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# Adult orbital xanthogranuloma – a case report

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## Summary

The periorbital plastic surgery, especially upper and lower blepharoplasty is a very common surgical procedure. Usually the preoperative finding is typical, the surgery routine without unexpected surprises and postoperative course is smooth, quick and without complications. However, periorbital area can also be the source of unexpected findings and peroperative surprises. In this article we present a rare case of a specific disorder – adult onset orbital xantogranuloma in a 37-year-old woman who was treated for recurrences of facial adult orbital xantogranuloma by surgical excisions at the Department of Plastic Surgery, University Hospital Bulovka.

**Key words**

xantogranuloma – orbital xanthogranuloma – histiocytosis – ocular xantogranuloma – giant xantogranuloma

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## Introduction

Adult orbital xantogranuloma (AOX) is present by progressively enlarging, yellowish to reddish-brown lesions of the orbit. The disease is isolated on periorbital area only, without significant systemic involvement. It is the least common one of the four uncommon syndromes called adult orbital xanthogranulomatous disease (AOXD).

AOXD is a very rare and poorly understood heterogeneous group of orbital and ocular adnexal disorders that are diagnosed histologically and classified as class II non-Langerhans cell histiocytosis, characterized by proliferation and accumulation of phagocytosing macrophages (foamy macrophages), Touton-type giant cells and varying degrees of fibrosis [1,2].

## Case description

We describe a case of a patient from the Czech Republic who permanently lives in Luxembourg and at 31 years of age, first presented to our clinic with bilateral eyelid swelling. The first symptoms started 5 years earlier with gradual swelling of both lower eyelids. The patient had no relevant personal and family histories, and presented only with lower eyelids puffiness, diagnosed as an early stage of lower blepharochalasis. The patient underwent surgery; a transconunctival blepharoplasty was performed under local anesthesia. The tissue wasn't subjected to histopathology. No complications were encountered during and after surgery and the patient returned back home to Luxembourg. Two years after the operation, the patient observed gradual swelling of both upper and lower eyelids. The swelling was painless with a yellowish discoloration without other abnormalities of the overlying skin. Two years prior to the periorbital swelling, the patient had developed rhinitis. By her physician, she had been investigated at a department of clinical immunology and allergology. Systemic evaluation revealed an allergic process: rhinitis due to sensitization to gramineous pollens. The patient was treated with corticosteroids with clinical improvement, but every time for a while. At the age of 37, she was referred again due to infiltration of the anterior upper and lower part of both orbits with yellowish plaques, which was so voluminous that it almost took up the closest surroundings and distorted the patient's physiognomy (Fig. 1).

On palpation, they were felt as anterior extensions, firm, painless, slightly lobular tissue. The patient's vision and ocular motility were not affected, although opening the eyes was limited. The rest of her physical examination was unremarkable. The patient underwent upper and lower blepharoplasty. The preseptal fat appeared infiltrated, as did the levator muscle. The resected skin of the upper and lower eyelids was also infiltrated by a firm, white, fibrous mass with minimal vascularity. The operation was accomplished without complications. The tissue was subjected to histopathology. The pathological diagnosis was benign orbital xanthogranuloma, based on the presence of foamy macrophages and Touton giant cells, admixed with chronic inflammatory cellular infiltrate such as lymphocytes, plasma cells and cholesterol. The Touton giant cells are multinucleate cells with the nuclei arranged in a wreath

**Fig. 1. Clinical picture of the patient before surgery with bilateral yellow, elevated, indurated xanthomatous eyelids and/or orbital masses.**

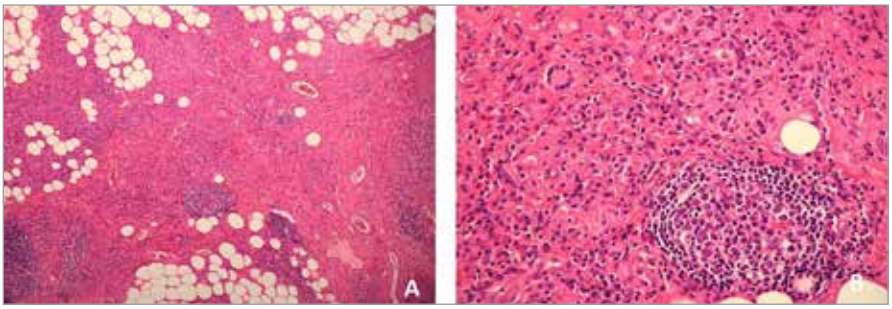


**Fig. 2. Diffuse infiltrate rich in lymphocytes, foamy histiocytes, and giant cells. A) Nodular lymphoid infiltrate; B) Touton-type giant cells.**

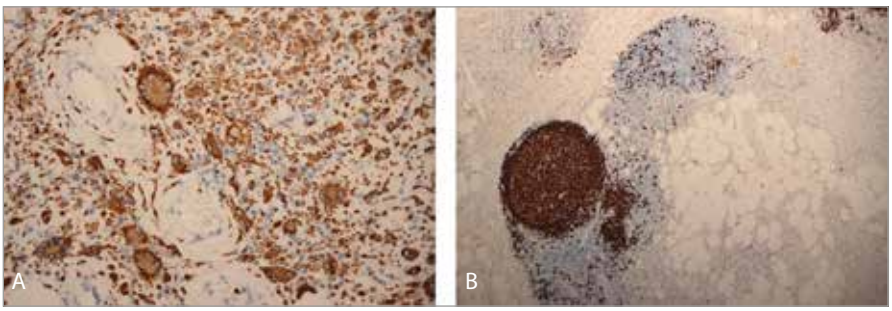


**Fig. 3. Immunohistochemically, the foamy histiocytes positive for A) CD68; B) CD20.**

**Fig. 4. Result 1 year after operation.**

around a nidus of eosinophilic cytoplasm and separated from the cell membrane by a rim of translucent foamy cytoplasm (Fig. 2).

