Smoldering multiple myeloma revealed by superior ophthalmic vein thrombosis

Nadia Bouattour, Nouha Farhat, Nouha Hamza, Hanen Hadjkacem, Olfa Hdiji, Salma Sakka, Mariem Dammak, Chokri Mhiri

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Abstract:

Superior ophthalmic vein thrombosis is a rare entity. It is associated with significant morbidities. It may present with dramatic clinical signs. It is frequently secondary to cavernous sinus pathology and it can be a harbinger of cavernous sinus thrombosis. We report an unusual case of superior ophthalmic vein thrombosis, as the first manifestation of multiple myeloma. As far as we know, this is the first case described in the literature. Here we describe a patient presented with a painful, visual blur and a right-sided proptosis due to superior ophthalmic vein thrombosis. Appropriate medical workup was conducted, and smoldering multiple myeloma was diagnosed as the underlying cause. We further discuss the possible involved mechanisms.

Keywords:

Hyperviscosity syndrome, multiple myeloma, ophthalmoplegia, proptosis, superior ophthalmic vein

Introduction

Superior ophthalmic vein thrombosis (SOVT) is an uncommon orbital pathology that can present with dramatic clinical signs: a sudden onset proptosis, a conjunctival injection and a loss of visual function. SOVT can have various causes such as an orbital infection, tumors, traumatic or spontaneous carotid cavernous fistulas or vascular and coagulation anomalies.[1-4] We report an unusual case of SOVT due to hyperviscosity syndrome, as the first manifestation of multiple myeloma. SOVT can be detected with contrast-enhanced computed tomography (CT) or a magnetic resonance imaging (MRI). Appropriate intervention and management strategies based on the underlying disease and the patient's clinical symptoms are required, otherwise severe complications can occur.[3]

Case Report

A 70-year-old man with a history of surgery of right-sided cataract, 2 years ago, presented to the emergency department with an ongoing headache for 2 weeks with a painful and progressive right-sided proptosis, associated to a visual blur, redness, and periorbital edema. He had neither a medical history of previous venous thromboembolism nor a known malignancy or a coagulopathy.

Ocular examination revealed right periorbital swelling with mild erythema. There was no palpable thrill or bruit heard in the affected globe, but it was proptosed by 6 mm. A significant conjunctival chemosis with dilated episcleral vessels was noted. The ocular motility examination showed a total right ophthalmoplegia [Figure 1].

The left eye examination was normal except for senile immature cataract. The Oropharynx examination was normal and especially nasal swabs and culture ruled out bacterial and fungal sinusitis. Suspecting an intraocular malignancy with orbital involvement, an urgent CT scan of the orbits was performed [Figure 2]. This showed a dilated and thrombosed superior ophthalmic vein with an associated swelling of the extraocular muscles. CT and MRI angiograms excluded a dural cavernous sinus fistula, a sino-orbital infection, but showed a partial extension of thrombus into the ipsilateral cavernous sinus [Figure 3]. Plasma

Department of Neurology's, Habib Bourguiba Hospital, Sfax, Tunisia

Address for correspondence:
Nadia Bouattour, Department of Neurology's, Habib Bourguiba Hospital, Ferdaous Street, Sfax 3029, Tunisia.
E-mail: bouattour_nadia@yahoo.fr

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protein electrophoresis showed paraprotein and increased gamma-globulins, whereas Bence-Jones proteins were not detected on urinary protein electrophoresis. Blood counts and investigations were as follows: Hemoglobin 14.5 g/dl, white blood cells 10000/µL, platelets 355000/µL, random sugar levels, renal function, and phosphocalcic balance were normal. Radiographs of the flat bones were normal. Bone marrow biopsy showed atypical plasma cells infiltration equal to 11%. On the basis of these findings, CRAB's criteria were absent and the diagnosis of smoldering (asymptomatic) IgG multiple myeloma was made.[5] As previously mentioned, smoldering multiple myeloma does not require treatment, so the patient, in this case, was not treated by chemotherapy.[5] He underwent IV heparin and bridged to the oral anticoagulant. Evolution was marked ultimately by the gradual disappearance of symptoms 1 week after starting anticoagulation therapy [Figure 4]. Close monitoring was indicated every 4 months including renal function, serum calcium, total blood count, serum protein electrophoresis with immunofixation, 24-hour

