

"Cauliflower Ears" in a Patient with Anterior Uveitis

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Summary

"Cauliflower ears" describes the appearance of swelling of the auricular cartilage resulting in gross distortion of the auricle. The underlying pathology is inflammation of the auricular cartilage. We report a case of a patient who presented to the ophthalmology clinic with an ocular inflammation, whose subsequent diagnosis of relapsing polychondritis was clinched with the finding of "cauliflower ears". This case emphasizes the importance of an appropriate general examination of ophthalmic patients for early diagnosis and prompt institution of medical treatment, which may prevent life-threatening complications to the patient.

Key Words: Cauliflower ears, Anterior uveitis, Relapsing polychondritis

Case Report

A 49-year-old man presented to the eye clinic in UMMC after noticing redness and mild discomfort in both eyes one month prior to his presentation. The redness worsened and was associated with photophobia two weeks prior to his presentation. There was no eye discharge or tearing. He also experienced pain in the small joints of some fingers in both hands. There was bilateral ear discomfort. He was otherwise well with no recent weight loss.

Ocular examination showed congested conjunctival and episcleral vessels in both eyes (Figure 1). Vision was 6/6 bilaterally. Pupils were symmetrical and reactive to light. Slit-lamp

biomicroscopy revealed evidence of anterior uveitis in both eyes with the presence of 1+ cells in the right anterior chamber and 2+ cells associated with minimal flare in the left. This was associated with secondary glaucoma, where the intraocular pressures were 30mmHg bilaterally. Gonioscopic examination showed open angles in both eyes with no peripheral anterior synechiae. The lens in both eyes were not cataractous. Funduscopy was also normal.

The underlying auricular cartilage of both his ears were grossly swollen resulting in the appearance of "cauliflower ears" (Figure 2). The nasal bridge was tender. Several small joints of both hands were inflamed and tender. There was also costochondral junction tenderness.

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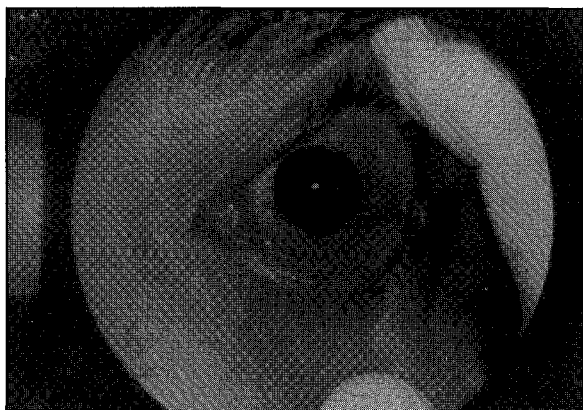


Fig. 1: The patient's left eye showing congested conjunctival and episcleral vessels.

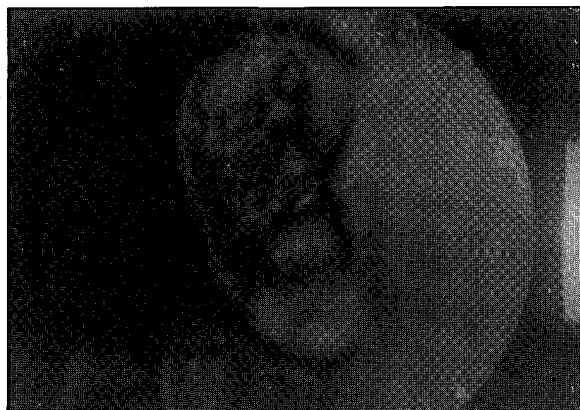


Fig. 2: The patient's right ear was grossly swollen as a result of inflammation of the underlying auricular cartilage, giving the appearance of "cauliflower ears".

A diagnosis of bilateral anterior uveitis secondary to relapsing polychondritis with secondary glaucoma was made. ESR was 75mm/1st hour. Full blood count, serum electrolytes were within normal limits. Anti-DNA antibody was negative. Rheumatoid factor was <20iu/ml. The C3 was 203mg/dL (normal range 83 - 177mg/dL) and C4 was 71mg/dL (normal range: 15 - 45mg/dL). The raised ESR and complement levels support the presence of an inflammatory process.

