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RECURRENT EPISCLERITIS - A RARE PRESENTATION OF MIXED CONNECTIVE TISSUE DISEASE.

Ophthalmology

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ABSTRACT

Episcleritis refers to inflammation of the loose connective tissue between the sclera and the conjunctiva. The majority of episcleritis cases are idiopathic, but 26% to 36% of patients have an associated systemic disorder that is responsible for the pathological process and development of episcleritis. It is also well established that the incidence and prevalence of episcleritis are higher in populations with systemic collagen-vascular disease and autoimmune diseases. However recurrent episcleritis in Mixed Connective Tissue Disease has not been reported before in literature. Here we are reporting a case of recurrent bilateral nodular episcleritis as a rare presentation of MCTD.

KEYWORDS

Case Presentation:

A 57-year-old male patient presented to our OPD with complaints of persistent redness in his right eye and headache since the past week.

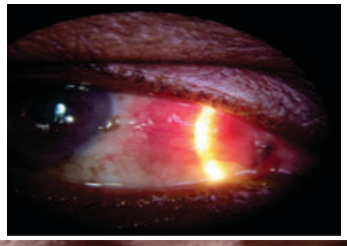
The redness was not associated with itching, discharge, photophobia or pain on extraocular movements. The headache was frontal and bitemporal, increasing in intensity at night, not associated with nausea or photophobia, and occurring about 7-8 times a month.

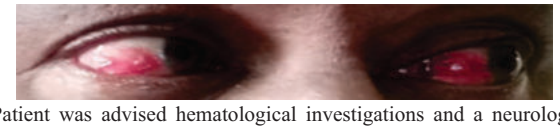
The patient has had similar complaints since the past 8 months, with the redness recurring in both eyes, either nasally or temporally. He has had no history of fever, seizures, cough, gastrointestinal symptoms or any oral or genital ulcers recently.

The patient had an episode of headache with diplopia in his right eye 8 months ago. He was investigated in another hospital, where MRI sequencing was performed. It showed enhancement in the intraconal fat at the right orbital apex and enhancement in the optic nerve sheath of posterior retrobulbar portion of the right optic nerve (Right optic perineuritis). There was also asymmetrical thick meningeal enhancement along the right anterior temporal lobe. Patient was prescribed oral prednisolone 60mg with rapid tapering over 20 days, which resolved his symptoms.

The eye redness and headache recurred after 2 months for which he was prescribed topical steroid eyedrops in multiple hospitals to which he was nonresponsive.

Patient then presented to our OPD; at presentation VA was 6/18 and 6/24 in RE and LE respectively, with a BCVA of 6/6 in both eyes. He had nodular episcleritis of the nasal aspect of RE and temporal aspect of LE. IOP was 16mm Hg and 18mm Hg in RE and LE. The rest of the anterior segment was normal. Fundus examination was within normal limits.

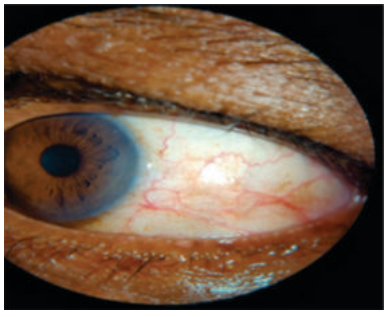




Patient was advised hematological investigations and a neurology opinion for the headache. ESR was elevated with a level of 20 mm at the end of 1 hour, and 42mm at the end of 2nd hour. ANA profile showed positivity for anti-U1-snRNP antibody. Viral profile, Mantoux test and Chest x-ray were normal. The neurologist carried out a lumbar puncture for CSF analysis which revealed slightly elevated proteins (95mg/dl), and a repeat MRI which revealed mild meningeal enhancement along the right anterior temporal lobe, s/o pachymeningitis.

Upon further evaluation, patient reported a history of swollen hands with a tightening sensation, which has reduced since initiation of treatment. By exclusion of other diseases, the patient was diagnosed with Mixed Connective Tissue Disease (MCTD) presenting with nodular episcleritis and pachymeningitis, with a high clinical suspicion of CNS vasculitis.

The patient was started on azathioprine 50mg OD and 5mg prednisolone every alternate day, with monthly checks of CBP, LFTs and IOP monitoring. The patient is symptomatically better with no headaches or episcleritis episodes in the 8 months since initiating treatment. IOP is maintained below 20mmHg in both eyes without any treatment.



DISCUSSION:

The episclera is a fibroelastic structure between the superficial scleral stroma and Tenon's capsule and consists of two layers loosely joined together- an outer parietal and an inner visceral layer. The vascular supply of both layers is derived from the anterior ciliary arteries, which start from the muscular branches of the ophthalmic artery.

Episcleritis is a self limiting inflammation of the superficial layers of the sclera, associated with a localized lymphocytic infiltration of episcleral tissue associated with edema and congestion of the overlying conjunctiva and tenons capsule. It is more commonly diagnosed in middle aged females; however there is no consensus on incidence and prevalence as a majority of patients self diagnose. There are two forms of episcleritis: nodular and simple. Nodular episcleritis presents as a discrete, elevated area of inflamed episcleral tissue. In simple episcleritis, vascular congestion is seen and there is absence of an obvious nodule. It is known that simple episcleritis is more common (70%) than nodular episcleritis (30%).

In cases of recurrent episcleritis with persistence of inflammation beyond 20 days, further systemic evaluation is warranted. In a study done by Sainz de La Maza et al., majority of cases were idiopathic, and an associated disease was found in about 27% of patients with episcleritis. A complete review of systems along with hematological and radiographic testing should be performed in accordance with other suspected symptoms which may include joint/muscle pain or weakness, skin rash, psoriasis, diarrhea, oral or genital ulcers, history of drug abuse or possibility for sexually transmitted infections or any other symptoms to suggest a systemic association.

Mixed Connective Tissue Disease (MCTD) is an autoimmune disorder defined as an individual disease by Sharp et al. in 1972. It is characterised by the presence of antibody u1 snRNP and shows a mix of symptoms of SLE, Scleroderma and Polymyositis. Diagnosis are according to various criteria, one which is the Alarcon-Segovia criteria, with a serologic criteria of u1 Sn-RNP antibody positivity and clinical criteria of swollen hands, synovitis, myositis, Raynauds phenomenon and acrosclerosis.

CONCLUSION:

We report a case of MCTD presenting with recurrent nodular episcleritis, who responded to treatment. Hypertrophic pachy meningitis has occasionally been associated with MCTD and has rarely even lead to the diagnosis. However, there have not been any reported cases of recurrent episcleritis with MCTD in current literature. The disease has an excellent prognosis with immuno suppression, and treatment should be initiated especially in patients with pachymeningitis and vasculitis, which can cause CNS complications.

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