

## Image Article

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## Erdheim- Chester Disease (Non-Langerhans Cell Histiocytosis)

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52 years old female with recurrent admission for myalgias, inflammatory arthritis, rash around the eyes and abdomen (A, B). Routine blood tests were either normal or negative apart from low C4 level. Pan CT of the body was unremarkable.

Biopsy from left medial canthus skin shows xanthoma cells occupying the dermis, few lymphocytes, no plasma cell infiltrates (negative CD138 stain) no eosinophils, and no Langerhans cells(D). Foamy cells with finely granular cytoplasm, positive for CD 168 and CD 163. The lesions have the appearance of xanthoma rather than xanthogranuloma (C). Skin biopsy from

right shin was consistent with dermatofibroma. A punch biopsy from the left arm showed eosinophilic histiocytoid and epithelioid dermatofibroma stained positively for CD68 and negative for S100/Langerhin. Biopsy from the abdomen was consistent with angiokeratoma. Patient was diagnosed as multifocal non-Langerhans cell histiocytosis with varying degrees of lipidization (xanthomatization). This is very rare case report of papular xanthoma (progressive nodular histiocytosis) occurring together with angiokeratoma. Patient was treated with steroids and steroid sparing agents with moderate amelioration of the symptoms.

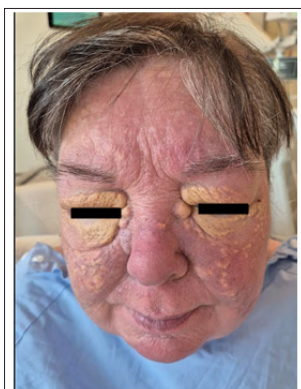


Figure A



Figure B

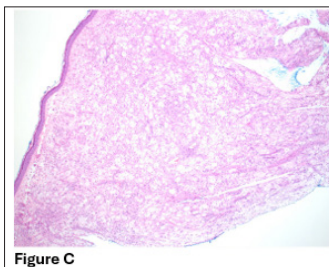


Figure C

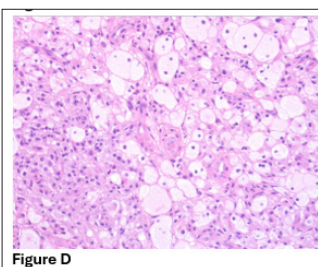


Figure D

Written Consent provided from patient to publish photo and Manuscript.

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