Idiopathic Orbital Myositis Presenting as Central Retinal Artery Occlusion

Shivcharan Lal Chandravanshi1*, Dinesh K Sahu1, Anup Chirayath1 and Aditi Dubey2

1Ahalia Foundation Eye Hospital, Palghat, Kerala, India
2Shyam Shah Medical College Rewa MP, India

Abstract

A 55-year-old man presented with central retinal artery occlusion secondary to idiopathic orbital myositis. The cardiac evaluation revealed co-existing myocarditis. The patient was treated with intravenous methyl prednisolone therapy which resolved the inflammatory process but during the course of treatment patent died due to cardiac complications. Few cases of central retinal artery occlusion secondary to orbital pseudotumors have been reported. In rare circumstances idiopathic orbital myositis may associated with giant cell myocarditis. We emphasized that idiopathic orbital myositis should be monitored for coexisting giant cell myocarditis because of its life threatening complications.

Introduction

Idiopathic orbital inflammation (inflammatory pseudo tumors) is a nonspecific benign orbital inflammatory process with no evidence of local and systemic cause [1]. Depending upon the predominant orbital structure involvement, it is classified into five groups: myositis, dacryoadenitis, anterior, diffuse, and apical. Orbital myositis is the most common nonspecific inflammatory disease of the orbit. Orbital myositis can be further subdivided into three forms: isolated, recurrent and atypical [2]. The typical presentation of the orbital myositis consists of periorbital inflammation and swelling, retrobulbar pain and pain on ocular movements, proptosis, diplopia, and occasionally diminution of vision [1]. Here we are describing a very rare case of idiopathic orbital myositis (IOM) that presented to us with complaints of total loss of vision secondary to central retinal artery occlusion (CRAO).

Case Report

A 55-years old man was admitted to the eye hospital because of sudden loss of vision in left eye associated with breathlessness since 3 days. There was no history of previous episodes of orbital inflammation in the past. There was no family history or past history of any cardiac or respiratory disease. Ophthalmic examination revealed the vision was 20/20 in right eye and no perception of light in left eye. The intraocular pressure by applanation tonometry was 16 mm of Hg in both eyes. The patient had left eye proptosis and 3 mm globe displaced laterally (Figure1). Hertel’s exophthalmometry revealed 15 mm in right eye and 19 mm in the left eye displacement, with a base measurement of 100 mm. The left eye ocular movements were painful and limited in all directions of gaze (Figure2). There was a moderate mechanical ptosis, conjunctival congestion, chemosis in left eye. Pupillary examination showed mid dilatation with gross relative afferent pupillary defect. Dilated fundus examination revealed left eye pale disc, cherry red spot and retinal edema along with narrowing of blood vessels with segmental blood flow (Figure3). Rest ocular examination was unremarkable. The patient was not evaluated with fundus fluorescein angiography and indocyanine green angiography due to his poor systemic conditions. Brain and orbital computerized tomogram scan demonstrated a fusiform enlargement, involving both tendon and belly of left medial rectus muscle, with moderate compression of the optic nerve and nerve sheath in region of orbital apex (Figure 4, 5). Brain and cavernous sinus are unremarkable on computed tomography scan.

Physician evaluation revealed breathlessness which was more on lying down and associated with edema of dependent part of body. Laboratory investigations such as blood sugar, complete haemogram, thyroid function, tuberculin test, angiotensin-converting system, laboratory evaluation revealed co-existing myocarditis. The patient was treated with intravenous methyl prednisolone therapy which resolved the inflammatory process but during the course of treatment patient died due to cardiac complications.

Figure 1: The Clinical photograph showing left eye periocular swelling proptosis, conjunctival congestion, lateral displacement of globe and mechanical ptosis.

