Orbital Arteriovenous Fistula Causing Only Visual Impairment due to Compression of the Optic Nerve by the Dilated Superior Ophthalmic Vein

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Abstract

Orbital arteriovenous fistula (AVF) is a rare disease, and its standard therapeutic strategy has not been established. A 70-year-old male consulted an ophthalmologist due to a visual field defect in his left eye. Neurological findings showed visual impairment but no symptoms such as exophthalmos, conjunctival congestion, or diplopia. Magnetic resonance imaging showed marked dilation of the left superior ophthalmic vein (SOV). Cerebral angiography revealed an AVF that was limited to the left orbit. The feeder was a branch of the ophthalmic artery that originated from the first portion, and the drainer was the SOV, which was meandering and significantly dilated. Since the only symptom was visual impairment, the etiology was considered to be compression of the optic nerve due to a dilated SOV rather than increased venous pressure. Transvenous embolization via the facial vein was performed, and a visual field examination 1 week after the operation revealed marked improvement. Orbital AVF that develops only with visual impairment is extremely rare. As demonstrated with this case, coil embolization for proper position and reduction of the venous pressure, which relieves compression on the optic nerve, may be useful in improving the visual impairment.

Keywords

- endovascular treatment
- orbital arteriovenous fistula
- superior ophthalmic vein
- transvenous embolization
- visual impairment

Introduction

Orbital arteriovenous fistula (AVF) is extremely rare, with approximately 20 cases reported so far.[1,2] The natural course of this disease is not known in detail, and the treatment method is controversial. While proptosis and chemosis are the most common symptoms,[2] only 4 cases of visual impairment have been reported in the past.[3–6] In addition, they also have other symptoms such as proptosis or chemosis. Herein, we report a case of an orbital AVF that develops only with visual impairment without proptosis or chemosis which is rare in that no similar reports have been reported in the past.

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Case Description

History

A 70-year-old man with no history of trauma was aware of the deterioration of his visual acuity 6 months before his visit. A close examination by an ophthalmologist revealed a decrease in his left visual acuity of 0.3, visual field impairment (►**Fig. 1A**), and no increase in intraocular pressure nor capillary dilation of the ocular conjunctiva. Also, there was no difference between left and right in the central retinal arteries and veins. The neurological findings at the time of admission to our hospital showed left visual impairment, but no chemosis, proptosis, or eye movement disorder were observed. Magnetic resonance (MR) angiography demonstrated flow void anterior to the left ophthalmic artery (►**Fig. 2A**) and T2-weighted image of MR imaging (T2WI) showed dilation of the left superior ophthalmic vein (SOV, ►**Fig. 2B**), suggesting an arteriovenous shunt and cerebral angiography was performed for further examination. Left internal carotid angiography revealed a shunt between the ophthalmic artery and SOV (►**Fig. 2C**) while there was no feeding from the external carotid artery system. Furthermore, after the SOV meandered markedly (*arrowhead*) according to the three-dimensional digital subtraction angiography (3D-DSA), it flowed out to the angular vein and facial vein (►**Fig. 2D**). No obvious cortical reflux or venous stasis was observed in the venous phase of cerebrovascular angiography (►**Fig. 2E**). Based on the above findings, a diagnosis of orbital AVF was made, and the cause of the visual impairment was considered to be compression of the optic nerve by dilated SOV rather than increased venous pressure. After admission, visual impairment gradually progressed, so transarterial embolization was performed for the purpose of improvement of visual impairment.

Endovascular procedure was performed under local anesthesia. A 4-French diagnostic catheter (Medikit co. ltd., Tokyo, Japan) was guided through the left femoral artery to the left internal carotid artery and roadmap images and control runs were obtained during the procedure. A 7-French guide catheter (Asahi Intec co. ltd., Aichi, Japan) was navigated through the right femoral vein to the left internal jugular vein. The microcatheter (Terumo MicroVention, Tustin, California, United States) was set up coaxially with the 3.4-Fr distal access catheter (Technocrat co., Aichi, Japan) and was guided to the immediate distal to the shunt point through the internal jugular vein, facial vein, angular vein, and SOV (►**Fig. 3A**). The outflow side of the shunt was obliterated by placing a total of six removable coils (►**Fig. 3B,C**) and the shunt blood flow had disappeared after embolization (►**Fig. 3D**).

