



Plastics Case Report

Infraorbital adult onset xanthogranuloma associated with eccentric proptosis – A rare case report

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Received : 03 October 2022
Accepted : 20 October 2022
Published : 28 November 2022

DOI
10.25259/LAJO_7_2022

Quick Response Code:



ABSTRACT

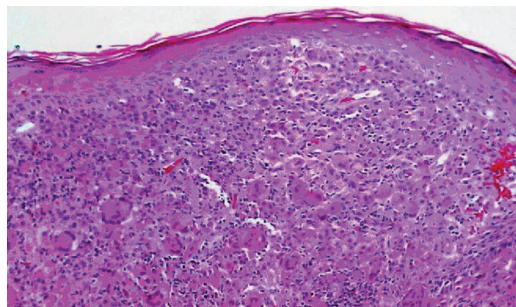
Adult onset xanthogranuloma is one of the very uncommon four varieties of adult xanthogranulomatous disease diagnosed mostly by characteristic histology. The work focuses on excision of the mass in the control of recurrence and associated periocular manifestations. The entity of above case report is very rare and unique and needs proper treatment and careful follow-up to prevent cosmetic disfigurement.

Keywords: Adult onset xanthogranuloma, Histiocytes, Proptosis, Incision, Excision biopsy

INTRODUCTION

Adult onset xanthogranuloma (AOX) is one of the four uncommon entities of AXD confirmed mostly by histologic examination. It is a very rare group of entity with multiple organs such as heart, lungs, bone, retroperitoneum, and orbit/ocular adnexa involvement. The symptoms vary depending on the part of the body involved to exophthalmos, proptosis, visual alteration, HTN, Heart and lung disease, etc.

Mostly AXD has four varieties-adult onset xanthogranuloma,^[1] necrobiotic xanthogranuloma, adult onset asthma and peri-ocular xanthogranuloma and Erdheim-Chester disease. Histopathologically, these varieties are mostly denoted by presence of “Hallmark cells” especially foamy histiocytes,^[2] Touton type giant cells, varying degree of fibrosis. This study focuses on unilateral infraorbital xanthogranuloma and associated recurrence and occurrence after functional endoscopic sinus surgery (FESS).



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CASE REPORT

A 36-year-old male visited the opd with lobulated mass lesion in medial extraconal space of the right orbit with peri-orbital swelling below right lower eyelid with partial decreased vision and restricted extraocular movement on medial side, pain over swelling.

The patient is a known case of undergoing microdebrider FESS twice, with ethmoidal polyp and fungal sinusitis in the right eye. The patient is not known case of diabetes mellitus, hypertension, and asthma.

On magnetic resonance imaging (MRI) paranasal sinuses (PNS) and MRI scan orbit, mass effect is seen over optic nerve which is displaced laterally. Mass effect was also seen over medial rectus muscle. Size of lesion is 6 cm*2.3 cm*3.1 cm. There is extension to medial aspect of pre-septal space and no intra-cranial extension. The mass is hyperintense in anterior part and hypointense on posterior part on T2.

On MRI brain, no significant abnormality was detected; mucosal thickening in the right PNS was seen.

On incision biopsy, histiocytes, multinucleate giant cells, granulomatous inflammatory, and dense collagenous cells were seen.

On microscopy, extensive areas of necrosis were seen with rim of epitheloid granulomatous composed of epitheloid cells, multinucleate giant cells, lymphocytes, and fibroblasts.

Eccentric proptosis was noted from lateral orbital margin to apex of cornea and was noted to be 23 mm. On palpation of orbital margin of lower lid, medial 1/3rd was obscured. Transillumination was absent. Nasolacrimal test showed regurgitation of clear fluid from lower punctum.

On fundus examination, fundic reflex was dull, cup disk ratio within normal limit, and background was normal.

Differential diagnosis

Mucormycosis, encephalocele, hydatid cyst, neurogenic tumor, and lymphoma.

Examination and investigations

Systemic examination

- CVS-S1 n S2 Heard
- P/A -soft, NAD
- R/S-B/L Air entry-Equal
- CNS-conscious and oriented.

Local examination

- Examination of eye-periorbital swelling present in the right eye
- Examination of ear-pre-auricular, pinna, post-auricular region, external auditory canal, tympanic membrane, and middle ear mucosa was normal for both ears
- Examination of nose-skin over root, bridge, dorsum of nose, columella, and vestibule was normal
- Right part of nose showed crust on anterior rhinoscopy.

Examination of throat

Oral cavity-teeth, lip, anterior 2/3rd of tongue, hard and soft palate, and buccal mucosa were normal.

Examination of oropharynx

Anterior pillar, tonsils, posterior pillar, and post pharyngeal wall were normal.

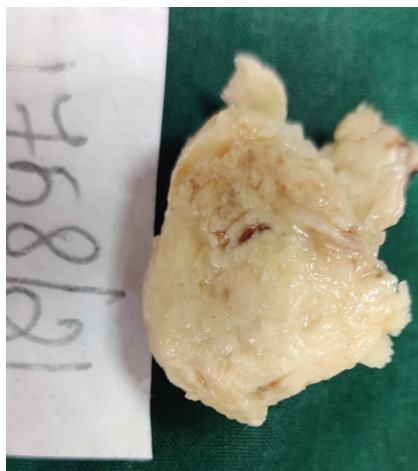
Investigations

- Urea-20
- Creatinine-0.6
- HbsAg and HIV 1,2-negative
- RBS-108 mg/dl
- Hb-14.2 mg/dl
- BT-1 min 30 s
- Computed tomography (CT)-4 min.

Pus culture and sensitivity-culture revealed contaminants

Treatment

- T. Pan 40 mg, tab Augmentin 625 mg, tab para 500 mg was given
- Intralesional triamcinolone, Tab. Zerodol SP, refresh tears were given



Course on hospital

The patient came with complaint of the right infract orbital swelling. The patient started on antibiotics. Condition improved somehow, CT showed mass in orbital space, histology done to reveal xanthogranuloma, and excision done. The patient was discharged.

RESULTS

After the incision biopsy, the yellow sample collected postoperatively, the most probable, and likely diagnosis is infraorbital AOX.

DISCUSSION

Eyelid or ocular adnexa skin lesion can be presented in the 4 syndromes. In 1930, 1st description of disease was done by William Chester and Jacob Erdheim.

Four types are discussed, AOX described as solitary lesion without systemic findings.

Adult-onset asthma and peri-ocular xanthogranuloma (AAPOX) associated with immune dysfunction such as asthma and lymphoproliferative disorders.

Necrobiotic xanthogranuloma is similar to AAPOX.

Erdheim-Chester disease adds systemic involvement as pericardial or pleural effusions, retroperitoneal involvement, diabetic insipidus, hepatosplenomegaly, etc.

CONCLUSION

Xanthogranuloma is a very rare case study topic. The case has been studied due to its rarity and ophthalmic significance. Such cases are also important from cosmetic point of view to prevent recurrence and conserve the eyesight

Acknowledgment

The above study has been acknowledged by all, it is done by great deal of patient care. There is no conflict of interest. All funds have been used by myself and some by patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Das BB. Infraorbital adult onset xanthogranuloma associated with eccentric proptosis – A rare case report. *Lat Am J Ophthalmol* 2022;5:5.