Management of Adult-onset Eyelid Xanthogranuloma: Debulking Surgery Coupled with Intralesional Corticosteroids

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ABSTRACT

Introduction: Adult-onset xanthogranuloma (AOX) is a rare, benign inflammatory disorder within the spectrum of non-Langerhans cell histiocytosis, often manifesting as localised skin lesions in middle-aged adults and frequently misdiagnosed due to its nonspecific presentation.

Case: A 43-year-old woman developed a painless, progressively enlarging mass on her right upper eyelid over four months, with histopathology confirming AOX.

Observation: Following surgical debulking and adjunctive intralesional corticosteroid (triamcinolone acetonide) injection, the patient exhibited significant clinical improvement and no recurrence at three months. This outcome supports emerging evidence that a combination of surgery and corticosteroid therapy effectively reduces lesion size and recurrence rates in localised AOX.

Conclusion: The multimodal approach combining surgical excision and intralesional corticosteroid injections provides a conservative and effective management strategy for AOX, with favourable outcomes and minimal complications. Further research is warranted to standardise treatment protocols for this rare orbital disease.

Key words: Adult onset xanthogranuloma; intralesional corticosteroids; non-langerhans cell histiocytosis; surgical debulking.

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INTRODUCTION

Adult orbital xanthogranulomatous disease (AOXD) is a rare spectrum of non-Langerhans cell histiocytic illnesses that mostly affect orbit and associated ocular tissues. Adult-onset xanthogranuloma (AOX), adult-onset asthma with periocular xanthogranuloma (AAPOX), necrobiotic xanthogranuloma (NXG), and Erdheim-Chester disease (ECD) are four primary subgroups of this disorder (Sivak-Callcott et al., 2006; Kiratli et al., 2015; Detiger et al., 2023). While each subtype has unique clinical, histopathological, and systemic characteristics, they share common features such as the existence of Touton large cells, foamy histiocytes, and varied levels of lymphocytic infiltration and fibrosis. Due to their rarity and overlapping clinical presentations, these conditions are often challenging to diagnose and manage effectively (Chicas Sett et al., 2016).

The least prevalent of these subtypes is AOX and typically presents as isolated orbital or eyelid lesions without systemic involvement. Accumulation of multinucleated giant cells and lipid-laden macrophages is AOX characteristic, often resulting in painless, slowly progressive masses (Kiratli et al., 2015). Its nonspecific symptoms and rarity frequently lead to misdiagnosis, making its management a significant clinical challenge. While surgical excision remains primary treatment, recurrence is common, prompting exploration of adjunctive therapies such as intralesional corticosteroids, which have shown promise in reducing lesion size and preventing relapse (Chicas Sett et al., 2016; Knani et al., 2023).

This case highlights the critical importance of multimodal approach in management of AOXD, with particular emphasis on synergistic role of combined surgical and medical therapies in achieving optimal outcomes. Integration of intralesional corticosteroids as adjunct to surgery provides conservative yet highly effective treatment strategy for localised AOX lesions, as evidenced by the success observed in this case. Furthermore, this work contributes to existing body of literature on this rare disease, underscoring the need for heightened awareness, early diagnosis, and the establishment of standardised treatment protocols to enhance patient outcomes.

CASE REPORT

A 43-year-old woman appeared with a progressively growing, painless mass located in the right upper eyelid (central third), present for four months (Figure 1). The lesion was first noted four months before presentation as a maize-seed–sized nodule and progressively enlarged over the subsequent four months to its current size, with associated mild pruritus. The patient denied any associated pain, visual disturbances, redness, excessive tearing, or discharge. Her past medical history was unremarkable for systemic conditions such as hypertension or diabetes mellitus. However, she reported a history of ocular trauma due to a wooden splinter injury to the right eye four years prior.

On examination, her right eyelid was enlarged. Eversion of the eyelids showed a uniform, smooth mass that was roughly 20 mm by 35 mm by 17 mm. When palpated, the lump felt rubbery and hard rather than soft. In both eyes, her best corrected visual acuity was 20/20, and her

intraocular pressure was within normal limits. In primary gaze, the vertical palpebral fissure height was 3 mm in the right eye compared with 11 mm in the left eye, consistent with severe right upper eyelid ptosis. Her pupils were equal and responsive to light, and there was no indication of a RAPD. She also possessed a full and equal range of extraocular muscle activity. She was afebrile and normotensive.