Visual acuity did not change soon after the operation, but a marked improvement in visual acuity, from 0.3 to 0.9, was observed on the seventh postoperative day. The SOV was thin on the T2WI, and the flow void disappeared on MR angiography 3 months after the procedure (►**Fig. 3E,F**). The visual field at 5 months after the operation also significantly improved compared to the preoperative examination (►**Fig. 1A,B**).

Discussion

Orbital AVF is a very rare disease, and only about 20 cases have been reported in the past.[1–4,6–19] Asymptomatic patients are often just followed up conservatively because it is a disease that may disappear spontaneously.[9,20] On the other hand, some patients have symptoms such as chemosis, proptosis, double vision, and visual impairment. Visual impairment is a rare symptom and only a few cases have been reported in the past, and all of them have other ocular symptoms simultaneously.[3–6] In the above cases, visual impairment was improved by endovascular or surgical intervention. This case is an orbital AVF that develops only with visual impairment and is rare in that no similar reports have been reported in the past.

In general, the symptoms of orbital AVF are presumed to differ depending on the pattern of venous drainage. Increased pressure of ophthalmic veins causes conjunctival hyperemia, chemosis, exophthalmos, and secondary glaucoma due to

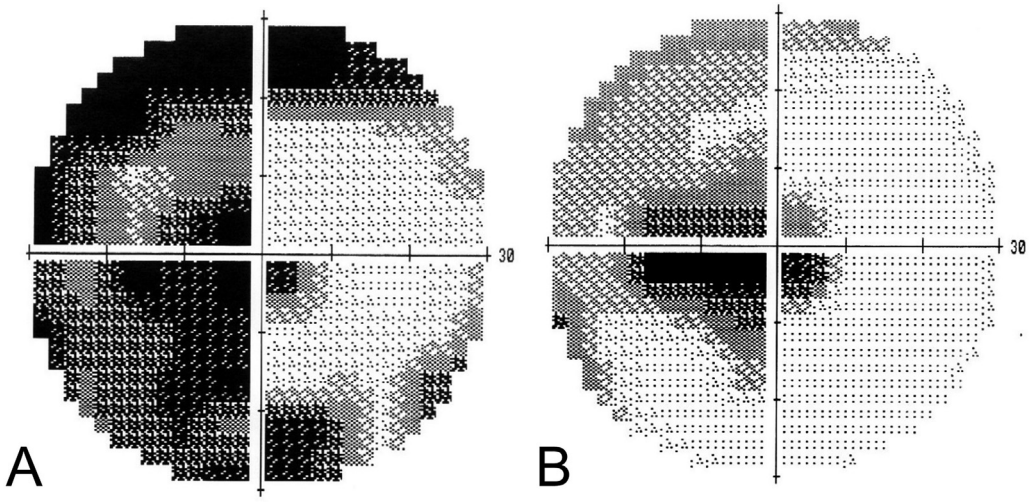

Fig. 1 Pre- and postoperative visual field examination of the left eye. The preoperative examination showed left side hemianopsia (**A**), which had significantly improved in the postoperative examination (**B**).

