Introduction

Persistence in terminological confusion of vascular anomalies has lead to inaccurate diagnosis and treatment. These lesions can occur anywhere in the body, and treatment becomes complicated when they are situated in the craniofacial region. An accurate diagnosis with appropriate investigations is of vital importance, and the use of the standardized craniofacial approach will result in a satisfactory outcome, as illustrated in this case report.

Case Report

A 17 year-old female presented with a 3-month history of itchiness and redness in the left eye with associated swelling. A CT scan showed a mass lesion in the left orbit. An attempt of excision was carried out at another institution through a lateral orbitotomy via a skin incision. This procedure was abandoned when profuse bleeding was encountered. The histology of a biopsy taken at the time was reported as a ‘cavernous haemangioma’. The patient was subsequently referred to our Centre. Examination revealed left globe dystopia, as well as an obvious temporal scar (Fig. 1). She had a normal visual acuity, and a full range of extra-ocular motion. Review of the CT scan showed a 2cm mass containing a phlebolith (arrow) in the floor of the orbit, displacing the globe upwards (Fig. 2). An MRI scan (Fig. 3) showed the mass, closely related to the inferior rectus muscle displacing the globe upwards. The lesion enhanced brightly with Gadolinium contrast on T1 sequences, and on T2 images. The phlebolith could be seen within the lesion (arrow). These features are those of a venous malformation.

Histology of the first specimen comprised fragments of bone & a fragment of grey-tan tissue in which there were dilated ectatic blood vessels lined by thin endothelium separated by fibrous stroma (Fig 6a). Histology of the definitive resection specimen showed similar features to the first specimen, with several large vascular channels & numerous small vessels within fibrous scar tissue. Immunoperoxidase staining for smooth muscle actin showed smooth muscle within the large vessels, the small vessels within the scar tissue & some of the dilated ectatic vessels (Fig 6b).

Discussion

This case highlights the importance of proper investigations to achieve an accurate diagnosis prior to treatment. The diagnosis of vascular anomalies can be made clinically by careful history and physical examination in most cases without invasive studies. Appropriate radiological studies are necessary to evaluate lesions in the craniofacial region. An MRI scan with Gadolinium contrast is considered the single most informative investigation. It verifies the clinical diagnosis, demonstrates the flow characteristics of the lesion and provides accurate anatomical details.