Immunohistochemically, the foamy histiocytes are strongly positive for CD68, CD 20, CD3, LCA, and CD 138 (Fig. 3).

In our patient, the final diagnosis was adult onset orbital xanthogranuloma. No other abnormities have been shown in this patient. After surgery and diagnosis statement the patient underwent complete medical examination – internal, allergological, and immunological with no pathology found and the conclusion of all specialist confirmed a histopathological diagnosis of periorbital xantogranuloma. One year after surgery, the patient is without any treatment, currently pregnant. Locally, only the left side lower eyelid shows gentle abundancy and elevation that will probably require another surgical correction (Fig. 4). Appropriate treatments for this condition included long-term closed follow-up for early detection of systemic involvements. Topic or systematic treatment with corticosteroids can be considered, but with all negative consequences. Repetitive surgical interventions are possible, though with increasing risks of complications.

## Discussion

In 1987, the Histiocyte Society Writing Group classified histiocytic disorders into three categories: class I (Langerhans cell histiocytosis – histiocytosis X spectrum), class II (histiocytoses of mononuclear phagocytes other than Langerhans cells), and class III (malignant histiocytic disorders) [3]. Adult xanthogranulomatous disease, which falls in class II, is an uncommon group of unknown etiology and pathogenesis affecting the skin and subcutaneous tissues of the periorbital areas and the ocular adnexa. Pathologically, they typically demonstrate an abundance of foamy histiocytes and Touton giant cells. It affects patients from 17 to 85 years of age with no significant gender preference [1]. AOXD is thought to

be caused by a stimulating agent that induces proliferation of histiocytes [2], although the nature of this stimulus is currently unknown.

Depending on clinical manifestations and characteristics, adult periorbital xanthogranulomatous disease comprises four forms: adult onset orbital xanthogranuloma, adult onset asthma and periocular xanthogranuloma, necrobiotic xanthogranuloma, and Erdheim-Chester disease (Tab. 1) [4–6].

Adult onset xanthogranuloma (AOX) is an isolated xanthogranulomatous lesion without systemic involvement. It is the rarest of the xanthogranulomatous lesions, with unknown etiology and incidence. AOX presents as soft yellow-brownish subcutaneous tumours of different sizes, mostly as solitary lesions, but also as multiple lesions with predilection areas on the face and the neck. It is often self-limiting; and no aggressive treatment is required. The diagnosis is made by biopsy of the lesion. Our patient had a periorbital disease without systemic involvement. Ulceration or ocular inflammation had never occurred in 8 years and no monoclonal B-cell abnormality could be observed. For those reasons we classified the patient as suffering from adult onset orbital xanthogranuloma. The treatment is usually empirical as the mechanisms in xanthogranulomatous disorders are poorly understood, and there is no information on the genetic basis of the disease. Intralesional corticosteroids (triamcinolone acetonide 40 mg/mL) have also been used to treat AOX, although they are less efficacious than systemic corticosteroids [7]. In systemic disease, corticosteroids are necessary. Progression during the therapy with corticosteroids would warrant cytostatic treatment [8]. Radiotherapy has been administered in recurrent cases, but it is considered empirical. Although exacerbation of cutaneous lesions after treatment has been reported [9], various treatment modalities have been tried including local excision, radiotherapy, intralesional corticosteroids, interferon alpha, plasmapheresis, extracorporeal photopheresis, laser therapy, radiotherapy, and psoralen plus ultraviolet A photochemotherapy [10–12]. In our case we completed the therapy by the excision of abundant mass as much as the diagnosis was established retrospectively by histopathological examination. The patient was informed about the diagnosis and prognosis. Long-term follow-up is mandatory.

**Tab. 1. Differential diagnosis of adult periorbital xanthogranulomatous disease [1,3].**

| Clinical form | Clinical features and systemic associations | Evaluation | Prognosis |
|---|---|---|---|
| adult onset orbital xanthogranuloma | localized to the eye (anterior orbit) | histopathological | good; limited to the eye |
| adult onset asthma and periocular xanthogranuloma | anterior orbit, asthma and lymphadenopathy | perform lung function testing | good; rarely associated with lung disorder |
| necrobiotic xanthogranuloma | anterior orbit ulceration, multiple myeloma, lymphoma, paraproteinemia | rule out paraproteinemia | poor; systemic lymphoproliferative disorders |
| Erdheim-Chester disease | intraconal, long bone sclerolytic destruction, retroperitoneal fibrosis, cardiac involvement | perform extension study | poor; mortality rate is increased |

## Conclusion

Adult onset xanthogranuloma is the most benign and rarest form from the four subtypes of adult orbital xanthogranulomatous disease – heterogeneous group of orbital and ocular adnexal disorders that are classified as class II non-Langerhans histiocytic proliferation, clinically presenting with progressively enlarging yellowish lesions of the orbit with unknown pathogenesis. Our case is the first reported case of adult onset xanthogranuloma without any systemic associations in the Czech Republic. Our experience emphasised that the diagnosis of xanthogranuloma may be considered in a patient with proptosis associated with periocular yellowish cutaneous plaques, histologically established by the presence of an inflammatory infiltrate of foam cells and Touton-type multinucleated giant cells. The prognosis of AOX is excellent, without extracutaneous manifestations [1]. Nevertheless, long-term follow-up is necessary to determine systemic involvement early.

**Roles of authors**

Elbek Rashidov – main author, preparation of manuscript

Madina Babakalanova – review of the literature, preparation of manuscript

Martin Molitor – treating doctor, preparation of manuscript, review of literature

Zuzana Špůrková – histopathological examination, diagnosis statement

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