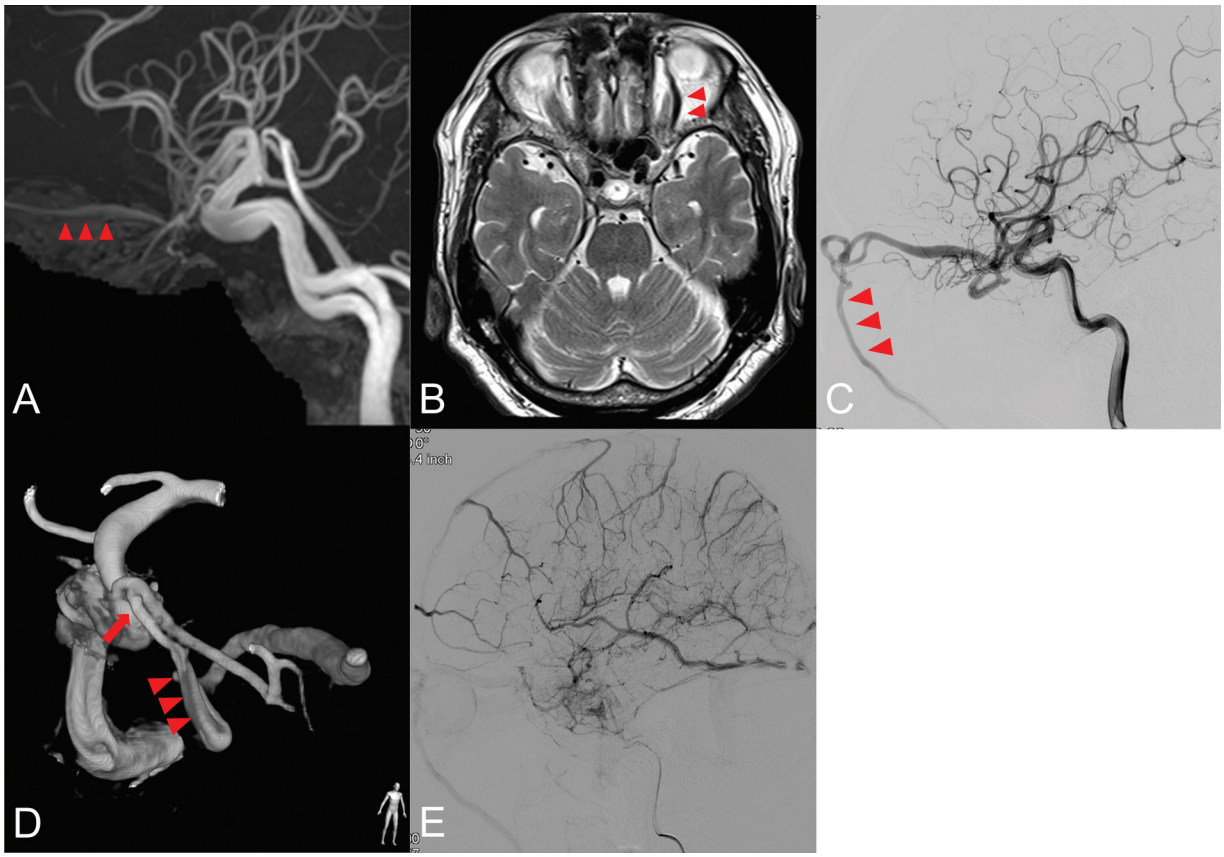

Fig. 2 Preoperative imaging. (**A**) Flow void in the orbit (*arrowheads*) is visualized in magnetic resonance (MR) angiography. (**B**) Axial T2-weighted MR image showing marked dilation of the left superior ophthalmic vein (SOV) (*arrowheads*). (**C**) Left internal carotid angiography revealed an arteriovenous shunt between the ophthalmic artery and SOV, which flowed out to the angular and facial veins (*arrowheads*). (**D**) Three-dimensional digital subtraction angiography (3D-DSA) showed significant meandering of the SOV (arrowheads) distal to the shunt point (*arrow*). (**E**) No obvious cortical reflux or venous stasis was observed in the venous phase of cerebrovascular angiography.

increased intraocular pressure. On the other hand, increased pressure of cavernous sinus causes extraocular muscle paralysis, ptosis, and anisocoria. In past case reports, symptoms corresponding to the pattern of venous return have appeared. In contrast to previous reports, this case is extremely rare in that it causes only visual impairment. In this case, the intraocular pressure was normal and there was no difference between left and right in the central retinal arteries and veins. These findings indicate there was no abnormality in venous reflux which is possibly due to significant development of drainage to the facial vein via the angular vein. On the other hand, markedly meandering SOV physically pressed the optic nerve, causing visual impairment. In other words, symptoms appear not due to increased venous pressure but due to the mechanical compression to the optic nerve.

