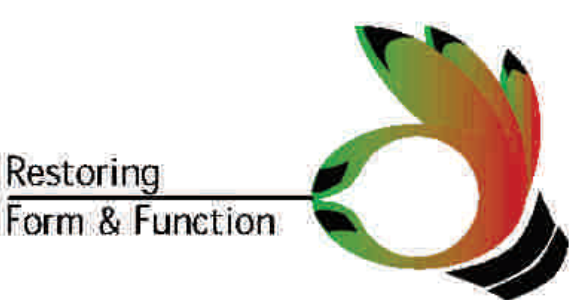


Venous Malformation of The Orbit

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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.



Fig. 1

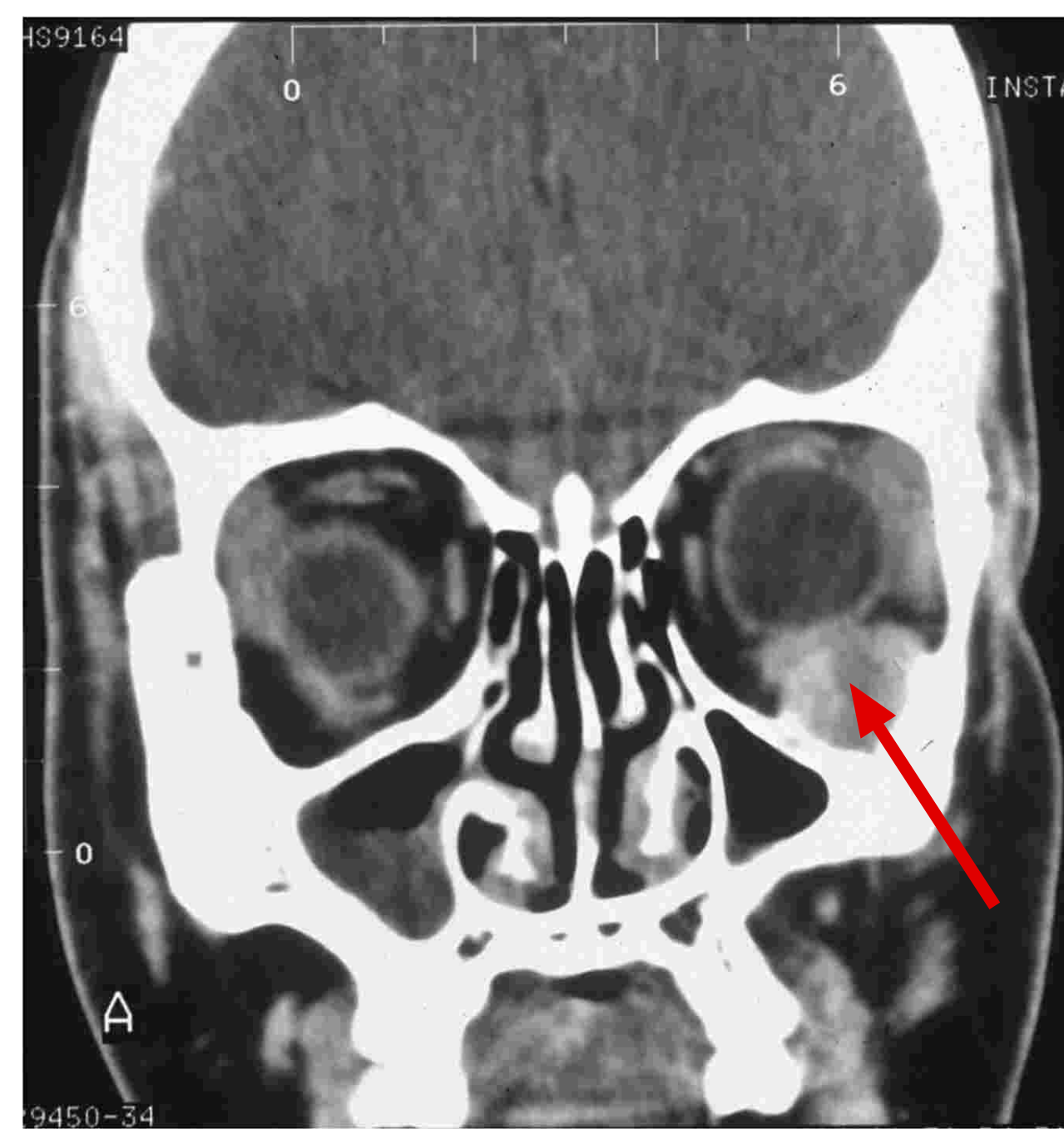


