

Orbital Venous Malformation Accompanied by Arteriovenous Fistula

Dear Editor,

Orbital venous malformation (OVM) is a hemodynamically low-flow vascular malformation caused by aberrant angiogenesis in the embryonic developmental stage [1,2]. It is a rare disease which accounts for 0% to 1.3% of orbital tumors, and it is usually managed conservatively [1,3]. However, when OVM is accompanied by acute thrombosis and hemorrhage or when it shows gradual enlargement, the lesion is surgically managed, as it causes intolerable pain, functional impairment, and cosmetic deficits [2,4]. Herein, we report a case of OVM fed from an arteriovenous fistula in a patient with acute proptosis and vision loss. Instead of surgical intervention on the orbital lesion, we chose to remove the feeder vessel via craniotomy, and the lesion regressed.

A 48-year-old man presented with right exophthalmos accompanied by intermittent eye pain and diplopia that had appeared 3 weeks previously (Fig. 1A, 1B). He had no history of trauma or previous diseases. Visual acuity was 18 / 20, and intraocular pressure was 14 mmHg in the right eye. A blue, vascular, cutaneous lesion was noted at the temporal area of the right face, and he reported that it had been present since he was a child (Fig. 1C, 1D). Hertel exophthalmometry revealed 12-mm proptosis on the right eye, and the globe was displaced inferiorly. Limited ocular movements in all directions were noted in his right eye. Orbital computed tomography with contrast revealed a right retrobulbar mass that simultaneously enhanced with an engorged superior ophthalmic vein (Fig. 1E).

Brain magnetic resonance angiography revealed a T1 hyperintense mass in the right retrobulbar space with internal hemorrhage and thrombi, and arteriovenous shunt-

ing from the right sphenoid bone to the lateral orbit was suspected (Fig. 1F, 1G). While awaiting cerebral angiography 2 weeks after the first visit, the patient reported a sudden loss of light perception in the right eye. Slit-lamp examination, intraocular pressure, and fundus examination were unremarkable, but the right pupil was fixed at 4 mm and had a relative afferent pupillary defect, suggesting compressive optic neuropathy. The patient was administered 500 mg of methylprednisolone intravenous twice a day. Cerebral angiography revealed a retrobulbar venous malformation that was fed from the branches of the right ophthalmic artery and recurrent meningeal artery (Fig. 1H, 1I).

On the next day, the patient underwent craniotomy via the pterional approach. The anterior clinoid process and part of the orbital roof of the sphenoid bone were removed for optic nerve decompression. Multiple epidural and intradural arteriovenous shunts were noted, and they were removed by bipolar cautery. Postoperative angiography performed 1 week after the surgery revealed no definite feeding vessels to the orbit. One year later, proptosis was completely resolved without limitation of ocular movement, although the patient had no light perception (Fig. 1J, 1K). The follow-up magnetic resonance angiography examination revealed a regressed retrobulbar venous malformation without enhancement, which confirmed occlusion of the feeding fistula (Fig. 1L, 1M).

The patient in the present report had a venous malformation in the retrobulbar space that was spontaneously combined with an arteriovenous fistula, causing severe proptosis and visual disturbance. The recent classification of orbital vascular anomalies by the International Society for the Study of Vascular Anomalies is based on the following pathological and hemodynamic characteristics: arteriovenous malformation, arteriovenous fistula, venous malformation, lymphatic malformation, and lymphaticovenous malformation [1,4]. The treatment strategy for OVM varies depending on the location and hemodynamics of the lesion, and the considerations include surgical excision, sclerotherapy using alcohol or bleomycin, embolization with a glue or coil, and laser ablation [1]. A similar case has been reported that an orbital lymphaticovenous malformation was accompanied by an arteriovenous fistula. In