A computed tomography (CT) scan of the orbits revealed thickness in the superior and inferior eyelids on the right side, with mild enlargement of the lacrimal gland (Figure 2). These findings suggested an underlying inflammatory process rather than a neoplastic lesion.

The patient underwent anterior orbitotomy through an upper eyelid crease incision under general anesthesia, followed by subtotal tumour excision (debulking). Intraoperatively, a yellowish, firm, lobulated xanthogranulomatous mass was identified predominantly within the right upper eyelid/anterior orbital compartment, extending through the subcutaneous and orbicularis plane with adherence to adjacent eyelid structures. The lesion's margins were ill-defined, and therefore complete excision was not attempted to avoid functional compromise of the eyelid; a debulking (subtotal excision) was performed until adequate decompression and safe planes were achieved. No gross involvement of the globe or extraocular muscles was noted. The excised tissue was sent for histopathological examination, and a gross specimen photograph is provided (Figure 3).

Histopathological examination of the excised tissue was consistent with adult-onset xanthogranuloma, characterised by a xanthogranulomatous infiltrate with foamy histiocytes and Touton-type giant cells, without evidence of a neurofibromatous lesion (Figure 4).

The patient's initial post-operative course was uneventful, with mild eyelid oedema and hyperaemia but no significant pain (Figure 5). Over the subsequent weeks, progressive improvement was observed, with minimal haematoma formation and resolution of eyelid swelling; however, minimal signs of inflammation were still present.

On post-operative day 16, intralesional triamcinolone acetonide (TA) injection was administered to reduce inflammation and prevent recurrence (Figure 5). Two intralesional corticosteroid injections were administered at 2-month intervals. At each session, triamcinolone acetonide 40 mg (total 1 mL of 40 mg/mL) was injected using a 25-gauge needle into the subcutaneous/preseptal plane of the right upper eyelid, using a multi-point technique to achieve uniform distribution within the affected area. The patient exhibited significant improvement, with increased interpalpebral fissure width from 3/11 mm preoperatively to 11/11 mm post-treatment. No complications, such as ocular hypertension or skin discoloration, were observed.

At the three-month follow-up after the second TA injection, there was no clinical evidence of lesion recurrence, and the eyelid had returned to a normal appearance (Figure 5). This case highlights the efficacy of intralesional corticosteroid therapy in managing adult-onset eyelid xanthogranuloma, notably in cases where total surgical excision is not possible because to significant tissue involvement.

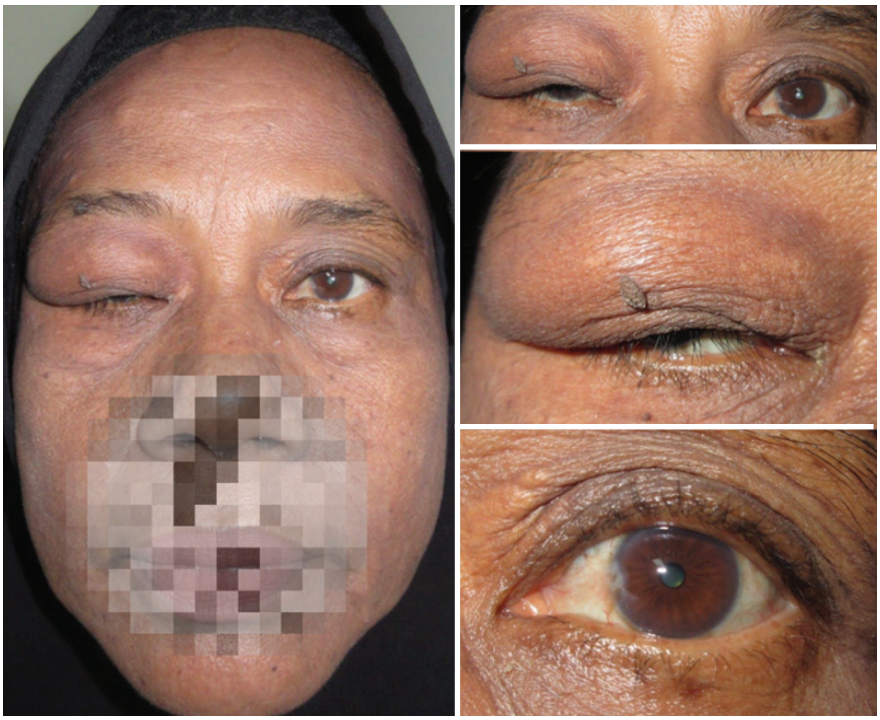

Figure 1: First clinical view of patient.