Treatment strategies for orbital AVF include surgical treatment, transarterial embolization, and TVE when the patient experiences progressive symptoms. Surgical treatment that exposes and treats the SOV is extremely invasive as is reported in the past.[5,12] Transarterial embolization from the ophthalmic artery, which is the most common feeder, proximal to the origin of central retinal artery may cause loss of vision.[21] Transarterial embolization is a treatment that can overcome the weaknesses of the above treatment policy, although it may be difficult to guide the catheter due to meandering of the vein. In fact, most of the past reports have been treated by transarterial embolization only, which followed good outcomes.[1,2,4,6,7,10,14,16,17] The most important risk of transarterial embolization is a hemorrhage associated with venous congestion by blocking the outflow. Originally, the SOV has blood flow from the orbit toward the cavernous sinus in normal condition. Therefore, embolization of the SOV may cause venous congestion of the central retinal vein and its branches, resulting in fundal hemorrhage. Preoperative fundus examination revealed no dilation of the central retinal vein or its branches so it was presumed that these veins flow into the cavernous sinus by a route unrelated to the SOV. Embryologically, orbital venous drainage and cavernous sinus form one continuous channel, but gradually differentiate into multiple channels.[17] In other words, even if the SOV is not involved in orbital venous drainage, the return from other veins is maintained. In this case, 3D-DSA revealed meandering of the proximal part of SOV which compressed the optic nerve from above. The treatment strategy was to reduce the pressure of the SOV and release the pressure on the optic nerve by embolizing the fistula. In order to achieve this goal, it is essential to embolize the coil at the appropriate site. In detail, the proximal portion of the lesion is the optic

