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Idiopathic Orbital Inflammatory Disease Mimicking a Carotid Cavernous Fistula

Nurhayati Abdul Kadir, Syed Shoeb Ahmad*, Shuaibah Abdul Ghani

Ophthalmology Department, Queen Elizabeth Hospital, Kota Kinabalu, Malaysia
*Corresponding author: syedshoebahmad@yahoo.com

Abstract Idiopathic orbital inflammatory disease (IOID) is a common orbital disorder. However, it has a highly variable clinical presentation depending on the site, degree of inflammation and size of the lesion, which produces a mass effect in the orbital cavity. IOID has no pathognomonic clinical features, thus radiologic and occasionally histopathologic studies are required to confirm the diagnosis of IOID. Here we report a case of a healthy elderly gentleman who presented to us with proptosis, dilated and tortuous conjunctival and retinal vessels and a positive response to Valsalva maneuver. These clinical features were suggestive of a carotid cavernous fistula (CCF). An initial CT scan showed an intraconal orbital mass. Hence, an MRI was subsequently performed which did not show features of CCF, however, there was again a large intraorbital mass noted. The radiologic differential diagnosis suggested was IOID, lymphoma and metastasis. Hence, a biopsy of the intraorbital mass through the transnasal approach was performed. That confirmed the lesion to be IOID and not CCF. Hence, although proptosis with conjunctival congestion, dilated tortuous conjunctival and retinal vessels with positive Valsalva maneuver are the usual clinical features of CCF, this case illustrates these features may also occur in patients with IOID.

Keywords: *orbital inflammatory pseudotumor, carotid cavernous sinus fistula, Valsalva maneuver, inflammation, proptosis*

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1. Introduction

Idiopathic orbital inflammatory disease (IOID) is a nonspecific, non-neoplastic inflammatory process of the orbit. [1] The orbital inflammation may range from idiopathic inflammatory disease to that secondary to systemic or local inflammatory conditions. Although the presentation is variable, the common symptoms are proptosis and ocular pain.

Carotid cavernous sinus fistulas (CCF) are abnormal communications between the cavernous sinus and the carotid arterial system. The presence of conjunctival congestion, dilated retinal vessels with a positive Valsalva maneuver are usual features in a CCF. These features have not been described in a case of IOID.

This case report describes a patient who had clinical features suggestive of CCF, but on investigation was diagnosed with IOID.

2. Case Report

A 70-year-old healthy gentleman presented to us with a sudden onset of proptosis of the right eye associated with diplopia, red eye and decreased vision for the last three weeks. There was no history of any systemic illness, fever, respiratory infection or ocular trauma. The visual acuity in the right eye was 6/9 and 6/6 in the left. The intraocular pressures were 17 and 11 mm Hg in right and left eyes respectively. The right eye was congested and proptosed. A Hertel exophthalmometry examination revealed that the right eye was 10mm and left eye 6mm. The conjunctiva was mildly congested with dilated, cork-screw vessels notably present towards the lower fornix. These vessels became more prominent on Valsalva maneuver (Figure 1). The pupil was reactive and a mild relative afferent pupillary defect was noted. The rest of the anterior segment evaluation was unremarkable. Fundus examination of the right eye showed a swollen optic disc and dilated tortuous retinal vessels in all quadrants (Figure 2). The anterior and posterior segments of the left eye were unremarkable. These features suggested a right sided CCF.

Erythrocyte sedimentation rate (ESR) was 43mm/H (normal < 35mm/H) and C-reactive protein was 0.77mg/dL (normal < 0.33mg/dL). Full blood count, renal and liver profiles, thyroid function tests and other investigations for autoimmune diseases were normal.

A computerized tomography (CT) scan showed there was an ill-defined enhancing mass in the intraconal space of the right eye. This mass extended posteriorly until the orbital apex. The optic canal and superior orbital fissure were not widened (Figure 3). CT angiography showed the superior orbital veins were not dilated and other cerebral vessels were normal. A magnetic resonance imaging (MRI) scan revealed the cavernous sinuses were symmetrically concave. Subsequently, a trans-nasal endoscopic biopsy was done and the histopathology examination showed lymphocytic inflammatory infiltrates with fibrosis (Figure 4).

