Adult-onset Xanthogranuloma

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The outlines

- The case
- Introduction
- Discussion
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Conclusion
The case

History

• A 44-year-old man.
• C/O multiple bilateral upper and lower eyelid lesions, started 7 years ago.
• Progressive in number and size over two years with no other associated symptoms.
• Allergic rhinitis
• No relevant ocular, social, or family history.
The case, cont.

**Examination**

- His vital signs were within normal limits.
- Visual acuity was 20/20 OU with no relative afferent pupillary defect.
- There was no restriction of ocular motility in all fields of gaze.
- There was no proptosis in either eye.
- Multiple yellowish to tan-colored raised cutaneous lesions that are variable in size were seen bilaterally on the patient’s lower eyelids.
- There were no clinical signs of an infectious or malignant process.
Clinical photograph at presentation
The case, cont.

**Diagnosis:**

- The clinical diagnosis was: *Xanthelasma palpebrarum-like lesions.*
- The patient was booked for excision of these lesions with full thickness skin graft under local anesthesia.
- The procedure went smoothly with no complications.
One week post excision with skin grafting

The case, cont.
The case, cont.

- **Histopathology**

  - Foamy macrophages & many Touton giant cells
  - **+ve:** CD68 and factor XIIIa.
  - **-ve:** CD34, S100 and CD1a.
  - No evidence of malignancy.
  - Consistent with:

    adult orbital xanthogranulomatous disease
The case, cont.

Thorough blood investigations were carried out along with Magnetic Resonance Imaging (MRI) with gadolinium contrast.

Results were within normal limits.

Dx: adult-onset xanthogranuloma (AOX)
The case, cont.

Follow up

The patient came 1 year later in the following visits with recurrent lesions bilaterally. Lesions were excised again and skin was left to heal by secondary intention.
Follow up

Recurrence one year after first surgery
Follow up

3 months post second excision only with no skin grafting (healing by granulation)
Follow up

Six months later, he presented with new small eyelid lesions. Intralesional Triamcisenolone Acetonide 20 mg/ml was injected in both upper and lower lids two times separated by three months.
Introduction

Adult Orbital Xanthogranulomatous Disease (AOXGD) is a rare group of orbital diseases that are classified as class II: Histiocytoses of mononuclear phagocytes other than Langerhans cells, in the classification proposed in 1987.¹

A recent classification published in 2016 based on histology, phenotype, molecular alterations, clinical and imaging characteristics was proposed. AOXGD’s fall in 2 groups, the “L” group and the “C” group. ²
Introduction, cont.

Four entities are listed under the umbrella of AOXGD based on the presence of other associated systemic conditions.²⁻⁴ Some of which can be fatal.⁵

1. Adult-onset asthma and periocular xanthogranuloma (AAPOX)
2. Necrobiotic xanthogranuloma (NXG)
3. Erdheim–Chester disease (ECD)
4. Adult-onset xanthogranuloma (AOX) being the least common.²
Discussion

**Pathogenesis**

- Reactive proliferation of the free tissue macrophages.
- The cause is not known currently.
- One report described a case of xanthogranuloma of the salivary gland in a 3-month-old infant presenting with cytomegalovirus (CMV) infection.\(^6\)
- Another study confirmed the presence of cytomegalovirus antigens immunohistochemically in a patient presenting with oral juvenile xanthogranuloma.\(^7\)
- A viral exposure is suggested to be a possible mechanism in the pathogenesis of xanthogranulomatous disease.
Discussion

Clinical presentation

There are overlapping features between these conditions and diagnosis of a single specific entity can be difficult initially.\(^4\)

However, there are some distinct dissimilarities in their clinical, histopathological, associated systemic features, the laboratory findings and the prognosis.
### Systemic associations

<table>
<thead>
<tr>
<th>Condition</th>
<th>Associated Conditions</th>
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<tbody>
<tr>
<td>Adult-onset xanthogranuloma</td>
<td>Typically none</td>
</tr>
<tr>
<td>Necrobiotic xanthogranuloma</td>
<td>Paraproteinemia (IgG monoclonal gammopathy), multiple myeloma.</td>
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<tr>
<td>Erdheim–Chester disease</td>
<td>The histiocytic proliferations seen occur mainly in the long bones of the lower extremities, the central nervous system, retroperitoneum, lung, heart, liver, spleen, skin, and the orbit.</td>
</tr>
<tr>
<td>Adult onset with asthma</td>
<td>Lymphadenopathy, paraproteinemia. chronic lymphocytic leukemia/ small lymphocytic lymphoma, multiple myeloma, and non-Hodgkin lymphoma.</td>
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Treatment

Because the mechanisms are poorly understood, to date there is no targeted therapy directed at the histiocytic proliferation.

Modalities:
- Intrallesional corticosteroids
- Systemic corticosteroids
- Debulking surgery
- High-dose corticosteroids with low-dose radiotherapy
- Methotrexate, interferon-alpha, cyclophosphamide, doxorubicin, vincristine and others.
Another case

Journal of Clinical Case Studies

Periocular Adult-Onset Xanthogranuloma (AOX) Initially Misdiagnosed as Xanthelasma: A Case Report

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Another case

A 54-year-old Saudi lady

Persistent multiple yellowish skin lesions involving all 4 eyelids, for 4 years

History of hyperlipidemia on treatment

Otherwise healthy

History of excisional biopsy elsewhere with the histopathological diagnosis of xanthelasma

Another case
Histopathology 1

- Initial skin lesions at our center were also diagnosed as xanthelasma
Histopathology 2

Further excisional biopsy of the skin lesions of the right lateral canthal area and the deep orbital component.

Sheets of foamy histiocytes with few aggregates of lymphocytes, some forming germinal centers.
Further excisional biopsy of the skin lesions of the right lateral canthal area and the deep orbital component.

Sheets of foamy histiocytes with few aggregates of lymphocytes, some forming germinal centers.

Giant cells including Touton giant cells.

Significant fibrosis was evident.

No evidence of necrobiosis.
The xanthoma cells showed positive staining with CD68 while they were negative to S100 and CD1a.
Impression

• AOXD

• Further evaluation medically to rule out asthma and/or systemic associations was done.

• AOX
Conclusion and take home message

- AOXGD’s are rare.
- Recognition is important.
- Treatment options vary.
- Prognosis ?
Thank YOU
References