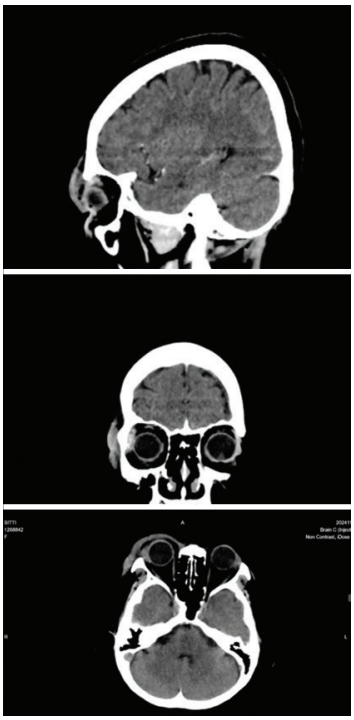

Figure 2: Contrast multislice computed tomography (MSCT) brain examination.

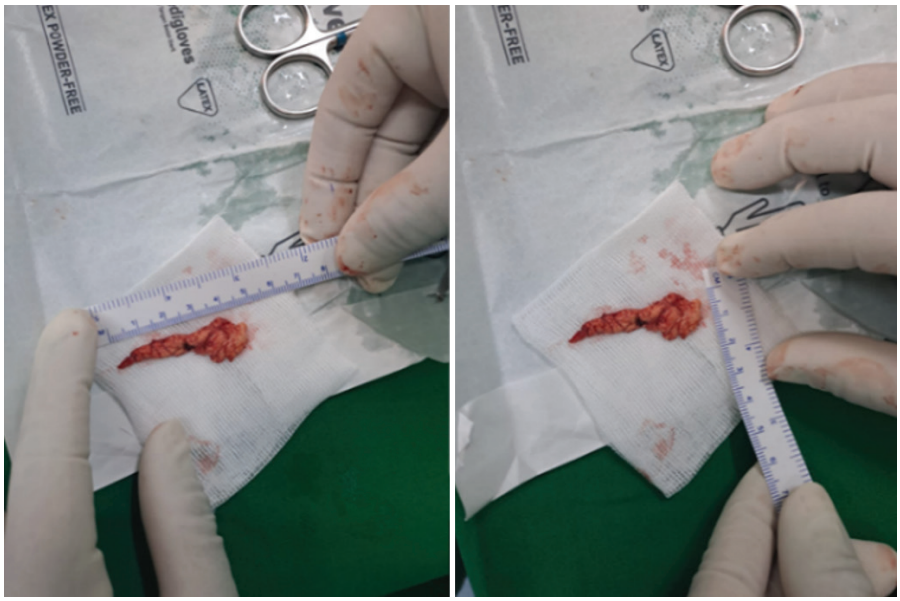

Figure 3: Tumour excision results.

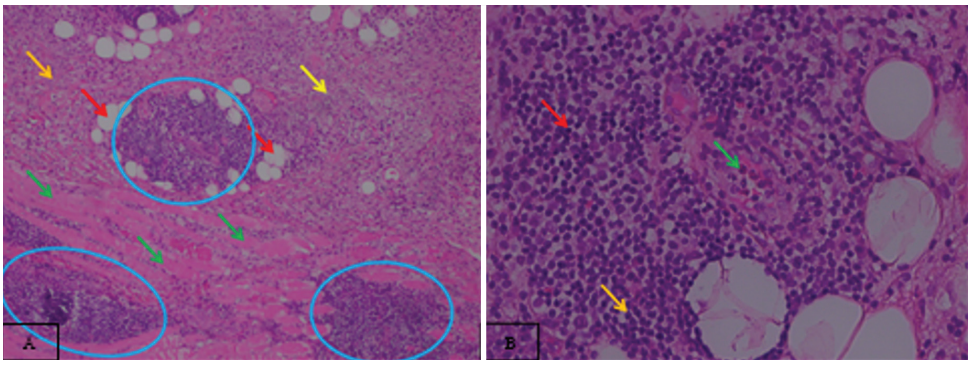

Figure 4: Histopathological Examination (Haematoxylin and Eosin stain (HandE)). A. 10x Magnification: In the tissue sample, xanthoma cells (blue circle) were mixed with lymphocytes, plasma cells, and neutrophils and invaded connective tissue (yellow arrow), adipose tissue (red arrow), and muscle tissue (green arrow). There was no tumour mass with characteristics of neurofibroma. B. 40X Magnification: The tissue sample revealed xanthoma cells infiltrating muscle, adipose, and connective tissue, along with neutrophils (green arrow), lymphocytes (red arrow), and plasma cells (yellow arrow). There was no tumour mass with characteristics of neurofibroma.

