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Background

Xanthogranulomas are the most common form of non-Langerhans cell histiocytosis.¹ The lesions are considered benign and frequently are self-limiting. They can present in childhood as well as in adulthood.¹ In adults with multiple lesions that erupt as progressive yellow-orange papules and plaques, Multiple Adult Xanthogranuloma (MAXG) should be considered, and monitoring is advised due to the occasional association with haematological malignancies.²

Patient History

- 88-year-old female patient.
- History of presenting complaint:
 - 8-year history of localized facial rash with slow progression.
 - No known triggers.
 - Occasional tenderness.
- Past medical history of eczema.
- Non-smoker.
- Minimal alcohol use.
- History of excessive sun exposure.

Prior Treatment:

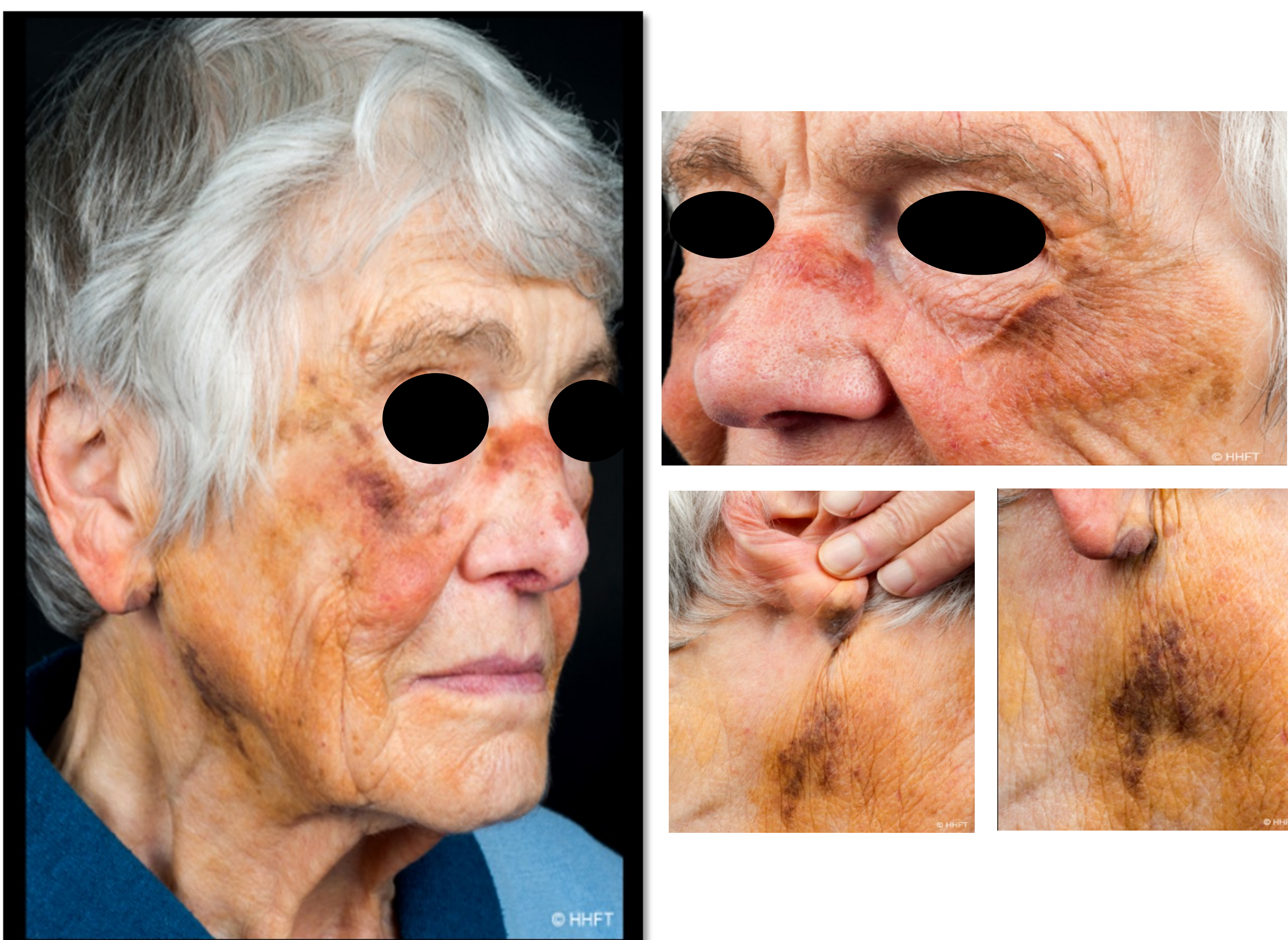
- Multiple previous topical treatments:
 - Topical steroids (Hydrocortisone 1%, Clobetasol butyrate 0.05% and Betamethasone valerate 1 in 4).
 - Emollients

These showed no improvement.

