Superior Orbital Fissure Syndrome in a Patient with Partially Treated Granulomatosis with Polyangiitis: A Case Report

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ARTICLE INFO
Article History
Submitted: 15 November, 2022
Accepted: 20 December, 2022
Published: 8 February, 2023

Source of support: None
Conflict of Interest: None

ABSTRACT

Superior orbital fissure syndrome (SOFS) is a collection of symptoms caused by compression of structures just anterior to the orbital apex. The optic nerve is spared which differentiates superior orbital fissure syndrome from orbital apex syndrome. The causes are trauma, neoplasms, infections, inflammation and vascular phenomena. SOFS in granulomatosis with polyangitis (GPA) is a rare association and has been found only in some case reports till date. This rare case highlights development of SOFS and multiple serious adverse effects of glucocorticoid in an inappropriately treated patient with GPA without renal or pulmonary involvement.

Keywords: Cataract; Glaucoma; Glucocorticoid-induced osteoporosis; Granulomatosis with polyangiitis (GPA); Superior Orbital Fissure Syndrome (SOFS).

INTRODUCTION

Granulomatosis with polyangitis (GPA) is an ANCA-associated small vessel vasculitis with multisystem involvement. In the decreasing order of frequencies, the systems involved are ENT, joints/muscles, kidneys, lungs, eyes, heart, skin, peripheral nerves, CNS and GI tract.[1] The annual incidence and prevalence rates of GPA are 2.1-14.4 and 46-184 per million respectively. Granulomatous ear, nose and throat (ENT) lesions are the most common manifestations with gradual destruction of nasal cartilage, which may lead to saddle-nose deformity.[3,4] Eye and orbital involvement may be a striking feature, and the frequency of such involvement ranges from 30%-60% over time. Constitutional symptoms can be present in >58% of patients.[5] The pathophysiology of inflammation involving the orbits may be from vasculitis or spread from paranasal sinuses.[6] Manifestations can be non-organ threatening, organ threatening or life threatening.[7,8] Progression to systemic vasculitis, particularly with glomerulonephritis or alveolar hemorrhage, remains a common route to diagnosis. Superior orbital fissure syndrome (SOFS) is caused by compression of structures just anterior to the orbital apex producing symptoms of cranial nerve, pupillary and extraocular muscles.[9] The optic nerve is spared which differentiates SOFS from orbital apex syndrome.[10] The most common cause of SOFS is trauma followed by neoplasms (particularly lymphoma and rhabdomyosarcoma), infections (such as meningitis, syphilis, sinusitis, herpes zoster), inflammation (such as SLE, sarcoidosis), and vascular phenomena (such as carotid-cavernous fistulas, retro-orbital haematoma and carotid aneurysms).[11-19]

CASE REPORT

A 35-year-old male was diagnosed as GPA six years back on the basis of bloody nasal discharge, headache,

purpuric skin rashes, inflammatory polyarthritis involving small and large joints and positive C-ANCA. Biopsy from nasal mucosa was consistent with small vessel vasculitis and extravascular granulomatous lesions. He was then put on oral cyclophosphamide 100 mg daily and prednisolone 60 mg daily, with which he had a good response as early as 1 month of treatment. Then prednisolone was gradually tapered to 20 mg daily and cyclophosphamide was stopped 6 months later without any maintenance immunosuppressive agent. He tried to stop prednisolone abruptly, but he failed to give up the drug as it was associated with recurrence of headache and bloody nasal discharge. He also developed an episode of dizziness trying to avoid prednisolone for a few days last year, and fell down from standing height. He felt severe pain in the back and radiological evaluation revealed osteoporotic fracture of 11th dorsal and 5th lumbar vertebrae. He also developed steroid-induced cataract and glaucoma. He then consulted Department of Rheumatology, BSMMU with complains of dizziness, shortening of height, epistaxis, nasal deformity and diplopia. On examination, he had nasal crusts with rhinitis, saddle-nose deformity, cushingoid face, central obesity with thin limbs and bilateral proptosis (Rt.>Lt.). Examination of cranial nerves revealed involvement of bilateral olfactory, oculomotor, divisions of trigeminal (ophthalmic, maxillary & mandibular) and abducens nerves (Figure 1). Sinus CT revealed thickening of mucosa of all paranasal sinuses and ethmoidal sinus mass enroaching the orbital apex (Figure 2). MRI of the orbits also showed similar findings. Culture of nasal discharge was done to exclude infection. Urine routine microscopic examination and chest X-ray findings were normal. Induction therapy was instituted with intravenous pulse cyclophosphamide 15 mg/kg and prednisolone 1 mg/kg/day. Vaccination against pneumococcus, influenza and hepatitis-B virus was given. He was put on adequate calcium, vitamin-D and risedronate. Pneumococcal carinii pneumonia prophylaxis was started simultaneously. Management of cataract and glaucoma was instituted according to ophthalmologist consultation. On his next follow up after 1 month, there was significant neurological improvement. Then, he scheduled his follow-up in vasculitis and glaucoma clinics in a tertiary care hospital.