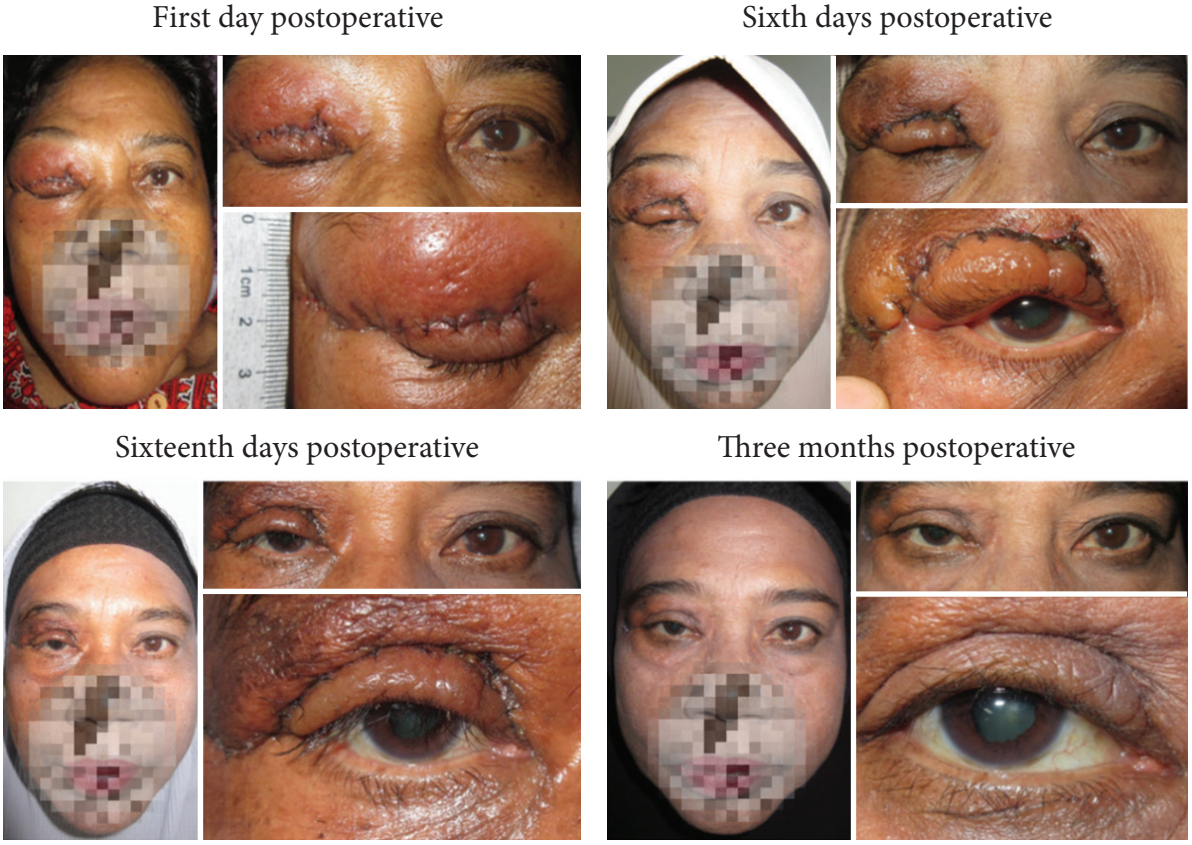

Figure 5: Post-operative clinical views showing the healing process following surgical intervention for ocular surgery.

DISCUSSION

The AOX is a localised entity within the spectrum of adult orbital xanthogranulomatous disease. Because these disorders can overlap clinically, diagnosis relies on histopathologic confirmation of xanthogranulomatous inflammation (foamy histiocytes with Touton-type giant cells) integrated with clinico-radiologic correlation and evaluation for systemic involvement. Management remains empiric and individualised, as no single regimen has been shown to be consistently superior; reported approaches include observation in limited disease, local and/or systemic corticosteroids, steroid-sparing immunosuppressive agents, radiotherapy, and surgical excision or debulking—often in combination depending on extent and systemic associations (Sivak-Callcott et al., 2006; Satchi et al., 2014; Topal et al., 2016).

