Unilateral isolated superior ophthalmic vein thrombosis

Raksha Rao, Yasmin Ali[1], Chinmay P Nagesh[2], Unnikrishnan Nair[1]

Superior ophthalmic vein (SOV) thrombosis is an uncommon orbital pathology that can present with sudden onset proptosis, conjunctival injection, and visual disturbance. SOV thrombosis is frequently secondary to a cavernous sinus pathology. A 32-year-old female with a known history of autoimmune hemolytic anemia presented with sudden painful proptosis left eye, and on imaging, she was found to have SOV thrombosis without cavernous sinus involvement. She was diagnosed with unilateral isolated SOV thrombosis and was managed conservatively. A careful history and clinical evaluation can help diagnose such rare disorders and initiate appropriate therapy.

Key words: Autoimmune hemolytic anemia, isolated, orbit, proptosis, superior ophthalmic vein, thrombosis

Isolated superior ophthalmic vein (SOV) thrombosis is an extremely rare entity.[1-4] SOV thrombosis (SOVT) is generally secondary to orbital inflammations and infections, tumoral compression or infiltration, and direct or indirect carotid cavernous fistula (CCF).[1-4] It may present with a sudden onset of painful proptosis, chemosis, conjunctival congestion, and visual disturbance and can be detected using computed tomography (CT) or magnetic resonance (MR) angiography.[1,3] Early diagnosis and appropriate management strategies must be based on the underlying causative factors to prevent visual loss and other complications. Herein, we present a case of isolated unilateral SOVT secondary to autoimmune hemolytic anemia (AIHA).

Case Report

A 32-year-old female presented with a sudden onset of redness in the left eye for 2 weeks. There was also a history of forward protrusion of the same eye with periorbital ache. She had been previously diagnosed with AIHA, for which she was undergoing regular blood transfusions for 15 years. There was no other significant medical, surgical, or family history.

On examination, the best-corrected visual acuity in both eyes was 20/20. Intraocular pressure in the right eye was 12 mmHg and in the left eye 14 mmHg. Anterior segment of the right eye was unremarkable. In the left eye, there was periorbital ecchymosis, diffuse conjunctival congestion, subconjunctival hemorrhage, and episcleral vascular engorgement and tortuosity [Fig. 1a and b]. On Hertel's exophthalmometry, a 4 mm axial proptosis left eye was noted [Fig. 1c]. There was no palpable thrill or bruit. Pupils were briskly reactive, and ocular movements were full. Gonioscopy showed open angles in both eyes. Fundus examination showed bilateral disc edema with multiple scattered intraretinal and subretinal hemorrhages. A dual phase CT angiography of the brain and orbits showed that the cavernous sinuses were symmetrical in appearance with no early arterial phase opacification of either the cavernous sinuses or the SOVs, with a clear-cut filling defect in the enlarged left SOV in the venous phase [Fig. 1d-f]. This was considered diagnostic of a thrombosis isolated to the left SOV alone. In addition, enlargement of the extraocular muscles in the left orbit was also noted. In view of the lack of involvement of the cavernous sinus, a diagnosis of unilateral isolated SOVT was made.

A neurology and hematology opinion was advised. Lumbar puncture revealed normal opening cerebrospinal fluid pressure, and MR imaging revealed normal brain study, hence ruling out papilledema. Hematological workup revealed chronic anemia with anticardiolipin antibody positivity. She was negative for rheumatoid factor, protein-C, protein-S, antithrombin-III, elevated homocysteine level, antiphospholipid antibodies, antinuclear antibody, ds-DNA, antineutrophil cytoplasmic antibody, and factor-V-Leiden mutation. She was treated with intravenous methylprednisolone (IVMP) 1 g for 3 days, followed by a tapering dose of oral steroids. Gradual resolution of symptoms was noted, and the vision was preserved at 20/20 both eyes at 8-week follow-up [Fig. 1g and h].