Fig. 2

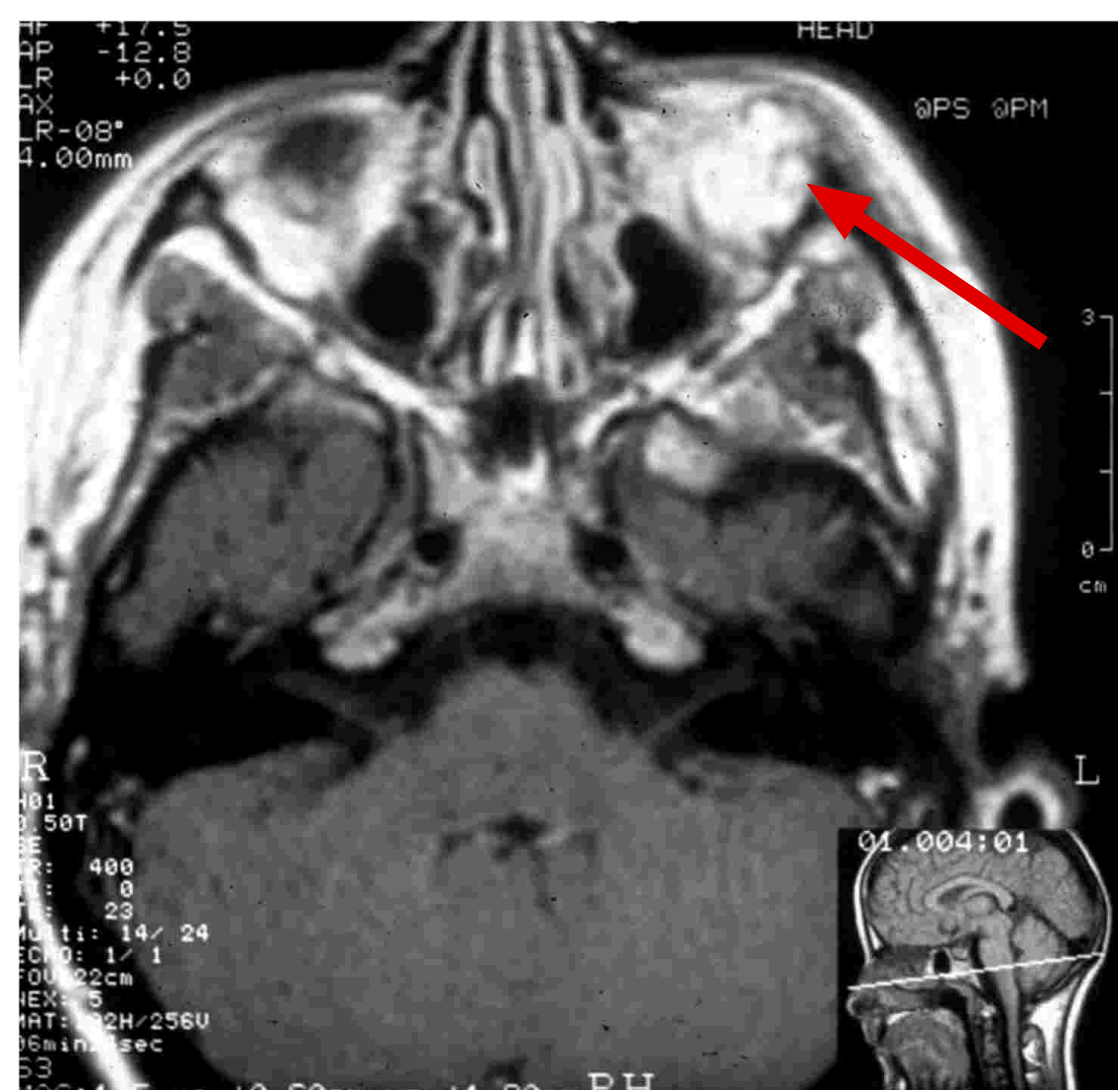


Fig. 3

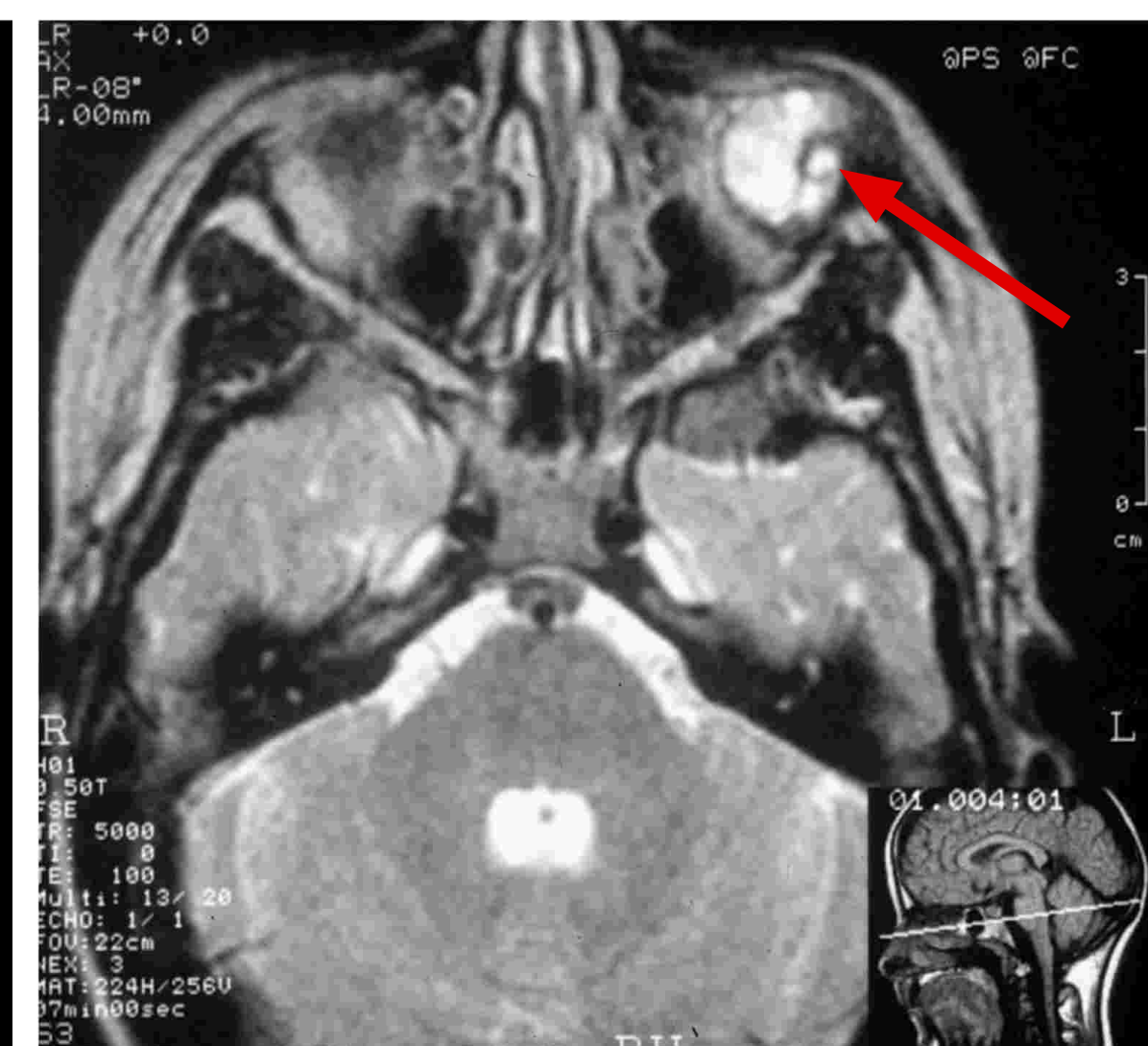


Fig 4

The patient underwent resection of the venous malformation through a subciliary incision (Fig 5). A sub-periosteal dissection was carried out along the lateral and inferior orbital margins, the floor and lateral orbital wall. Excision of the lesion was facilitated by excision of a bony spur at the inferior orbital margin.

