Langerhans Cell Histiocytosis with Multiple Adult Xanthogranuloma

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ABSTRACT

- A 61 year old man presented to the clinic with yellow-red papules coalescing into nodules and associated alopecia covering the scalp, ears, face, neck, trunk, and extremities. Histopathology of lesions biopsyed from the chest, back, and face exhibited a dense dermal infiltrate of foamy histiocytes with stains consistent with xanthogranulomas. However, there were also multiple eosinophils within the dermal infiltrate and striking follicular mucinosis with a proliferation of oval to round histocytes, confirmed to be folliculotropic Langerhans cells with appropriate staining. Literature reports multiple adult xanthogranuloma in association with lymphoproliferative disorders as well as cases of LCH in children that later presents with juvenile xanthogranulomas following chemotherapy. However, to our knowledge this is the first case of folliculotropic LCH presenting simultaneously with multiple adult xanthogranulomas in a middle-aged patient.

BACKGROUND

- Langerhans cell histiocytosis is a rare disease characterized by S100, CD1a, and langerin positivity and caused by clonal proliferation of dendritic cells. The disease is much more often seen in children than in adults. Any organ in the body may be affected (more commonly bone, skin, pituitary gland, lymph nodes, spleen, liver, CNS, and lungs) and extent varies. Prognosis correlates with number of organ systems involved. Therapy is also dependent upon organ involvement, but proper lab and imaging work-up is always primary. This includes CBC with diff, CMP, UA, TSH/T4, skeletal survey, coagulation studies, chest x-ray, abdominal and thyroid ultrasound, and head CT. Medical intervention may include methotrexate, azathioprine, diltiazem, hydrochlorothiazide, and coumadin.