Discussion

Although SOVT in association with cavernous sinus thrombosis is known to occur, isolated SOVT (SOVT in the absence of cavernous sinus pathology) is an extremely rare entity.[1,3] Risk factors implicated in the development of SOVT contribute to at least one of the elements comprising the Virchow's triad (vascular damage, stasis, and hypercoagulability).[1] The etiology of SOVT can be broadly categorized as septic and aseptic.[1-5] Septic causes include infectious diseases such as

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_791_17

Department of Orbit, Oculoplasty and Ocular Oncology, [1]Department of Retina, Chaithanya Eye Hospital and Research Institute, [2]Department of Imaging Sciences and Interventional Radiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum, Kerala, India

Correspondence to: Dr. Raksha Rao, Orbit, Oculoplasty and Ocular Oncology Service, Chaithanya Eye Hospital and Research Institute, Kesavadasapuram Junction, Trivandrum - 695 004, Kerala, India. E-mail: raoraksha@gmail.com

Manuscript received: 25.08.17; Revision accepted: 25.10.17

Cite this article as: Rao R, Ali Y, Nagesh CP, Nair U. Unilateral isolated superior ophthalmic vein thrombosis. Indian J Ophthalmol 2018;66:155-7.

orbital cellulitis, paranasal sinusitis, and septic cavernous sinus thrombosis with the extension of the thrombus into the SOV, whereas aseptic causes include spontaneous thrombosis of the dural cavernous fistula, tumors of the cavernous sinus or the orbit, and Tolosa–Hunt syndrome.[1-3]

Apart from orbital diseases, certain systemic factors can be attributed to the development of SOVT. These include hypercoagulable states (use of oral contraceptive pills, pregnancy, thrombocytosis, sickle cell trait, and hereditary hemorrhagic telangiectasia), inflammatory diseases (Graves' disease, systemic lupus erythematosus, sarcoidosis, Behçet syndrome, and amyloidosis), systemic malignancies (leukemia, lymphoma, and multiple myeloma), and amyloidosis.[1-4] While orbital pathologies commonly lead to unilateral SOVT, the systemic conditions can cause unilateral or bilateral involvement. Thrombotic events in young patients are frequently associated with potentially life-threatening systemic conditions, necessitating a detailed assessment to diagnose the underlying disorder.[6] In our case, the patient was only 32 years old with a history of AIHA.

AIHA is an autoimmune disorder, in which the autoantibodies target the surface antigens of the red blood cells (RBCs), causing peripheral destruction of RBCs leading to anemia.[5-7] It is an uncommon systemic condition, with a low incidence of 1–3 persons in 100,000 annually.[5-7] In a large meta-analysis, it was demonstrated that venous thromboembolism in patients with AIHA is 2.6 times higher in comparison to patients without AIHA.[5] The mechanism behind the thrombotic events is suggested to be a hyperactivated coagulation system.[7] Despite this, the use of antithrombotic prophylaxis in AIHA is debatable.[7]

Although AIHA causing venous and arterial thromboembolism is a known entity, to the best of our knowledge, SOVT in association with AIHA has not been reported till date. The presence of anticardiolipin antibodies, a frequent occurrence in AIHA, caused a higher risk of thrombosis in our case.[8,9] SOVT led to proptosis and dilated episcleral veins, but without secondary glaucoma, probably due to the short course of the condition. The bilateral disc edema seen in our patient is possibly secondary to either optic nerve ischemia or raised venous pressure without actual venous thrombosis, entities that have been described in chronic anemia.[10]

Contrast-enhanced MR venography or dual-phase CT angiography helps confirm the diagnosis of isolated SOVT by ruling out shunting lesions such as CCF, thrombus extension from the cavernous sinus, and sino-orbital infections.[1,3] An MR venography is not only more sensitive than CT venography but maybe more superior in demonstrating patency of the cavernous sinuses that may otherwise be obscured by artifacts from the adjacent bones on the latter.[1,3] Additional sequences such as diffusion-weighted imaging may also be useful when contrast medium administration is contraindicated.[11] Oral antibiotics may benefit patients with SOVT secondary to septic causes.[4] IVMP given in patients presenting with SOVT aids in symptomatic improvement by relieving orbital congestion and reducing proptosis.[1,2] Although some authors recommend the use of intravenous heparin or oral aspirin, their role in SOVT is yet to established.[1-3,6]