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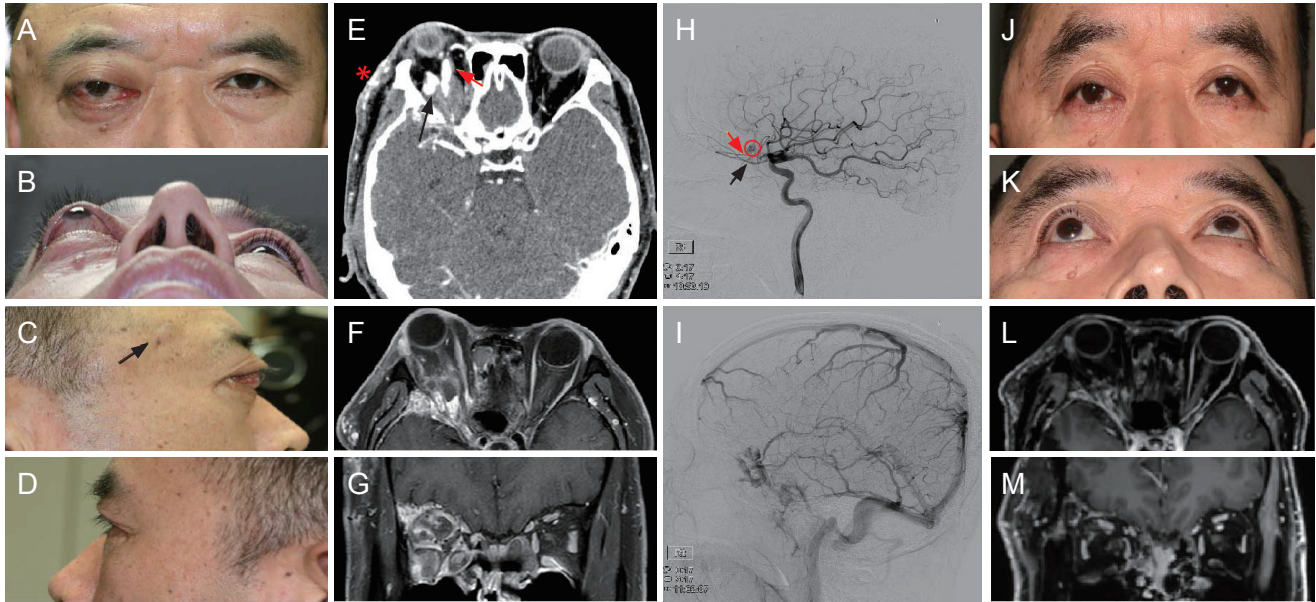


Fig. 1. Patient with orbital venous malformation accompanied by arteriovenous fistula. (A,B) Patient with severe proptosis and injection of right eye on the first day of visit. (C,D) Blue cutaneous lesion was presented at right temporal area of the face (arrow). (E) Axial view of computed tomography scan showed enhanced superior ophthalmic vein (red arrow), orbital mass (black arrow) and correlated enhancement of the cutaneous lesion (red asterisk). (F,G) Axial and coronal view of T1-weighted Gadolinium enhanced fat suppression magnetic resonance angiography respectively demonstrated mass with internal hemorrhage and thrombosis in intraconal space. (H) Preoperative cerebral angiography at arterial phase showed venous malformation (red circle) fed from ophthalmic artery (black arrow) and recurrent meningeal artery (red arrow). (I) Preoperative cerebral angiography at venous phase showed enhanced venous malformation at retrobulbar space. (J,K) After a year of the surgery, proptosis was improved. (L,M) Axial and coronal view of T1-weighted Gadolinium enhanced fat suppression magnetic resonance angiography, respectively showed decreased retrobulbar vascular structure. A written consent for the images was obtained from the patient.

that case, coil embolization and sclerotherapy were performed followed by resection [5]. The patient in the present case had a relatively well-demarcated OVM but without surgical intervention of the intraorbital lesion, it was successfully regressed after intracranial feeder vessel cauterization. Although we considered embolization, it was not feasible owing to the presence of multiple feeder vessels.

In conclusion, when an OVM is suspected based on acute symptoms and a prior computed tomography examination, angiography is essential for recognizing the underlying high-flow input, and a multidisciplinary treatment approach is needed to minimize the risk for complications and recurrence.

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Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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