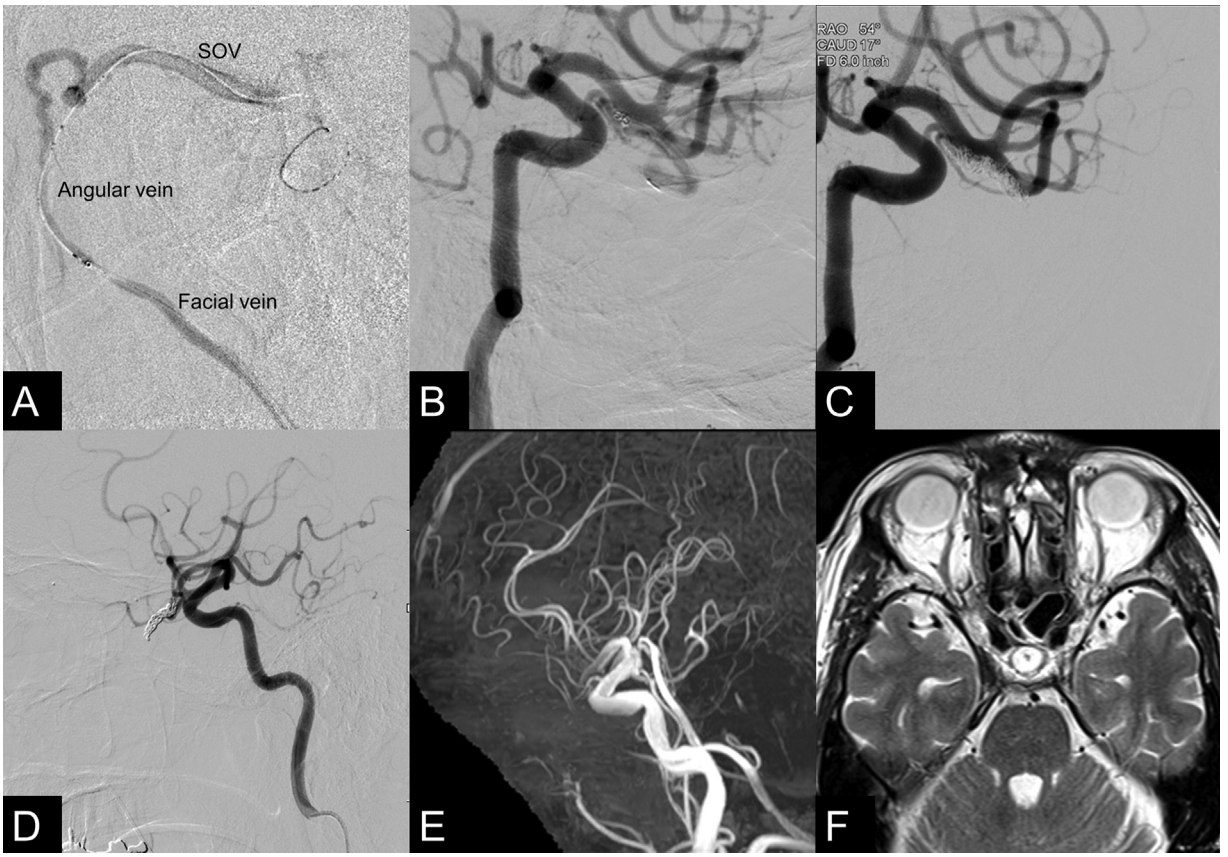

Fig. 3 Intra- and postoperative imaging. (**A**–**D**) Lateral (**A**,**D**) and right anterior oblique view (**B**,**C**) of the intraoperative cerebral angiography. A microcatheter was placed just distal to the shunt point via the facial, angular, and superior ophthalmic veins (SOV) (**A**). Coil embolization was started just distal to the shunt point (**B**), and a total of six coils was deployed (**C**). The shunt blood flow had disappeared after embolization (**D**). (**E**,**F**) Postoperative imaging at 3 months after the procedure. The flow void had disappeared on magnetic resonance (MR) angiography (**E**), and the SOV was thin on the coronal T2-weighted MR image (**F**).

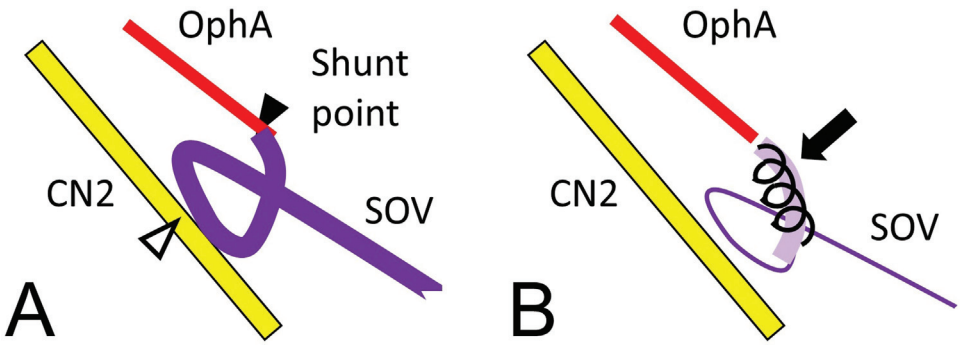

Fig. 4 Schema of this case. It was essential to embolize the coil at an appropriate site just distal to the shunt point (**A**, *black arrowhead*). In this case, the proximal portion of the lesion is the optic canal, in which runs a number of important nerves, arteries, and veins in a very narrow space, and the distal portion is the part where the superior ophthalmic vein (SOV) squeezes the optic nerve (**A**, *white arrowhead*). The coil was deployed in the SOV, just distal to the shunt point (**B**, *arrow*). The meandering SOV shrank, and the pressure on the optic nerve was released.

canal, which runs optic nerve, arteries, and veins in a very narrow space and the distal portion is the part where the SOV squeezes the optic nerve. By packing coils in those areas, the optic nerve is unintentionally compressed, which may worsen the symptoms. By performing necessary and sufficient embolization on the planned site via the facial vein, we succeeded in improving the symptoms (►**Fig. 4**).

Conclusion

Orbital AVF is a very rare disease and which causes only visual impairment has not been reported in the past. The etiology of this case is nerve compression by dilated meandering SOV. There are various nerves and blood vessels around the shunt point, and it is necessary to evaluate the pathological condition

in detail before treatment. In the case of this mechanism, coil embolization for proper position and reduction of venous pressure which relieves compression on the optic nerve may be useful in improving visual impairment.

Informed Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of Interest

None declared.

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