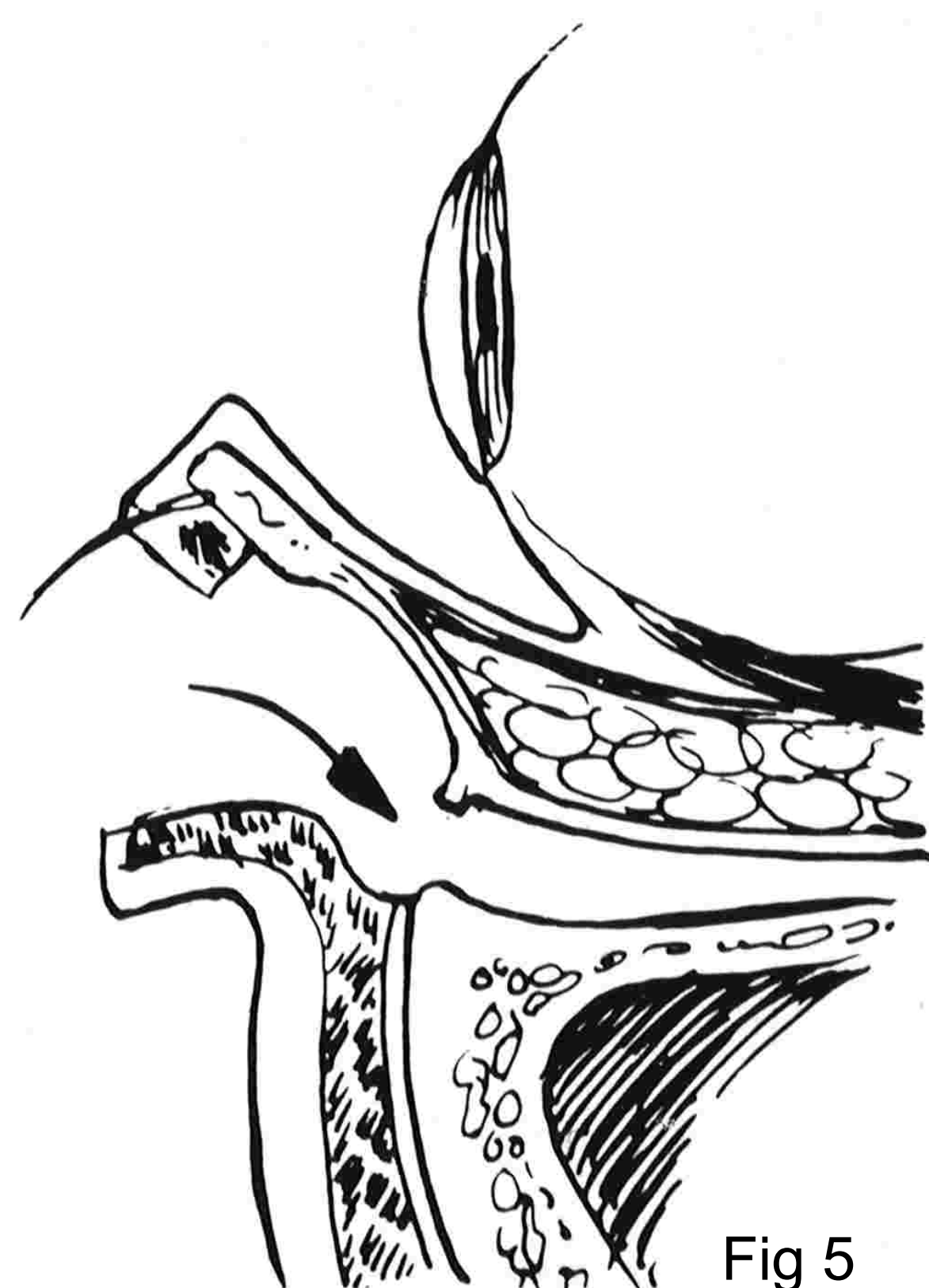


Fig 5

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).

