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Sarcoidosis Affecting Bilateral Lacrimal Gland: A not so Common entity in Indian Context

Pathology

KEYWORDS:

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ABSTRACT

Sarcoidosis is a multisystem disease with variable incidence rate throughout the world. It is relatively rare in indian population. In absence of clear cut clinical criteria demonstration of multiple confluent granulomas in histopathology is essential. We here present a case of sarcoidosis involving lacrimal gland in a middle aged woman which is relatively rare presentation of this disease.

Introduction:

Sarcoidosis is a relatively less common entity in Indian population, which is characterised histologically by non caseating granuloma. Sarcoidosis typically affects hilar lymph node, lung and skin, although almost every single organ involvement is reported. Isolated orbital involvement is rare. We hereby describe a case which we encountered at our department with bilateral lacrimal gland involvement in sarcoidosis.

Case report

A 45 year old women presented to the department of ophthalmology of our institution with chief complains of blurred vision, diplopia and mild redness and swelling of bilateral eyelids. The swelling was painless although there was history of sense of fullness of the eyelids. There was no history of trauma, past history of ocular surgery, past or family history of tuberculosis infection. Constitutional symptoms like cough, weight loss, haemoptysis were also absent. She was hypertensive for last 6 years, under medication (calcium channel blocker).

Physical examination showed corrected visual acuity within normal range in both eye, intraocular pressure by applanation tonometry showed normal bilateral intraocular pressure. No afferent pupillary defect in either eye.

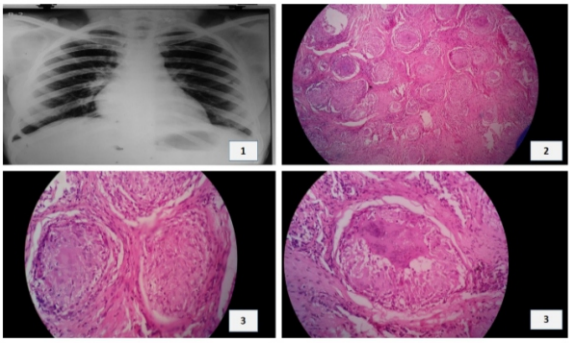
External examination showed mild bilateral upper lid oedema associated with erythema and a nodular mass in the lacrimal gland region.

Anterior orbitotomy was done and biopsy from the lacrimal gland was sent to our department.

On H&E section along with normal lacrimal gland structure there was presence of confluent non caseating granulomas composed of epithelioid cells and scattered multinucleated giant cells, some of which showed presence of round structures with concentric laminations resembling schaumann's bodies. No other organism was found in tissue sections. AFB stain showed no acid fast bacilli. Based on these histopathological findings strong suspicion of Sarcoidosis was made which was correlated with laboratory finding of increased lysozymes, (23 microgram/ml) however serum ACE level was within normal range (30 U/L).

Mantoux test result was also negative. Chest X-Ray did not show any pulmonary involvement.

Based on clinical, laboratory and histopathological findings Sarcoidosis involving bilateral lacrimal gland without systemic involvement was made.



Pic 1: Chest x-ray showing no abnormalities; Pic 2: H&E stained section showing confluent granulomas,

Pic 3: Same area in higher magnification, Pic 4: High power view showing asteroid body within giant cell

Discussion

Sarcoidosis is by definition a systemic disease of unknown cause characterised by non caseating granulomas in many tissues and organs. Exact etiology of Sarcoidosis is still an enigma, it has been postulated that it is a disease of disordered immune regulation in genetically susceptible individuals triggered by environmental factors. Improper cell mediated immune response by CD4+ T cells along with Th1 cell mediated cytokines are the key players in development of sarcoid granulomas against undetermined antigens[1].Histopathological diagnosis of sarcoidosis is basically a diagnosis of exclusion. Although it is defined as a systemic disease ocular involvement may precede systemic Sarcoidosis by years[2]. Sarcoidosis is relatively more common in western countries and exact data regarding its incidence and prevalence in Indian context are unavailable. In their study by Surendra K. Sharma et al. they sought rare clinical presentation of Sarcoidosis in 164 Indian patients & among those 164 cases 19 cases presented with ocular symptoms[3]. According to literature anterior uveitis is the most common ocular manifestation[4]. In their multicentric retrospective study of 26 patients Prabhakaran et al[5] found lacrimal gland involvement in 42.3% cases. Among those cases 54.5% cases were associated with hilar lymphadenopathy & majority of the cases showed raised serum ACE level, serum lysozyme were elevated in only 20 of the cases. In another retrospective study by Halil Yanardag et al[6], 9 out of 516 patients of sarcoidosis had lacrimal gland involvement, and of those 9 cases, 5 cases had normal chest x-ray. Murphy C et al[7] also reported a similar case of lacrimal gland involvement by sarcoidosis.

Summary

Sarcoidosis is relatively rare disease in Indian population and primary lacrimal gland involvement is even rarer. Though histopathological demonstration of non caseating granuloma is hallmark of sarcoidosis, it is a disease of exclusion, especially when lacrimal glands are only involved. Other inflammatory disorders and infective agents like Tuberculosis, atypical mycobacterial infections must also be ruled out.

References

1. Kumar V, Abbas A, Fausto N, Robbins S, Cotran R. Robbins and Cotran pathologic basis of disease. Philadelphia: Elsevier Saunders; 2005.
2. Iannuzzi M. Sarcoidosis. JAMA. 2011;305(4):391.
3. Surendra K. Sharma, Manish Soneja, Abhishek Sharma , Mehar C. Sharma & Smriti Hari. Rare Manifestation Of Sarcoidosis in Modern era of new diagnostic tool. Indian J Med Res 135: 621-9.
4. Obenauf C, Shaw H, Sydnor C, Klintworth G. Sarcoidosis and Its Ophthalmic Manifestations. American Journal of Ophthalmology. 1978;86(5):648-655.
5. Prabhakaran V. Orbital and Adnexal Sarcoidosis. Archives of Ophthalmology. 2007;125(12):1657.
6. Yanardag H and Pamuk ON. Lacrimal gland involvement in sarcoidosis. Swiss Med Wkly 2003; 133:388-391
7. Murphy C, Murgatroyd H. Sarcoidosis of the lacrimal sac--an unusual case. British Journal of Ophthalmology. 2013;97(10):1351-1351.