Figure 1. Pre- and post- Valsalva maneuver appearance

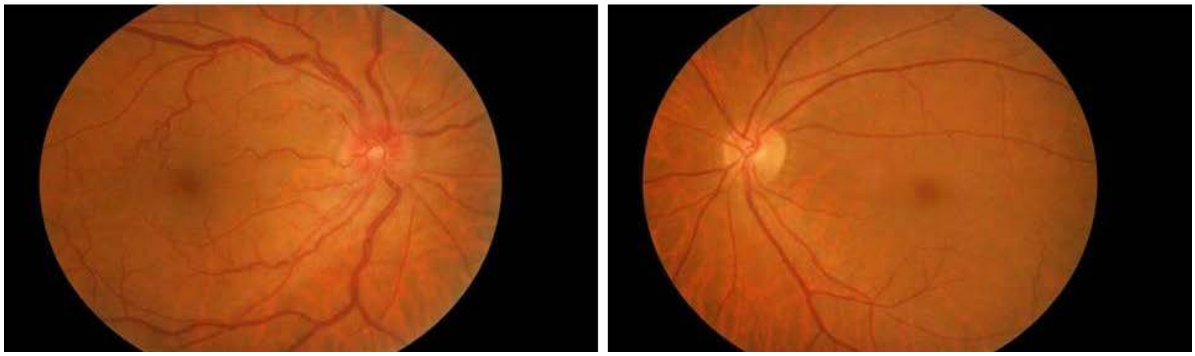

Figure 2. Tortuous retinal vessels in right eye

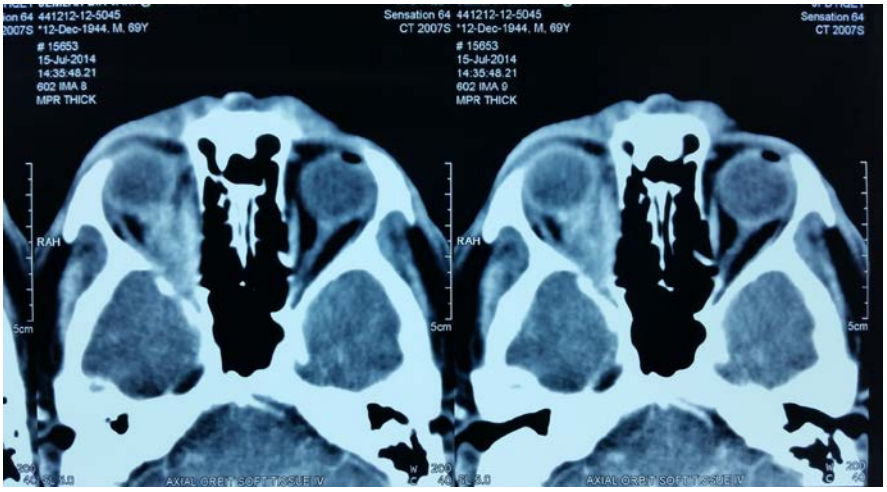

Figure 3. CT scan showing ill-defined enhancing intraconal mass

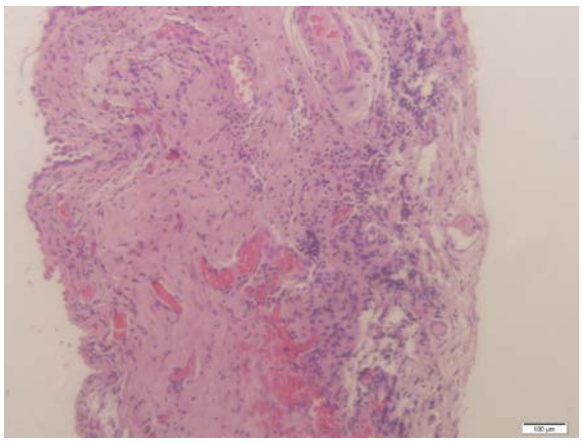

Figure 4. HPE of the lesion

Based on the radiologic and histopathologic findings a final diagnosis of IOID was made. The patient was started on Tab. Prednisolone 40mg per day and tapered by 10mg every week for a month. A significant improvement was noted in the patient's condition. His vision returned to 6/6 in the right eye, with a full range of ocular movements and resolution of the dilated conjunctival and retinal vessels (Figure 5). A repeat MRI scan showed the orbital mass had considerably shrunk in size (Figure 6).