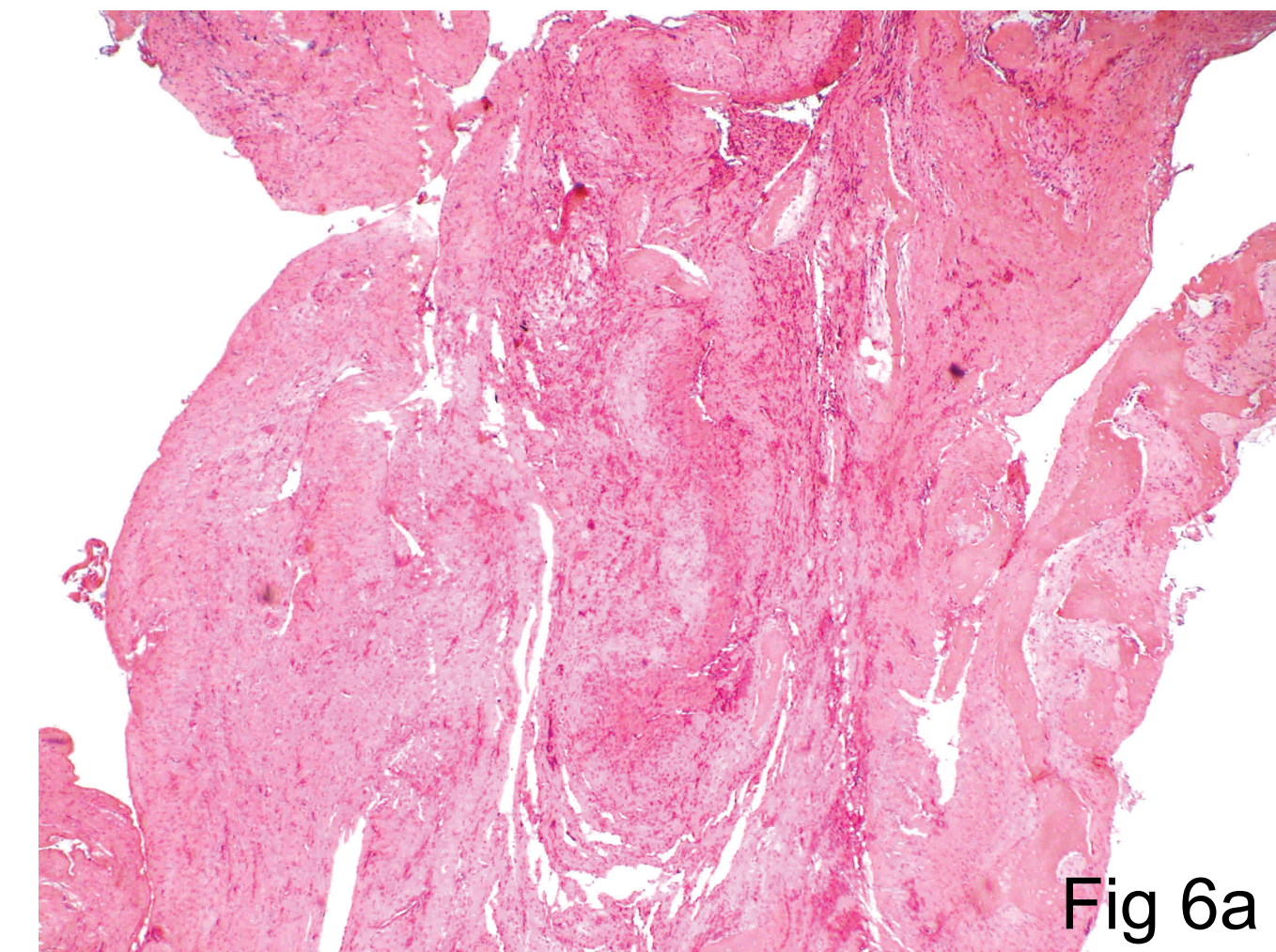


Fig 6a
Photomicrograph showing large & small blood vessels within woven bone. (H&E, Original magnification x40)