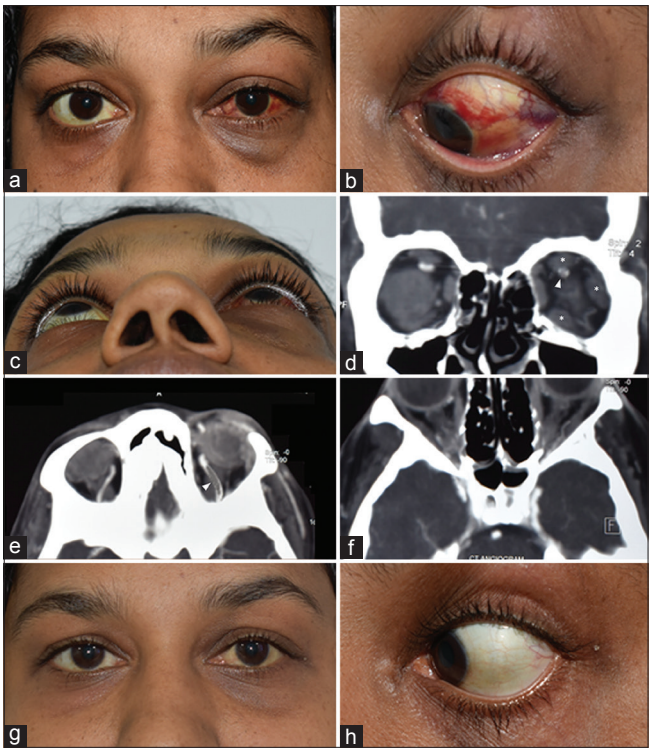

Figure 1: (a and b) External photographs illustrate left periorbital ecchymosis, subconjunctival hemorrhage, and dilated and tortuous episcleral veins; (c) external photograph with worm's eye view reveals left eye proptosis; (d) coronal reconstruction; and (e) axial section of the venous phase of computed tomography angiography shows dilated left superior ophthalmic vein thrombosis (white arrowhead) and enlarged extraocular muscles (*); (f) a more inferior axial section illustrates a normally opacified symmetric cavernous sinuses without any filling defect to suggest thrombosis. (g and h) External photographs at 8-week follow-up

Conclusion

Isolated SOVT is an infrequent diagnosis, and the more common differential diagnosis of cavernous sinus pathology must be ruled out. SOVT must be considered in all cases of sudden onset of painful proptosis, and a careful evaluation to find the underlying cause must be done to prevent life-threatening thromboembolic events.

Declaration of patient 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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Lim LH, Scawn RL, Whipple KM, Oh SR, Lucarelli MJ, Korn BS, *et al.* Spontaneous superior ophthalmic vein thrombosis: A rare

entity with potentially devastating consequences. Eye (Lond) 2014;28:348-51.

2. Mandić JJ, Mandić K, Mrazovac D. Superior ophthalmic vein thrombosis with complete loss of vision as a complication of autoimmune and infective conditions. Ocul Immunol Inflamm 2017;24:1-3.
3. Cumurcu T, Demirel S, Keser S, Bulut T, Cavdar M, Dogan M, *et al*. Superior ophthalmic vein thrombosis developed after orbital cellulitis. Semin Ophthalmol 2013;28:58-60.
4. Michaelides M, Aclimandos W. Bilateral superior ophthalmic vein thrombosis in a young woman. Acta Ophthalmol Scand 2003;81:88-90.
5. Ungprasert P, Tanratana P, Srivali N. Autoimmune hemolytic anemia and venous thromboembolism: A systematic review and meta-analysis. Thromb Res 2015;136:1013-7.
6. Hendrick AM. Auto-immune haemolytic anaemia – A high-risk disorder for thromboembolism? Hematology 2003;8:53-6.
7. Ruggeri M, Rodeghiero F. Thrombotic risk in patients with immune haemolytic anaemia. Br J Haematol 2016;172:144-6.
8. Lang B, Straub RH, Weber S, Röther E, Fleck M, Peter HH, *et al.* Elevated anticardiolipin antibodies in autoimmune haemolytic anaemia irrespective of underlying systemic lupus erythematosus. Lupus 1997;6:652-5.
9. Bick RL, Baker WF. Anticardiolipin antibodies and thrombosis. Hematol Oncol Clin North Am 1992;6:1287-99.
10. Biousse V, Rucker JC, Vignal C, Crassard I, Katz BJ, Newman NJ, *et al.* Anemia and papilledema. Am J Ophthalmol 2003;135:437-46.
11. Pendharkar HS, Gupta AK, Bodhey N, Nair M. Diffusion restriction in thrombosed superior ophthalmic veins: Two cases of diverse etiology and literature review. J Radiol Case Rep 2011;5:8-16.