Examination:

- Bilateral, diffuse, indurated red, brown and yellow plaques.
- Affected areas: cheeks, lateral upper eyelids, bridge of nose, jaw, upper neck and right ear lobe.
- There was no change in skin sensation.

Patient Images



Results

- Serum proteins (total – globulins), reticulocytes, electrophoreses, IgG, IgA, IgM, U&Es, LFTs, CRP, calcium, bone profile, FBC, ACE, iron, transferrin, gamma-GT, LDH as well as connective tissue screen were all unremarkable.
- Lipids were slightly raised:
 - Cholesterol: 6.5mmol/L (nv: 0-5.2).
- B12: slightly raised at 1110ng/L (nv: 211-911).

Immunohistochemistry

- Scattered spindle cells:
 - Positive for factor XIIIa.
 - Negative for S100 and CD34.
 - Foamy histiocytes and multinucleated giant cells are highlighted by CD68.

Histology

Two punch biopsies were taken from the Rt cheek and Rt earlobe.

Epidermis:

- Orthokeratotic epidermis.
- Increased basal cell layer pigmentation.

Dermis:

- Sheets of foamy histiocytes.
- Touton giant cells (figure 2a).
- Mixed inflammatory infiltrate rich in lymphocytes and pigment laden macrophages (figure 2b).

Results within the spectrum of juvenile xanthogranuloma. No features of necrobiotic xanthogranuloma.