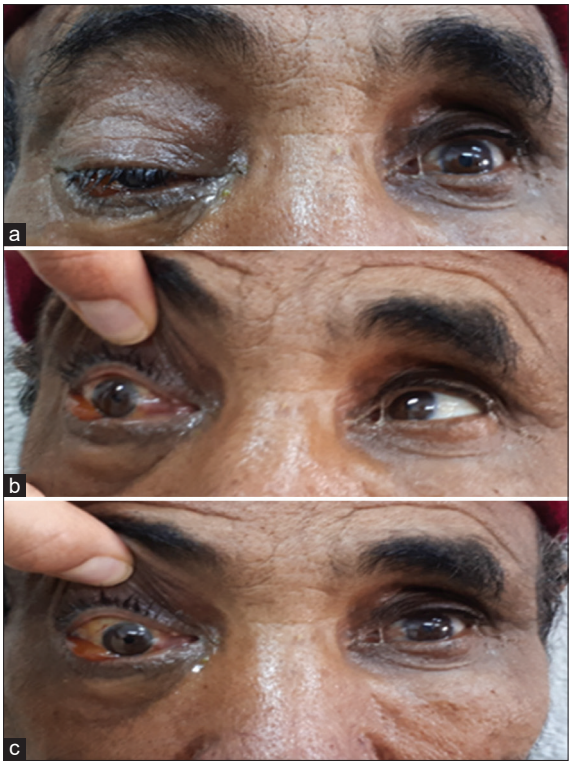

Figure 1: A photograph showing: (a) Right periorbital swelling with mild erythema, ecchymosis, subconjunctival hemorrhage, dilated and tortuous episcleral veins; (b and c) showed a restricted ocular motility

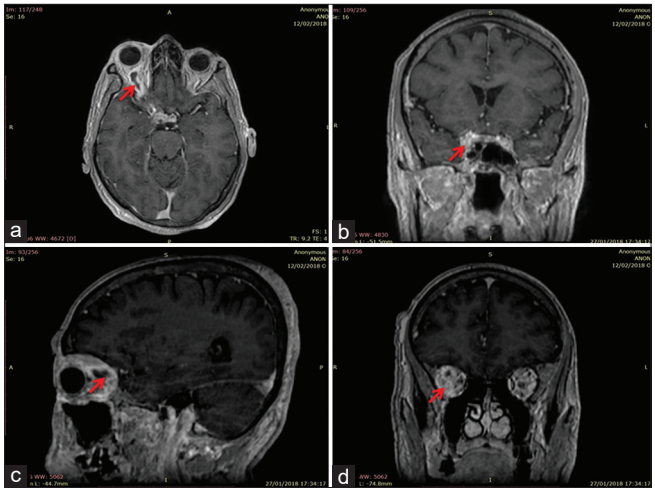

Figure 3: Brain MRI (T1 Gadolinium-enhanced): (a) axial section, (b and d), coronal section, (c) sagittal section: the arrow is pointing to right SOVT, (b) T1 coronal section: the arrow is pointing to the partial extension of thrombus into the right cavernous sinus

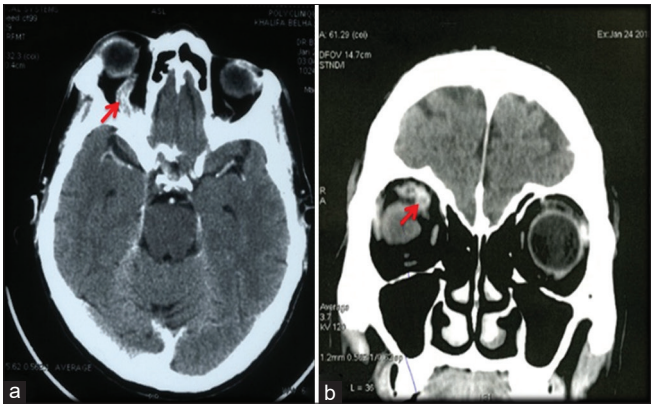

Figure 2: Computed Tomography Scan of the orbits: (a) axial section, (b) coronal reconstruction of the venous phase of computed tomography angiography: The arrow is pointing to dilated right superior ophthalmic vein thrombosis and enlarged extraocular muscles

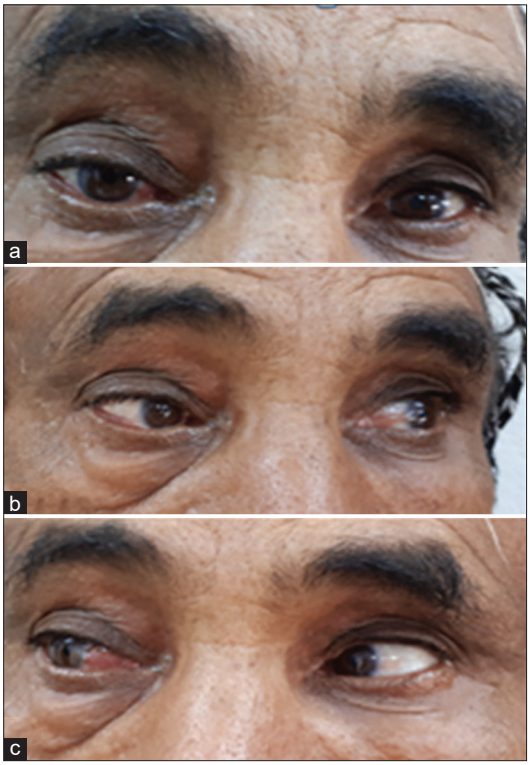

Figure 4: External photographs at 1-week follow-up; (b and c): no limitation of ocular motility

