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CASE REPORT

Challenges in the management of massive intraorbital and hemifacial arteriovenous malformation as causing life-threatening epistaxis

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Summary Arteriovenous malformations are congenital lesions that may evolve with time and manifest in a plethora of presentations. They can occur as torrential epistaxis when it extensively involves the facial region. Multi-imaging modalities are available to assist in characterizing the structure of the lesion as well as its location and extent. This complex disease requires a multidisciplinary team approach with preoperative embolization and surgery. We present a rare cause of life-threatening epistaxis in a gentleman with a longstanding orbital and hemifacial arteriovenous malformation and discuss the complexities involved in its management.

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1. Introduction

Arteriovenous malformations (AVMs) are a subclassification of vascular malformations proposed by Mulliken and Glowacki in 1982.[1] Although vascular malformations are commonly seen in the head and neck region, the incidence of AVMs in the head and neck region is rare in comparison with hemangiomas and venous malformations.[2] Intraorbital AVMs usually present with orbital symptoms such as proptosis, diplopia, and facial deformity. Because of its anatomical proximity to the nose, epistaxis may be a presenting feature. The clinical progression of an AVM is scored from I to IV based on its severity using the Schobinger clinical staging.[2] We report a case of a stage III rare intraorbital AVM with epistaxis as a presenting feature and its treatment challenges.

2. Case report

A 35-year-old man first presented with a progressive swelling of his left eye lid for 10 years, and was treated then with embolization. His vision at that time was normal. He presented again 5 years later with a large nonpulsatile proptosis and blindness of his left eye, dilated tortuous vessels over the scalp and forehead (Fig. 1), and life-threatening epistaxis requiring repeated nasal packing. An urgent cerebral angiogram revealed an extensive left intraorbital and hemifacial AVM supplied by the ophthalmic, maxillary, facial, lingual, and superficial temporal arteries from both sides (Figs. 2 and 3) with no contrast extravasation noted. Both sphenopalatine arteries were empirically embolized with Histoacryl [*n*-butyl-2-cyanoacrylate (NBCA)] to prevent further episodes of epistaxis. Magnetic resonance imaging (MRI) showed a heterogeneously enhancing left orbital mass extensively involving the left periorbital and hemifacial area, containing dilated tortuous vessels with a "signal void" (Fig. 4). A multidisciplinary team decision resulted in preoperative embolization by the interventional radiologist using Onyx 18 (ev3, Irvine, CA, USA), a nonadhesive liquid embolic agent consisting of ethylene vinyl alcohol copolymer dissolved in dimethyl sulfoxide and micronized tantalum powder, via a sonic detachable microcatheter (Balt Extrusion, Montmorency, France), followed by radical surgery the next day.

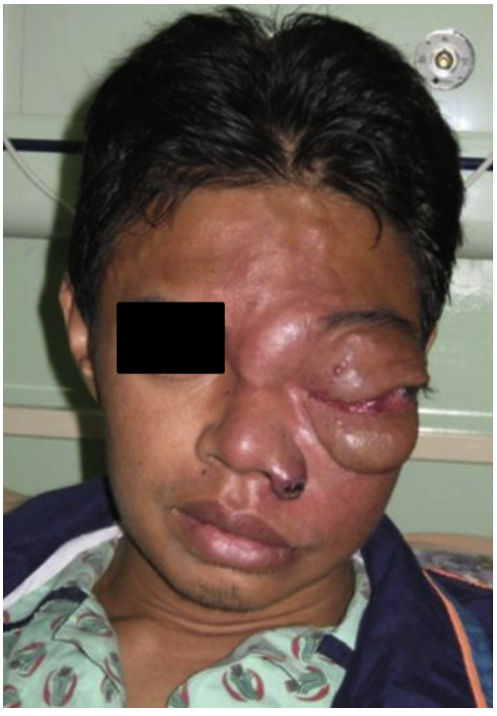

Figure 1 Large left proptosis with tortuous vessels over the forehead, extending to the nasion and involving the left ala nasi.

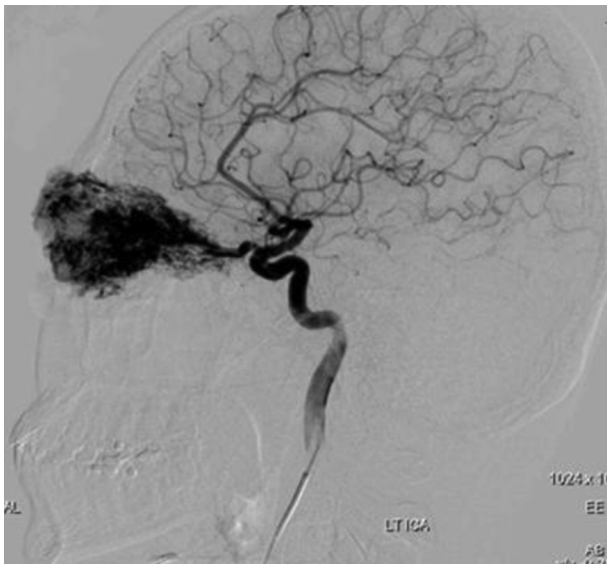

Figure 2 Internal carotid artery run showing arteriovenous malformation feeder vessels from the ophthalmic artery.

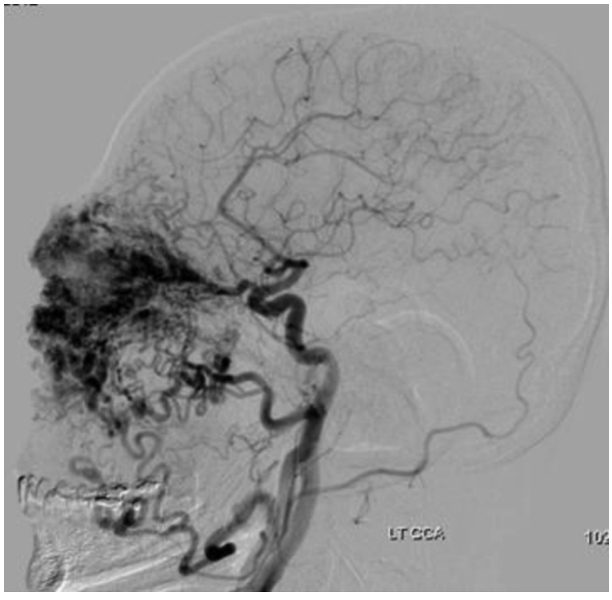

Figure 3 Left common carotid angiogram run shows extracranial arterial feeder of the AVM from the left maxillary and facial arteries branch.