Orbital Alveolar Rhabdomyosarcoma Mimicking a Capillary Haemangioma: A Case Report

Abdulsalam HO[1], Chinda D[1], Abubakar ML[2], Jimoh AO[3] Aliyu HO[4] Umar SS[5]

[1]Department of Ophthalmology, Ahmadu Bello University & Ahmadu Bello University Teaching Hospital, Zaria
[2]Division of Plastic surgery, Department of Surgery, Ahmadu Bello University & Ahmadu Bello University Teaching Hospital, Zaria
[3]Division of Neurosurgery, Department of Surgery, Ahmadu Bello University & Ahmadu Bello University Teaching Hospital, Zaria
[4]Department of Pathology, Ahmadu Bello University & Ahmadu Bello University Teaching Hospital, Zaria
[5]Department of Radiotherapy and Oncology, Ahmadu Bello University & Ahmadu Bello University Teaching Hospital, Zaria

Corresponding author: *Abdulsalam HO Email: halimaabdulsalami@yahoo.com*

Introduction: Rhabdomyosarcoma (RMS) is the most common malignant tumour of childhood[1]. Orbital RMS is a rare tumour with annual incidence of 4.3 cases per million children [1]. In approximately 10% of cases the primary site is the orbit [2]. Orbital RMS is usually seen in the first decade of life. Primary orbital RMS involves the orbit, eyelid, conjunctiva, and rarely, the uveal tract. The typical presentation is a rapid onset of unilateral proptosis, eyelid edema and ptosis.
Capillary haemangioma (CH) is a common benign orbital vascular tumour of childhood characterized by appearance at birth or shortly after birth, rapid increase in size for 6 to 12 months and then gradual involution over 5 to 7 years [3]. CH may cause proptosis or globe displacement [4].

Case Presentation: A twelve-year-old girl, presented with right eye protrusion of seven years' duration. As the protrusion increased in size, she gradually lost vision in the eye. The left eye was normal.
There was no history of headache, syncopal attacks and seizures or remarkable weight loss, there was no swelling of any other part of the body. She had presented four years earlier with three years' history of protrusion, was assessed and diagnosis of capillary haemangioma (based on clinical findings and imaging) was made at the time. She responded to oral propranolol with some regression of the proptosis, however, she defaulted from follow up.

Figure 1: The patient with massive proptosis of the right eye

Figures 2&3: Angiogram showing dilated right facial artery, right superficial temporal artery and left facial artery

On examination, there was proptosis of the right eye, with a mass measuring 12 cm X 14cm (Figure 1). The mass was firm, mildly tender with visible dilated superficial vessels on the upper lid. There was severe chemosis of the conjunctiva and the cornea was opaque. The left eye was essentially normal. There were no masses on the scalp, no

significant pre auricular, post auricular, mandibular or cervical lymphadenopathy. A clinical assessment of capillary hemangioma was made. Computed tomographic angiography showed dilated right facial artery, right superficial temporal artery and left facial artery (Figures 2 and 3).

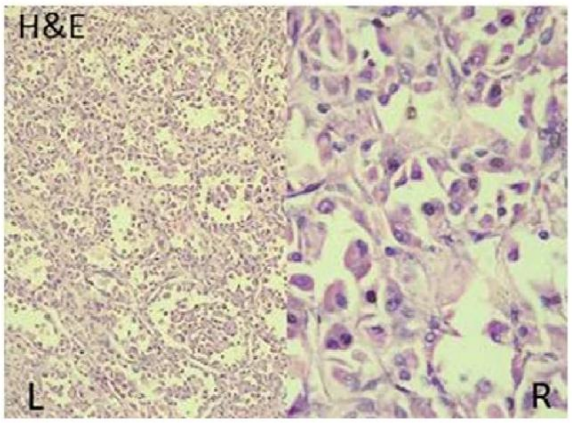

Figure 4: Micrograph showing cells in nests and diffuse sheets, pleomorphic cells with eosinophilic cytoplasm. L: X100, R: X400. These findings were consistent with that of Alveolar Rhabdomyosarcoma.

The management of the patient was multidisciplinary, involving the Ophthalmologists, Neurosurgeons, Plastic surgeons and Radio-oncologist. She had a two staged surgery: first, the feeding branches of the superficial temporal and facial arteries were ligated. A modified exenteration was done two weeks later. Histopathology of the mass showed pleomorphic cells with eosinophilic cytoplasm, in nests and diffuse sheets strongly suggestive of Alveolar Rhabdomyosarcoma (Figure 4). The patient was diagnosed according to the International Rhabdomyosarcoma Study Group (IRSG) as Stage I, Group II with microscopic residual disease remaining after surgery.

Discussion: This patient presented with a tumour which was purely orbital, causing uniocular proptosis, at five years of age. The mass increased in size to become massive, it was highly vascular as showed in the angiogram. There was expansion of the bony orbit due to the size of the tumour. These characteristics can suggest either CH or and RMS; and growth was slow, over seven years. Also, earlier investigation had diagnosed CH and she had responded to systemic Propanolol (a feature unique to CH). The clinical diagnosis of CH was made based on these reasons. Rhabdomyosarcoma is a close differential, however, it is typically a fast growing highly invasive tumour, though it can also be very vascular [5,6].

Conclusion: Orbital CH and RMS can have very similar features. To the best of our knowledge this might be the first case of ARMS mimicking CH to be reported. Therefore, ophthalmologists should always keep an open mind, especially in long standing cases like this one. RMS can be slow growing as seen in this case.

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