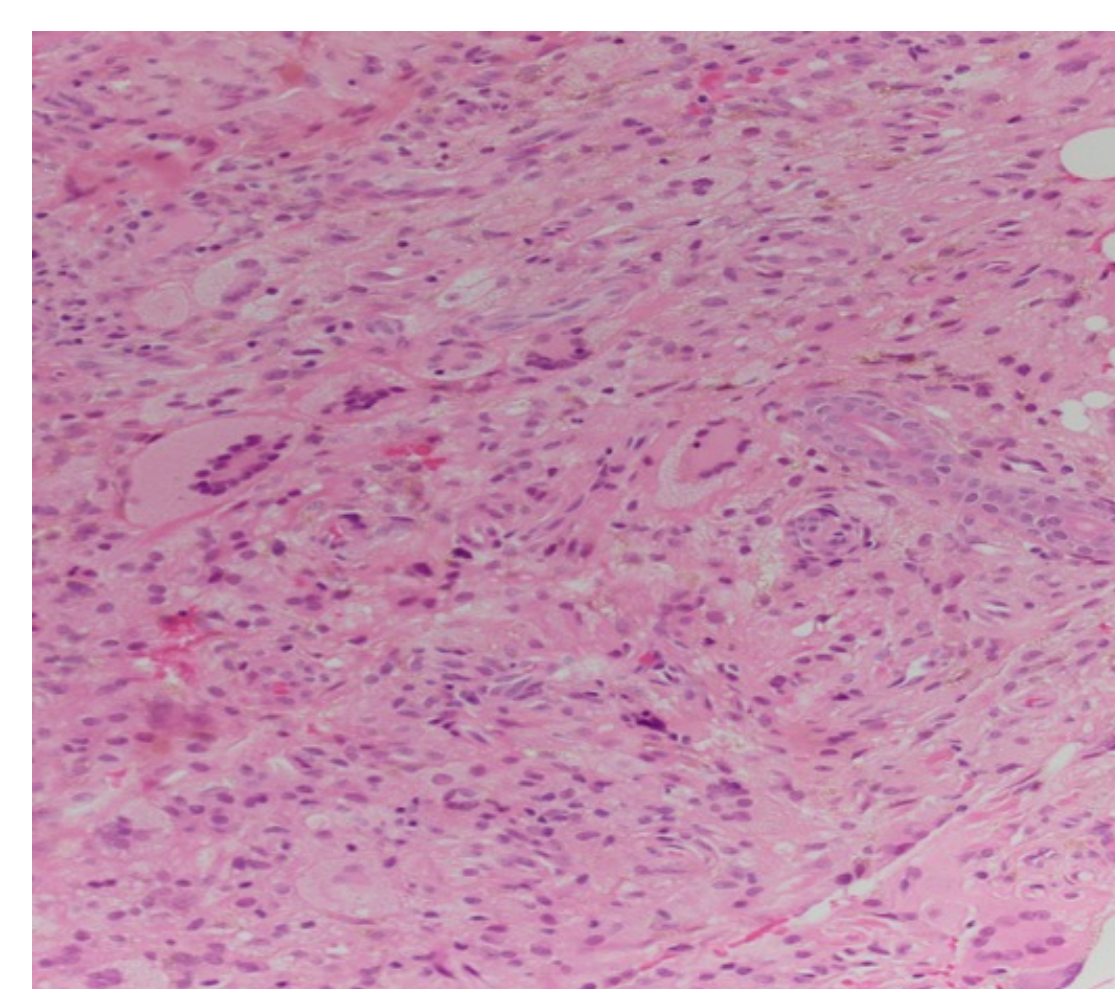


Figure 2a

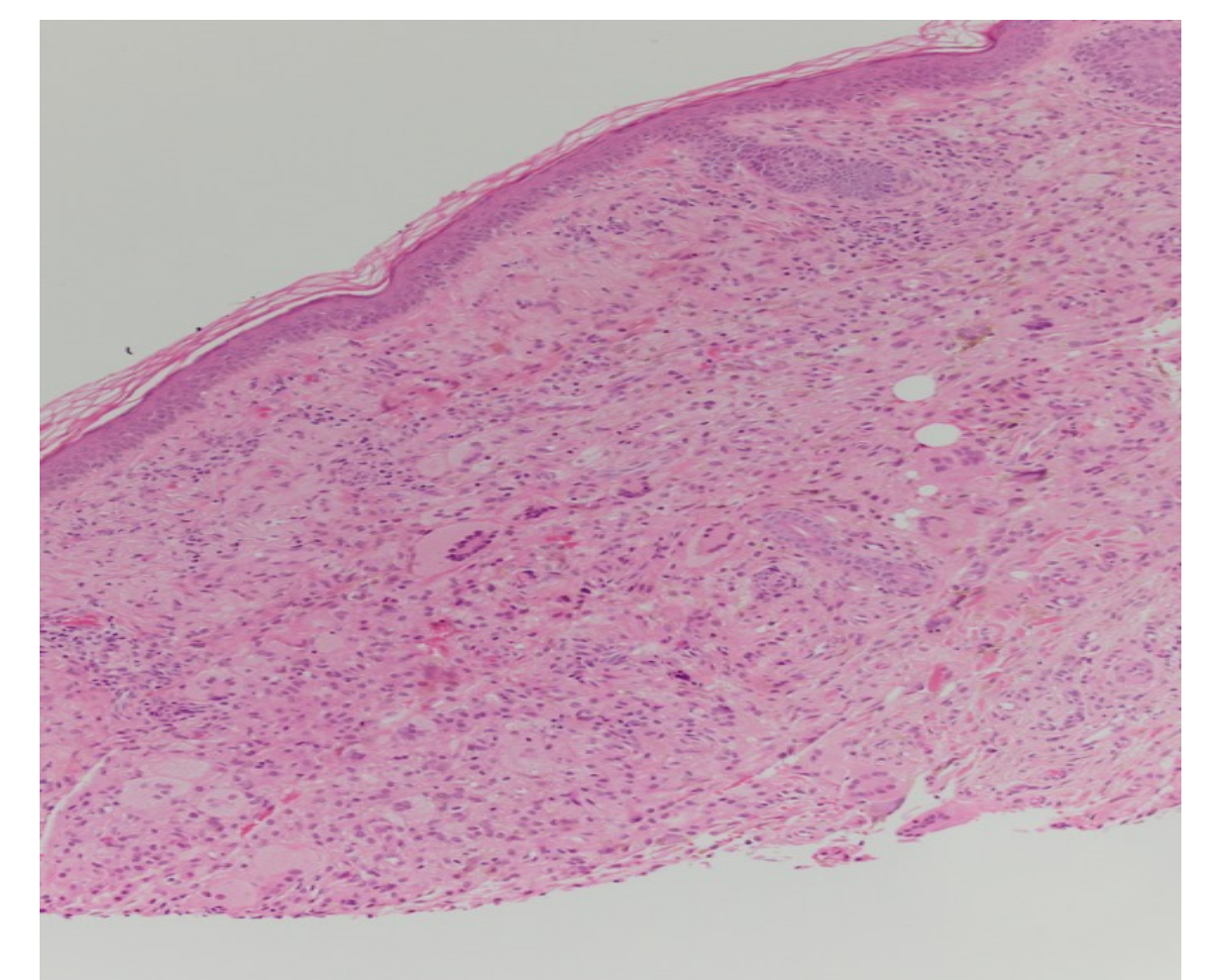


Figure 2b

Conclusion

This case is atypical due to its late onset and diffuse presentation, posing a differential diagnostic challenge alongside limited treatment options. Although, clinically the rash presented with plaques as would be seen in xanthoma, histologically, the findings were more consistent with xanthogranuloma. This case had factor XIIIa positive cells on IHC which would be negative for xanthoma. There was also a good number of touton giant cells which is not typical for xanthoma. Despite multiple investigations, no association with haematological malignancy was found; however, continued monitoring is advised.

References

1. Sun L, Cláudia Brazão, Mancha D, Luís Soares-de-Almeida, Tapadinhas C, Filipe P. Multiple adult xanthogranuloma as the first sign of chronic myelomonocytic leukaemia. *Clinical and Experimental Dermatology*.
2. Tan L, Aw C. Unusual presentation of adult xanthogranuloma. *Singapore Medical Journal* 2014 Feb;55(22): e25-7