In the case presented, a 43-year-old female developed a solitary xanthogranuloma on her right upper eyelid, consistent with a diagnosis of AOX. The lesion was initially painless and exhibited a slow, progressive enlargement over a four-month period. Clinical examination revealed a firm and rubbery mass, measuring approximately 20 mm × 35 mm × 17 mm, without any associated visual disturbances or systemic symptoms.

Histopathological evaluation of the excised tissue demonstrated a typical invasion of foamy histiocytes and Touton large cells. In addition, the lesion had an admixture of lymphocytes and plasma cells, confirming the diagnosis of AOX. Immunohistochemical staining was instrumental in excluding other histiocytic disorders, as negative results for CD1a and S100 ruled out Langerhans cell histiocytosis (Burris et al., 2016; Detiger et al., 2023).

Management strategies for AOXD are tailored according to the specific subtype and disease extent (Chicas Sett et al., 2016). In this case, the patient initially underwent a surgical debulking procedure via orbitotomy. Recognising the potential for recurrence, the surgical intervention was complemented with intralesional corticosteroid injections, specifically triamcinolone acetonide, to further control the lesion (Knani et al., 2023).

Recent studies have underscored the efficacy of combining surgical excision with adjunct intralesional corticosteroid therapy in managing localised lesions of AOX (Elner et al., 2006; Burris et al., 2016; Miguel et al., 2017; Knani et al., 2023). This approach aims to minimise recurrence while preserving function and aesthetics. In this case, the patient received two intralesional injections at specified intervals, which resulted in a marked reduction in lesion size and alleviation of symptoms.

Notably, the combined treatment strategy yielded a highly favourable outcome, as evidenced by the substantial improvement in eyelid function and cosmesis (Detiger et al., 2023; Knani et al., 2023). The patient exhibited no significant complications such as ocular hypertension or skin pigmentation changes, and follow-up evaluations confirmed the absence of lesion recurrence three months after the second injection.

Differential diagnosis for orbital xanthogranulomatous disease includes other

histiocytic conditions such as Langerhans cell histiocytosis, Rosai-Dorfman disease, and IgG4-related disease (Detiger et al., 2023). Therefore, accurate classification relies on careful integration of clinical context with histopathological evaluation and, when required, immunohistochemical profiling, as these conditions may overlap clinically but differ in prognosis and management.

In the present case, histopathology demonstrated a predominantly xanthogranulomatous infiltrate composed of mononuclear foamy histiocytes (xanthoma cells) with frequent Touton-type multinucleated giant cells. The infiltrate extended into connective tissue, adipose tissue, and skeletal muscle, accompanied by a mixed inflammatory background including lymphocytes, plasma cells, and neutrophils. At higher magnification, Touton giant cells showed the characteristic wreath-like nuclear arrangement with abundant lipidised cytoplasm. Importantly, no morphological features suggestive of a neurofibromatous tumour were identified, supporting the diagnosis of adult-onset xanthogranuloma.

This case highlights key clinical and histologic features of AOX and demonstrates favourable short-term response to a combined approach of surgical debulking and intralesional corticosteroid injection. Debulking was preferred because complete excision was not feasible without undue risk to eyelid/orbital structures and allowed both symptom reduction and definitive tissue diagnosis. Intralesional corticosteroid was used as an adjunct to target residual localised inflammatory disease while minimising systemic corticosteroid exposure. However, recurrence after surgical management—particularly when complete excision is not achievable—has been reported, and may occur months to years after treatment; therefore, our 3-month follow-up should be interpreted as an early outcome, and longer clinical (and imaging when indicated) surveillance is required to confirm durable disease control.

CONCLUSION

Adult-onset xanthogranuloma of the eyelid is a rare condition that should be considered when diagnosing orbital and eyelid tumours in adults. The management of these lesions can be challenging, and a multidisciplinary approach involving ophthalmologists, dermatologists, and pathologists is often necessary. The combination of surgical debulking and intralesional corticosteroids, as demonstrated in this case, offers a promising treatment strategy for localised AOX lesions. Additional study is required to better understand the pathophysiology of these disorders and to develop consistent treatment methods.

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