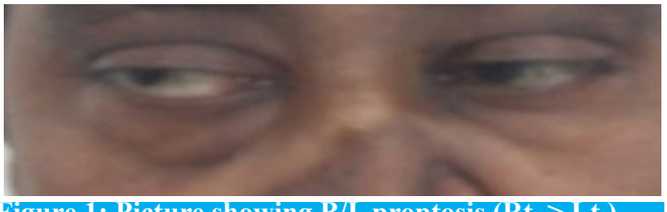

Figure 1: Picture showing B/L proptosis (Rt. > Lt.), B/L ptosis, partial abducens nerve palsy of Rt. eye and saddle nose deformity

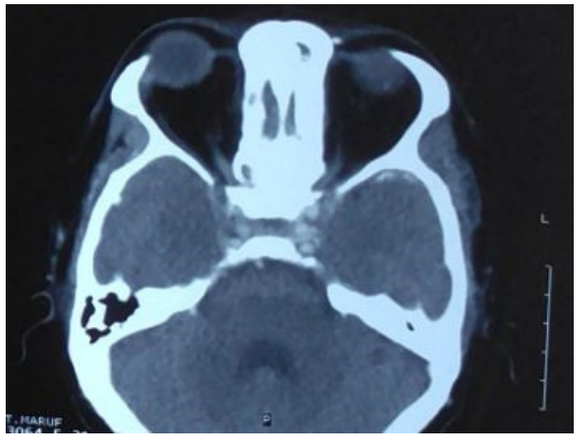

Figure 2: CT scan head Transverse view showing ethmoidal sinus mass enroching orbital apex (white arrow)

DISCUSSION

GPA is an uncommon cause of SOFS. The peculiarity of this case is the paranasal sinus disease without involvement of other systems. There are very few published case reports of SOFS in association with GPA. The common route of diagnosis of GPA is renal and/or pulmonary involvement. Limited form of GPA involves head and neck without any renal involvement and has a better prognosis. SOFS can lead to extraocular muscles involvement or visual impairment, which is really important in this case. Diagnosis can be best established on the basis of clinical features and imaging. Early use of plain film with superior orbital fissure views (20-25° head tilt) has largely been replaced by widespread availability of CT and MRI. Currently, the most common imaging modality is CT with 2mm fine cuts looking for bony fragments or compressive masses. When a vascular cause is suspected, angiography is recommended to define any carotid-cavernous fistula or carotid aneurysm.[12]

Management of SOFS includes medical and surgical options. Indications for urgent intervention are vascular causes (requiring embolization) and bony fractures with displaced fragments or severe foraminal narrowing.[11] Medical treatment is done with mega doses of steroids, especially for patients not meeting criteria for urgent surgical intervention. The common regime is IV methylprednisolone 30mg/kg bolus followed by a 5.4mg/kg 48 hour infusion and eventual oral prednisone tapered over 2 weeks.[12,20] Choice of immunosuppressive therapy includes induction with either of IV cyclophosphamide (CYCLOPS protocol) or IV rituximab (RITUXIVAS protocol) followed by maintenance therapy.[21,22] Surgical management includes both orbital and cranial extradural approaches or extranasal intraorbital to access the

lateral wall, extranasal transethomoidal to access the medial wall, a modified extranasal intraorbital route, transtemporal route in causes of infectious collections and combined orbital and cranial for deep decompressions.[23,24] Complete recovery of all nerves has been reported in 24-40% of patients receiving steroid treatment compared to 21.4% in those without. Recovery is usually extended over a period of months with progress plateauing at 6 months. Abducens nerve shows the best recovery. Possible sequelae from superior orbital fissure syndrome may include remaining deficit requiring further strabismus or ptosis surgery.[11,12]

CONCLUSION

Superior orbital fissure syndrome (SOFS) is caused by compression of structures just anterior to the orbital apex producing symptoms of cranial nerve, pupillary and extraocular muscles. This report shows that there is development of SOFS and multiple serious adverse effects of glucocorticoid in an inappropriately treated patient with GPA without renal or pulmonary involvement.

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