Table 1: Review of the literature

Publications	Number of cases	Age (years)	Etiologies	Treatment	Evolution
Schmitt NJ (2005)[19]	1	71	Dacryocystitis-induced orbital cellulitis	Intravenous vancomycin Dacryocystorhinostomy Postoperative intravenous dexamethasone	Favorable after 2 weeks
Carrim ZI (2006)[20]	1	78	Idiopathic orbital inflammatory disease	Oral steroid and anticoagulants	Favorable after 8 weeks
Iseki S (2010)[21]	1	77	Traumatic carotid-cavernous sinus fistula	Surgery	Favorable
Shinder R (2010)[22]	1	-	Hypercoagulability due to Chronic Myeloid Leukemia	Anticoagulant therapies	Unspecified
Vyas S (2011)[1]	1	45	Cavernous sinus meningioma with ipsilateral superior and inferior vein thrombosis	Surgery	Unspecified
Wade RG (2013)[23]	1	84	Idiopathic orbital inflammation	Anticoagulant therapies	Favorable
Lim L (2013)[18]	3	77 58 42	Hereditary Hemorrhagic Telangiectasia (HHT) Coagulopathy Systemic malignancy	Anticoagulant therapies	Favorable
Mishima M (2015)[7]	1	77	Severe facial trauma	Surgery	Favorable
Baidoun F (2015)[24]	1	60	Nephrotic syndrome from Class IV Lupus Nephritis	Anticoagulant	Favorable
Sambhav K (2015)[25]	1	68	Systemic lupus erythematosus bilateral consecutive superior ophthalmic vein thrombosis	Oral steroids/anticoagulant	Favorable
Pawar B (2017)[26]	1	71	Orbital inflammation	Oral steroids/anticoagulant	Lost to follow-up
Rao R (2018)[27]	1	32	AutoImmune Hemolytic Anemia (AIHA)	Intravenous steroid/oral steroids	Favorable after 8 weeks
Our case	1	70	Smoldering multiple myeloma	Anticoagulant	Favorable after a week

urine protein electrophoresis with immunofixation and skeletal X-ray survey.

Discussion

SOVT is an extremely rare entity resulting from orbital congestion, such as that caused by infectious diseases, cavernous sinus thrombosis, skull base tumors, arteriovenous malformations, after facial trauma and in a patient with hypercoagulability.[1-3,6,7] It has been reported to occur in a wide range of diseases, as summarized in Table 1.

Multiple myeloma is a frequent cause of the hyperviscosity syndrome.[8] Venous thromboembolism is highest during the first four months following the initial diagnosis.[9] SOVT occurred in our patient as a result of hyperviscosity because of an increased amount of circulating IgG paraprotein and cellular blood components.[10-12] Ocular manifestations can be the first signs of a patient developing multiple myeloma,[13] but SOVT wasn't reported in the literature. Multiple myeloma can be active (symptomatic) or smoldering (asymptomatic). Therapy should be started immediately for symptomatic disease, whereas asymptomatic disease requires only close monitoring.[14,15] As in the case of our patient.

Patients with SOVT may present with an orbital pain, diplopia or a decreased vision. Clinical findings may include proptosis, chemosis, ophthalmoplegia, and ptosis. The optic nerve may be affected by compression.[16]

SOVT is usually identified by contrast-enhanced CT or MRI followed by an assessment of the patient's clinical symptoms.[17] In our patient, the brain scanner showed engorgement and thrombosis of the SOV. The normal maximal diameter of the SOV was reported to be 3.5 mm.[2] In view of the possible systemic associations of orbital inflammation, inflammatory and autoimmune workup may be indicated. A chest x-ray is indicated to evaluate for sarcoidosis.[18] The optimal treatment for SOVT depends on the etiology and clinical symptoms of the condition [Table 1]. We treated our patient with heparin followed by anticoagulant therapy. Regardless of etiology of SOVT, if there is no contraindication, anticoagulant therapy should be initiated by the clinician. According to the literature evolution under treatment was generally favorable [Table 1].

In conclusion, we described the first case report of SOVT as the first manifestation of multiple myeloma. Cases with SOVT require interdisciplinary collaboration and more detailed and extensive investigations to determine the underlying pathology and to achieve the best clinical outcome. Our case was very unusual in that, the etiology was unusual, hyperviscosity syndrome due to smoldering multiple myeloma.

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Conflicts of interest

There are no conflicts of interest.

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