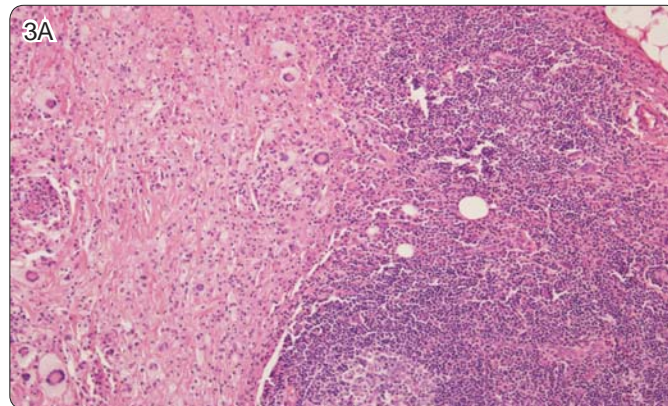


Photoclinic



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A 53-year-old man, a known case of adult onset asthma, presented with bilateral bulging of eyelids (Figure 1A). The lateral rectus muscle of the left eye was thickened on axial CT scan (Figure 1B). Histopathologic evaluation of eyelid and the lateral

rectus muscle showed infiltration of orbital tissue by xanthoma cells (mononuclear foamy histiocytes), dispersed Touton giant cells (giant cells with ring of nuclei around central homogenous eosinophilic zone and a pale foamy area at the periphery) and aggregates of lymphoid cells forming lymphoid follicles (Figure 1C)

Ali Zare-Mirzaie¹, Mohammad Ahangarani Farahani², Gholamhossein Aghaei³, Katayoun Ziari⁴, Moeinadin Safavi¹

Authors' affiliations: ¹Fellow of Molecular Pathology and Cytogenetics, Pathology Department, Medical Faculty, Tehran University of Medical Sciences, Tehran, Iran. ²Pathology Department, Medical Faculty, Iran University of Medical Sciences, Tehran, Iran. ³Ophthalmology Department, Medical Faculty, Iran University of Medical Sciences, Tehran, Iran. ⁴Be'sat Hospital, Pathology Department, Medical Faculty, AJA University of Medical Sciences, Tehran, Iran

•Corresponding author and reprints: Moeinadin Safavi MD, Department of Pathology, Medical Faculty, Tehran University of Medical Sciences, Tehran, Iran. E-mail: safavi_moeinadin@yahoo.com, moein.safavi@gmail.com
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**What is your diagnosis?
See the next page for diagnosis.**

Diagnosis:**Adult Orbital Xanthogranulomatous Disease**

Adult orbital xanthogranulomatous disease is rare and constitutes a group of four heterogeneous syndromes: Adult onset xanthogranuloma (AOX), necrobiotic xanthogranuloma (NBX), adult onset asthma with periocular xanthogranuloma (AAPOX), and Erdheim Chester disease (ECD). It is diagnosed based on the presence of xanthoma cells and Touton giant cells, both of which are considered as hallmark cells in its histopathologic findings. Lymphoid follicles with or without germinal centers are also commonly encountered, especially in adult onset asthma with periocular xanthogranuloma.¹ The latter finding was prominent in the present case and could imply its association with asthma.

AOX presents as an isolated xanthogranulomatous lesion. It has a self-limiting course and does not need aggressive treatment.^{1,2} NBX is a subcutaneous nodule with a tendency for ulceration and fibrosis. It is frequently accompanied by paraproteinemia and multiple myeloma. ECD usually causes a multisystemic

involvement. It is characterized by lymphohistiocytic infiltration along with fibrosis and Touton giant cell formation in heart, lung, retroperitoneum and other tissues. It is a fatal condition; thus, bilateral orbital mass should prompt the clinician to rule out this serious systemic disease. Finally, AAPOX is a rare and recently defined disease whose manifestations include bilateral yellow orange, non-ulcerated xanthomatous eyelids with or without orbital mass, and subsequent adult onset asthma.

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