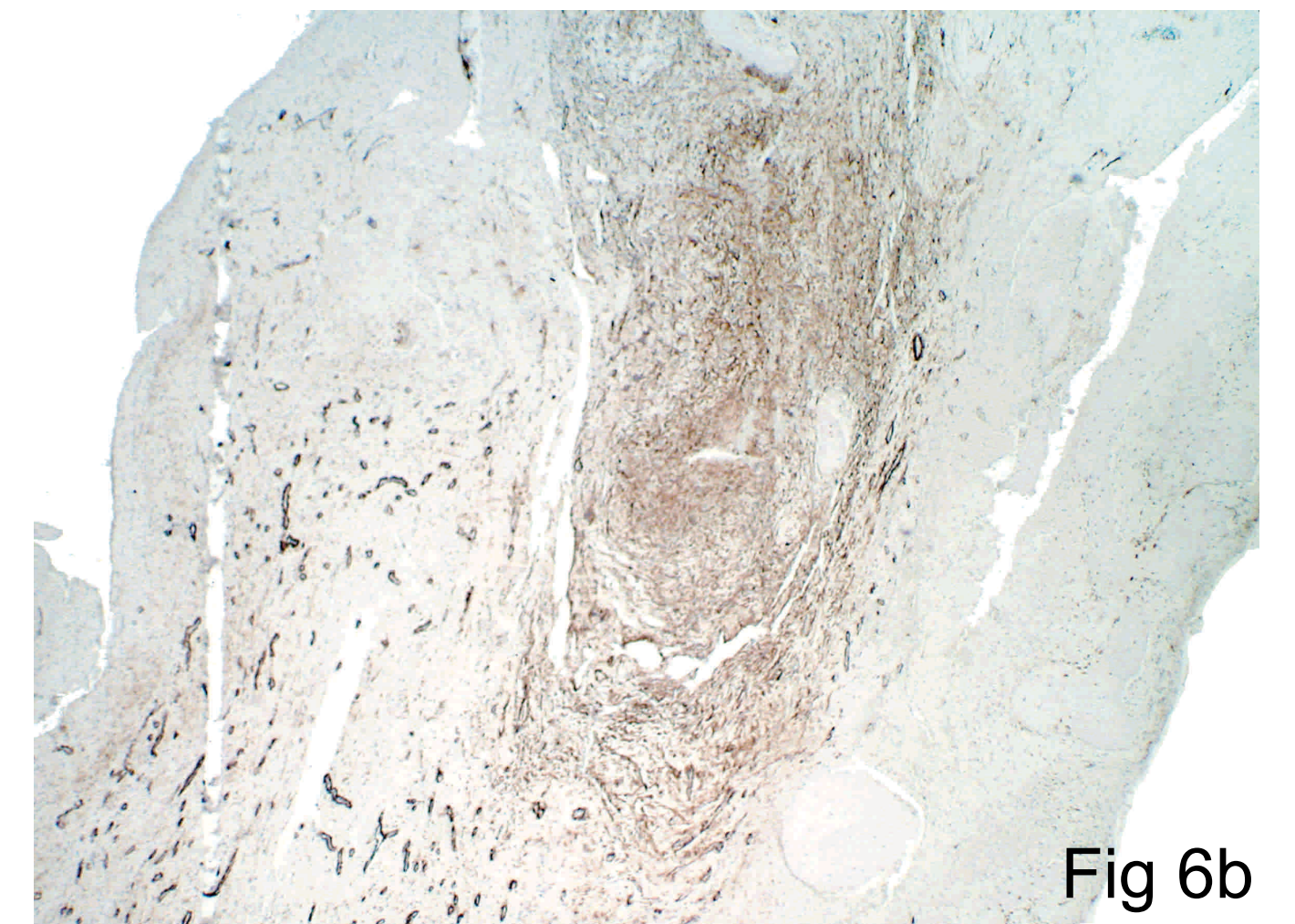


Fig 6b
Photomicrograph showing positive staining for smooth muscle actin within the walls of large & small blood vessels. (Original magnification x40)

There was no evidence of persistence of the venous malformation, both clinically and on repeat MRI scan at follow-up 2 years later. She had normal visual acuity and a full range of extra-ocular motion without diplopia. The left globe was in a satisfactory position (Fig 7a). The visible temporal scar had improved, but remained conspicuous (Fig 7b).



Fig 7a



Fig 7b

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.