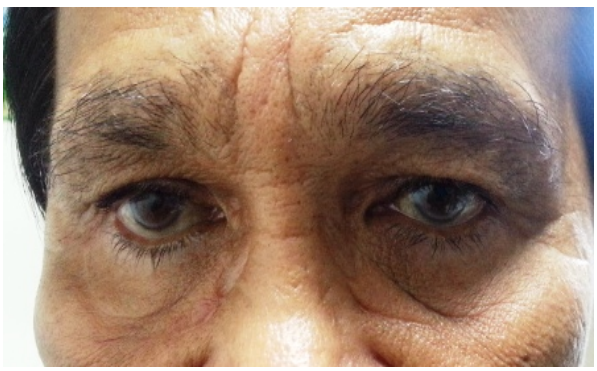

Figure 5. Post-treatment appearance

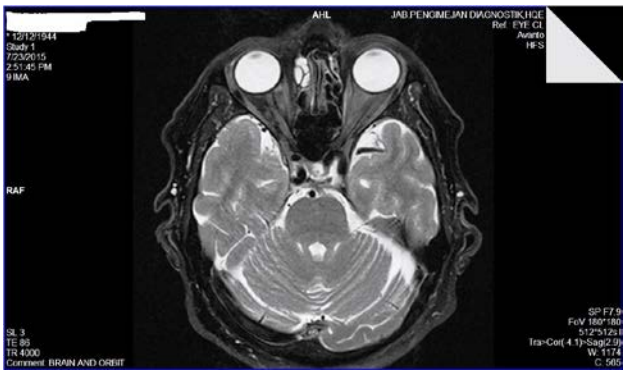

Figure 6. Post-treatment MRI

3. Discussion

This case report describes a patient who had clinical features suggestive of CCF. However, further investigations proved it to be IOID. Thus, this case report shows that IOID can have a varied presentation and can mimic CCF.

IOID encompasses a group of heterogeneous inflammatory diseases of the orbit of unknown etiology. It accounts for about 6% of all orbital disorders. [2] The condition is commonly seen in middle aged females, with a predominantly unilateral involvement.

This condition is characterized by focal or diffuse inflammation and infiltration of the orbit. Adnexal structures such as the lacrimal gland and extraocular muscles may also get involved. The disease typically presents with an abrupt onset of periorbital pain, swelling, proptosis, red eye, restricted extraocular movements, ptosis and decreased vision. The clinical presentation can vary according to the specific location and degree of inflammation, fibrosis and mass effect in the orbit. [2,3,4,5] The etiology is likely autoimmune in origin with viral, genetic and environmental factors proposed as possible trigger factors.

The differential diagnosis of IOID includes: idiopathic inflammatory disease; systemic or local inflammatory conditions; and other diseases such as neoplasms, infections, congenital malformations and trauma. (Table 1). However, it is not reported if IOID can present with features suggestive of CCF. Our patient had dilated corkscrew conjunctival vessels inferiorly which demonstrated dynamic changes on Valsalva maneuver. These features are more common in vascular lesions.

CCF is an abnormal communication between the cavernous sinus and the carotid arterial system. It is characterized by a high-flow system arising from direct communication between the intracavernous internal carotid artery and the cavernous sinus, or dural fistula arising from a communication between dural arteries and cavernous sinus (also known as indirect CCF). The dural fistulas usually have low rates of arterial blood flow and may be difficult to diagnose clinically due to much more subtle clinical features than direct fistulas. [6,7]

The clinical features of CCF include corkscrew episcleral blood vessels, conjunctival chemosis, proptosis; with or without pulsation, diplopia, ophthalmoplegia, orbital pain, audible bruits and blindness. [7] However dural fistulas usually present with less severe features. They have an insidious onset, mild orbital congestion with dilated conjunctival vessels, proptosis and low or no bruit. [6,7,8] As in our case, the patient presented with clinical features suggestive of dural CCF. The diagnosis was only ascertained following radiologic and histologic studies.

Table 1. Differential diagnosis of IOID

Idiopathic Inflammatory Disease
Systemic inflammatory disease
Autoimmune thyroid disease: endocrine exophthalmos
Sarcoidosis
Wegener's granulomatosis
Crohn's disease
Systemic lupus erythematosis
Churg-Strauss syndrome
Erdheim-Chester syndrome
Histiocytosis X
Giant cell arteritis
Idiopathic fibrosclerotic disorders
Periarteritis nodosa
Scleroderma
Sclerosing cholangitis
Neoplasm
Lymphoma
Lymphoproliferative disorders
Rhabdomyosarcoma
Choroidal malignant melanoma with extrascleral spread
Metastatic disease
Congenital malformation
Dermoid cyst
Lymphangioma
Infectious disease
Trauma

Preliminary investigations in patients presenting with orbital lesions should include a careful history and complete physical examination followed by full blood count, ESR, C-Reactive Protein (CRP) and radiologic imaging with a CT and/or MRI scan of the affected orbit. Further investigation depends on the individual clinical features. In equivocal cases, as in our patient, it is recommended to perform a biopsy to confirm the diagnosis of IOID, except in the case of pure myositic locations and posteriorly located tumors where there is a significant risk of damage to the optic nerve [9].

4. Conclusion

In summary, orbital inflammation can have a varied presentation. As this case report has shown, IOID can also present with features mimicking a CCF. Thus, these conditions should be included in their differential diagnosis.

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