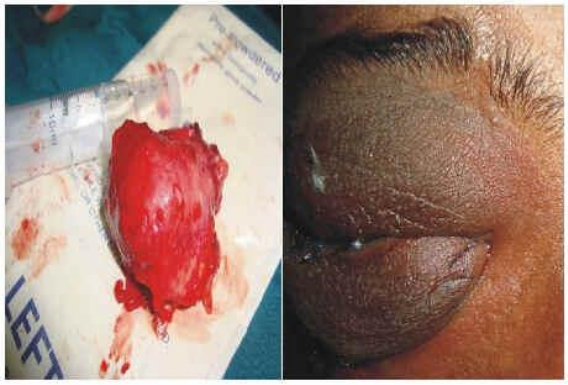

Fig. 3: Mass delivered en block and picture on the 1st post-operative day with a lot of redundant skin in the upper and lower lid

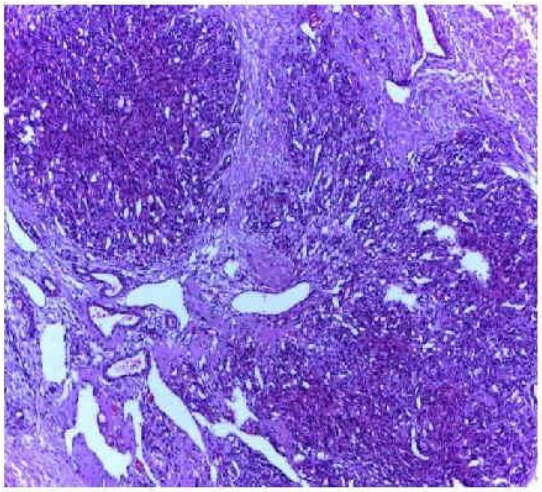

Fig. 4: Photomicrograph showing the histological findings of capillary haemangioma

SPECIMEN: *Received a soft, irregular grey brown tissue that measures 4x3.5x3cm and weighted 22g. Cut sections show tan brown microcystic appeareances. PEx4*

MICROSCOPIC DESCRIPTION: *Sections shows a fairly encapsulated lesion composed of mostly capillary-sized vascular channels containing red blood cells with foci showing dilated vascular channels at the periphery of the lesion. The stroma is fibrous and infiltrated by mononuclear cells. Other areas shows lobules of mature adipocytes and haemorrhage.*

The last visit was three months after surgery. Examination at the last visit revealed visual acuity of "Counting Fingers" at 2m, central corneal opacity, and keratoconus. Histology showed findings consistent with capillary haemangioma (Figure 4).

Discussion: From this report the unusual features of presentation include: Adult Presentation– based on documented natural history, this presentation probably accounts for the outlying 25% that failed to spontaneously regress after 7 years of age[1,2]. The initial swelling was noticed when patient was about 16 years old and the recurrent swelling was noticed at thirty years of age. The index patient is a Male. This finding is at variance with documented female preponderance (documented Male: Female ratio = 1:3)[2]. Patri-lineal history of similar presentation should arouse suspicion of some hereditary pattern not yet reported. Astigmatism has been reported in 20 – 46% of peri-ocular capillary haemangioma[5]. Keratoconus is usually associated

with severe astigmatism. As may be the case in this report it is noteworthy that large haemangiomas are associated with functional defects such as amblyopia, strabismus and optic atrophy[3,4].

References

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