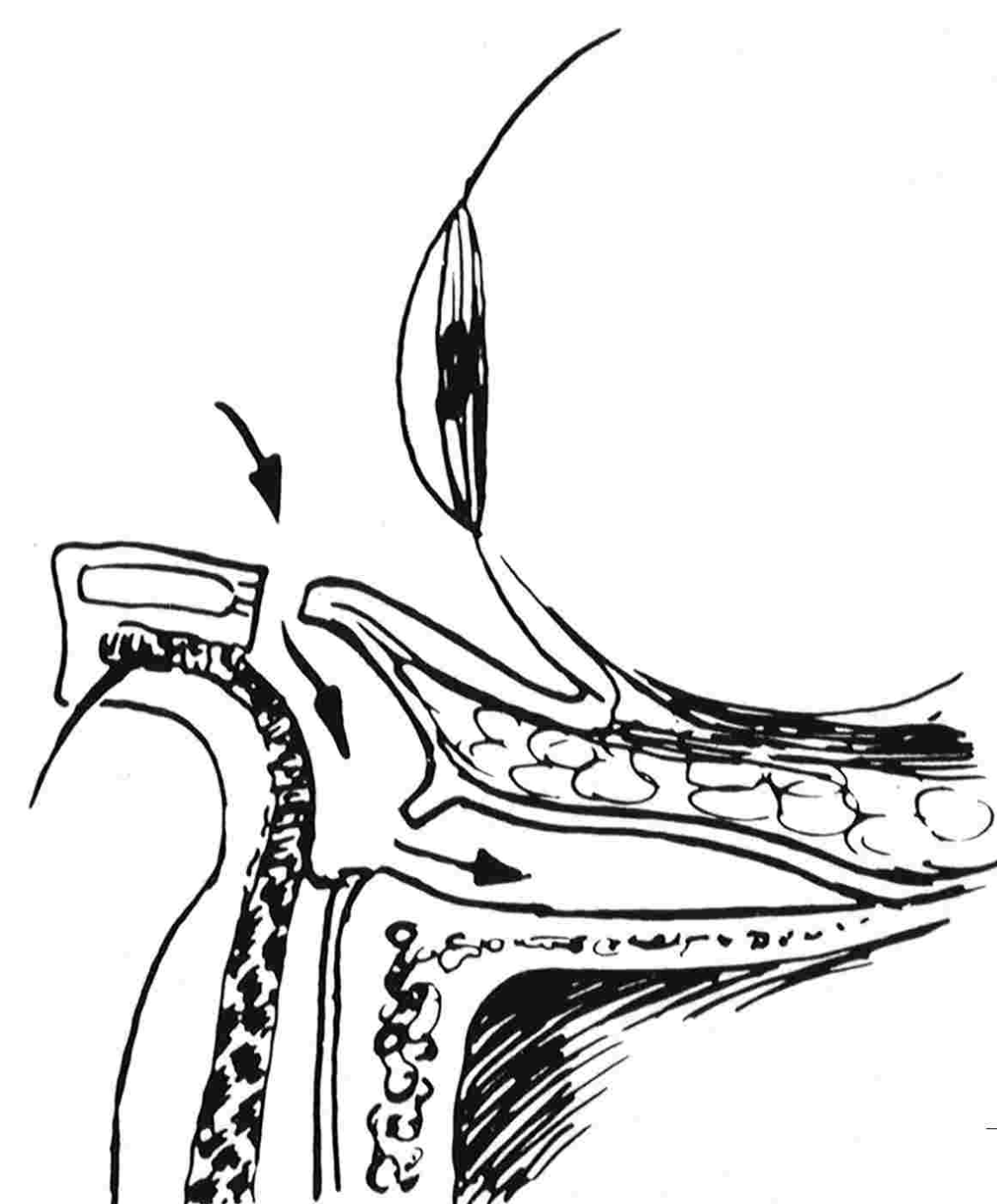


Fig. 8

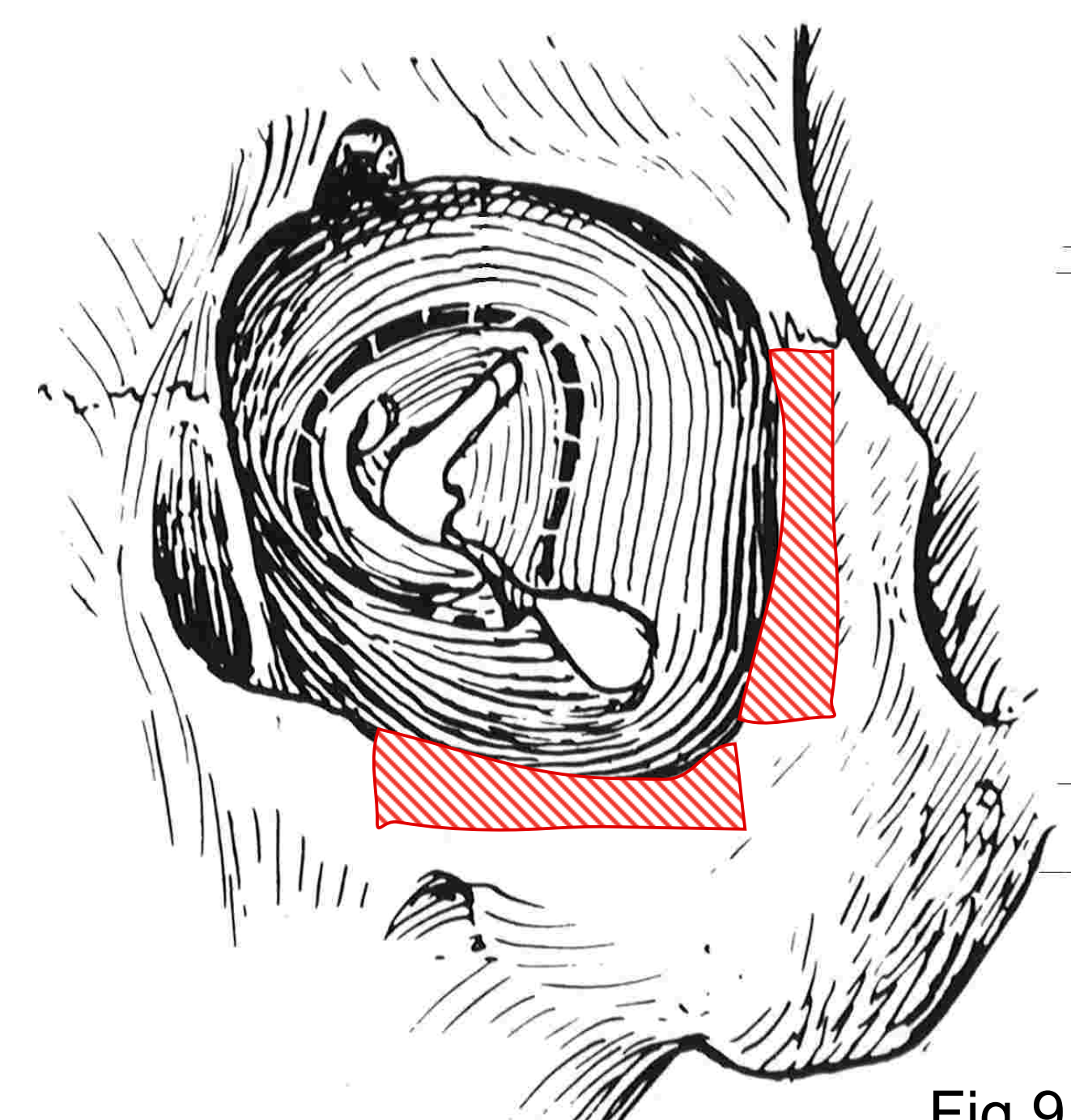


Fig 9

Craniofacial approach to the orbit is now standardized. Lesions in the orbit can be excised through hidden incisions with or without appropriate orbitotomies. The lesion such as presented in this case can be excised through a subciliary or a transconjunctival incision (Fig. 8). Occasionally improved access can be achieved by an inferior orbitotomy (Fig 9). If necessary, a lateral orbitotomy can be performed to remove a larger orbital floor lesion. If the lesion encroaches on the lateral orbital wall, this can be done through a coronal incision, thus avoiding a visible scar (Fig 10).

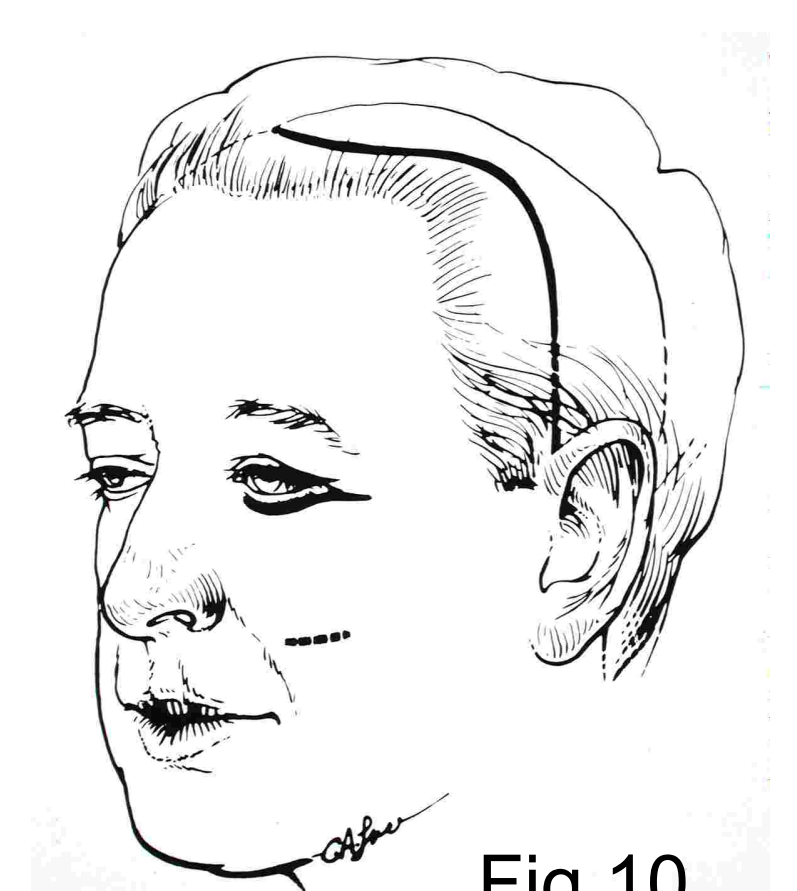


Fig 10