Figure 2: The Clinical photograph showing restricted extra ocular movements.
antiphospholipids antibodies, and ANCA are unremarkable. X-ray chest and para nasal sinuses were normal. Electrocardiography showed tachyarrhythmia and echocardiography suggested normal valves and ejection fraction of 24%. Patient was treated in cardiac intensive care unit under physician supervision with intravenous methyl prednisolone (IVMP) 1gm daily for 3 consecutive days followed by oral prednisolone 60 mg daily along with symptomatic and supportive treatment [3]. There was no improvement in vision; however proptosis and other inflammatory signs almost subsided after IVMP therapy (Figure 6). Patient showed symptomatic improvement with no functional cardiac improvement on echocardiography. Intensive monitoring of the patient did not reveal any serious life threatening side effect of IVMP therapy. In view of sudden onset of painless unilateral loss of vision, proptosis, limitation of extraocular movements in all directions of gaze, diagnosis of IOM with CRAO was made. The patient died after 7 day due to congestive cardiac failure secondary to myocarditis. Postmortem was not done in our case because next-of-kin had refused for same. Institution ethical committee had approved it for publication. Next-of-kin of deceased had issued written consent for publication of photographs.

Discussion
Van Graefe described CRAO first in 1859, as an embolic event to the central retinal artery in a patient with endocarditis [3]. CRAO patients typically present with sudden, severe, and painless loss of vision in one eye. It had multi-factorial etiology which can be divided into four categories: obstructive, spasmic, compressive and direct injury to artery [4]. Compressive causes of CRAO included orbital tumor especially at orbital apex [5]. There are a few reports of CRAO secondary to pseudotumor [6,7]. In our case, the cause of CRAO is compression at orbital apex secondary to IOM. IOM is the most common type of nonspecific idiopathic orbital inflammation. The differential diagnosis of IOM includes; Thyroid related orbitopathy (TRO), Tolosa-Hunt syndrome, primary or secondary tumor of extraocular muscles, orbital lymphoma, arteriovenous fistulas or malformation, orbital metastases, myasthenia gravis, and amyloid deposition [1]. However, TRO usually painless in onset, bilateral, asymmetrical, associated specific signs such as lid retraction, lid lag in down gaze, and abnormal serum T3, T4 and TSH level. On computed tomography scan in the TRO, the extraocular muscle enlargement is usually in belly in contrast to IOM both tendon and belly are involved. Tolosa-Hunt syndrome usually has deeper, more constant orbital pain and associated with multiple cranial nerve palsies. Imaging study can revealed either cavernous sinus enlargement or normal orbital findings. In our patient there was no cavernous sinus enlargement of computed tomography scan.

Metastases or local malignancy of extraocular muscle may masquerade as an IOM but acute and painful onset is very rare presentation. The first line of management of idiopathic orbital inflammation is oral corticosteroids in high doses followed by gradual tapering [1]. In our case we had chosen IVMP therapy in place of oral steroids because of severity of disease. Immunosuppressive are reserved for steroid resistant cases. Biopsy is usually indicated for recurrent and steroid resistant cases [2]. Usually IOM does not have any direct association with other systemic disorders except underlying immune disorders such as allergy, collagen vascular disorders, linear scleroderma and Crohns disease [8,9]. Rarely it may be associated with giant cell myocarditis [9]. In our patient ECG and Echocardiography

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**Figure 3:** Fundus color montage photograph of the left eye showing pale disc, cherry red spot and retinal edema along with narrowing of blood vessels with segmental blood flow.

**Figure 4:** Axial computed tomography of orbit showing homogeneous fusiform enlargement of medial rectus with involvement of belly and tendon both.

**Figure 5:** Coronal computed tomography of orbit showing crowding at orbital apex secondary to idiopathic isolated myositis of medial rectus muscle.

**Figure 6:** The clinical photograph showing marked improvement in proptosis and other inflammatory signs of idiopathic orbital myositis after intravenous methyl prednisolone therapy.
were suggestive of myocarditis. No other cause of myocarditis was found on extensive cardiac evaluation. The cause of Idiopathic orbital myositis and giant cell myocarditis is not known but often presumed to be autoimmune [9]. We presume that our patient died due to congestive cardiac failure secondary to giant cell myocarditis associated with IOM. However it may have only been confirmed by histopathological examination which was not done in our case [10]. Giant cell myocarditis had very high mortality rate, however timely cardiac transplantation may save life in these patients [10].

We suggest giant cell myocarditis should be monitored for in the course of IOM because of its life-threatening fulminant course.

References