Topical prednisolone was commenced four times daily for the control of anterior uveitis while gutt timolol 0.5% twice daily was given to control the high intraocular pressures. Gutt homatropine 2% twice daily was added to prevent formation of synechiae in a constricted pupil.

The patient was referred to the rheumatologist and otolaryngologist. He was started on oral prednisolone 30mg daily and warned of the possibility of breathing difficulties. The oral prednisolone was given as prophylaxis against worsening of inflammation by the attending rheumatologist.

Two weeks later the patient presented to the casualty with chest pain. Electrocardiogram showed sinus bradycardia with a heart rate of 52 beats/minutes. No ischaemic changes were recorded during observation in the cardiac unit. The patient had defaulted medication after discharge. Pulsed intravenous infusion of 500mg methylprednisolone was given for 3 days to control flaring up of inflammation involving the joints, costochondral cartilages, heart and the eyes. He also received a stat intravenous infusion of 150mg cyclophosphamide. Oral Azathioprine 50mg BD was given in addition to the prednisolone. The bradycardia resolved. Five months after initial presentation in the eye clinic, the patient's eyes were still showing signs of uveitis and required topical steroids twice daily as maintenance.

Discussion

Relapsing polychondritis is a rare disease manifesting as an episodic, generally progressive, inflammation of cartilagenous structures throughout the body. The eye and ear undergo

recurrent inflammation¹. This disease entity was first described by Pearson et. al in 1960. This disease usually manifests itself between 40 to 60 years of age. Auricular chondritis and arthritis are the most common presenting signs^{1,2}.

In this patient, the diagnosis of anterior uveitis secondary to relapsing polychondritis was made supported by the presence of aural and nasal cartilage inflammation, migratory arthritis and costochondritis. The "cauliflower ears" was a very helpful sign. In this case, such a finding helped clinched the medical diagnosis, thus avoiding unnecessary investigations to determine the aetiology of his anterior uveitis.

Ocular findings, such as conjunctivitis, episcleritis, scleritis, iritis, corneal infiltrates and melting, keratoconjunctivitis sicca, optic neuritis, exudative retinal detachment, chorioretinitis, retinal infiltrates, extraocular muscle palsy and exophthalmos, were reported in 50% - 72% of cases in various series^{1,2}. In 19% of patients, ocular manifestations were the first presenting sign of which the most common were episcleritis and scleritis. Iridocyclitis was seen in 22% to 30% of patients². The ocular findings in this patient were anterior uveitis with secondary glaucoma and episcleritis.

Auricular inflammation and arthritis are self-limiting over weeks. Inflammation involving tracheal and laryngeal cartilage can result in fatal airways obstruction. Cardiac valvular inflammation, coronary arterial vasculitis and complete heart block are potentially serious complications. Our patient presented with

bradycardia and chest pains a week after defaulting oral prednisolone that resolved when treatment was re-commenced. Neurological complications which can occur in this condition include cranial nerve disorders, hemiplegia, seizures, hallucination, confusion and cerebellar signs, were not observed in this case.

Serious ocular complications which can develop include secondary glaucoma from severe iritis, phthisis bulbi, corneal perforation, secondary cataracts, retinal vasculitis and optic neuritis. Therefore regular ocular examinations of such patients are essential, especially during episodes of recurrent inflammation.

There was a 30% mortality rate in McAdam's series¹. The causes of death included infection, acute respiratory failure from airway obstruction, dissection or rupture of large artery aneurysm, renal failure and systemic vasculitis.

The sclera and cartilage share a common phylogenetic origin. Therefore the presence of scleritis may indicate the severity of underlying connective tissue disorders and vasculitic syndromes³. The development of necrotizing scleritis during the course of a disease such as relapsing polychondritis, may reflect the development of a systemic vasculitic process.

In conclusion, a team approach between the physician and ophthalmologist is essential in the management of such a condition. A prompt referral to the respective specialty will provide early institution of treatment, thus preventing dreadful complications.

References

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