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Was it Just an Orbital Inflammation or Tip of the Iceberg - A Case Report

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Abstract: ***Orbital inflammation could be of various etiologies. We report a case of a young female presented to us with recurrent episodes of redness, pain, swelling in right eye for 3 weeks. Presence of joint pains and swelling in the parotid gland area called for an additional workup. Investigations were positive for Granulomatosis with polyangitis(GPA). Final diagnosis of orbital inflammation secondary to GPA was made and patient was treated. To conclude, though the condition is rare ophthalmologist should have a high index of suspicion in such a case scenario because of significant morbidity and loss of vision.***

Keywords: orbital inflammation, Granulomatosis with polyangitis

1. Introduction

Orbital inflammation could be of various etiologies. However, Cause of this inflammation could range from a simple local cause to a potentially morbid systemic cause. Granulomatosis with polyangitis (GPA) though uncommon can present with orbital inflammation. GPA is an autoimmune condition resulting in granulomatous vasculitis. It involves multiple systems. However there could be other systemic manifestations associated but in 8–16% of cases ocular involvement might be the only presenting feature. Orbital GPA can cause significant morbidity and might even result in complete loss of vision. Here we report a case of unusual presentation of orbital inflammation secondary to GPA. We mainly emphasise on the importance of high suspicion of condition, its early diagnosis and treatment.

2. Case Report

A 22 year female, presented to us with recurrent episodes of redness, pain, swelling in right eye since 3 weeks. Furtherly, Patient had history of ear discharge, was evaluated and underwent right sided mastoidectomy 2 months ago and complains of fullness of right cheek which was there since post surgery. Patient also had complains of joint pains on and off.

On examination, best corrected visual acuity was 6/6 in both eyes, extra-ocular movements were restricted in all directions of gaze in right eye and was normal in left eye. On slit lamp examination right eye proptosis noted with lid oedema and chemosis.(figure 1). Orbital fullness was noted superior laterally in right eye. Left eye was within normal limits. Based of clinical findings tentative diagnosis of right eye orbital cellulitis was made and she was admitted for further management. Patient was started on IV antibiotic (augmentin 1.2mg tid) with topical antibiotic steroid drops in right eye.

Figure 1: Clinical picture showing right eye proptosis, lid oedema and chemosis

ENT opinion was sought in view to rule out sinus causes of orbital cellulitis and right cheek fullness (figure 2), patient was diagnosed with right sided parotitis and adviced a CT scan.

Figure 2: Clinical picture showing conjunctival chemosis with congestion and fullness in the parotid area of right side

Rheumatology opinion was sought in view of join pains and was suspected to have granulomatosis with polyangitis with present clinical manisfestations.

Blood investigations including CBC, C-ANCA, P-ANCA, and were asked, which reported high WBC count, ESR and CRP values with C-ANCA and P-ANCA being strongly positive. CT scan suggested right lacrimal gland inflammation involving lateral rectus muscle with periosteal involvement in lateral wall of right orbit. A diagnosis of acute granulomatosis with polyangitis was made and 3 doses of inj. solumedrol 750mg in 500ml ns was given.

Meanwhile patient was responding well to treatment, extra ocular movements improved, lid oedema and chemosis resolved. Patient improved and was discharged.

3. Discussion

Granulomatosis with polyangiitis is a systemic disease. It involves multiple-systems in the body. It mainly affect small to medium sized arteries, capillaries and veins. Necrotizing non-caseating granulomatous vasculitis is seen.

50–60% of patients with this disease are reported to have ocular involvement. In 8–16% of cases ocular involvement might be the only presenting feature.[1] Clinical findings of ocular GPA arise from the inflammation of ocular structures, orbital contents including lacrimal glands, and optic nerve. Clinical presentation of these patients with ocular involvement can range from simple ocular pain, erythemato severe proptosis, diplopia, and vision loss.[2],[3],[4]

Ocular inflammation, along with systemic symptoms as in this case, is a red flag for initiation of systemic investigation for underlying disease. Antineutrophil cytoplasmic antibody serologies which include cytoplasmic ANCA and perinuclear ANCA are elevated and highly useful in GPA diagnosis.[3],[5]

In our case, positive C-ANCA, elevated erythrocyte sedimentation rate (ESR), c-reactive protein (CRP) supported the diagnosis of GPA.

CT findings of orbital GPA usually shows granulomatous infiltration of the orbit, at times even resulting in bony destruction. Magnetic resonance imaging of orbital inflammation can reveal hypointense lesions on T2-weighted studies with variable contrast enhancement.[4]

Without systemic treatment, ocular disease may be recurrent whenever ocular treatment is tapered, so treatment of ocular symptoms is usually in the context of systemic treatment.GPA responds well to systemic corticosteroids and cytotoxins (cyclophosphamide, methotrexate, or azathioprine). Treatment is given as induction dose and maintenance dose to prevent remission.[4],[6]

However, in patients presenting with sight threatening orbital involvement leading to severe proptosis and optic nerve compression surgical decompression of the orbit need to be considered.

4. Conclusion

Limited orbital granulomatosis with polyangiitis (GPA) is an uncommon and challenging disease the diagnosis of which may be delayed, especially in patients with isolated orbital involvement as the initial presentation, because clinical manifestations are non-specific and may mimic other less significant illnesses, and systemic diagnostic criteria are not applicable.

Making an early correct diagnosis of GPA limited to orbital structures despite the absence of systemic progression is extremely important because it can be locally destructive, with irreversible visual and functional loss.

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