An unusual case of adult onset multi-systemic Langerhans cell histiocytosis with perianal and subsequent incident thyroid involvement

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Introduction

- Langerhans cell histiocytosis (LCH) is a rare sporadic disease characterized by histiocytic neoplastic infiltration of various organ systems and a wide spectrum of clinical manifestations,ranging from benign and self-limiting to lethal.
- Involvement of the gastrointestinal tract is very rare in LCH, especially in adults, along with only a few isolated case reports in the literature indicating involvement of the perianal skin and rectum. Likewise, albeit reported to be more common in adults than children, involvement of the thyroid gland in LCH has been considered to be extremely rare, even in the setting of multifocal disease.
- Herein, we report a rare case of adult onset multi-systemic LCH with an initial perianal presentation and incidental finding of subsequent thyroid gland involvement in the follow up period.

Case

- A 36-year-old male patient with history of perianal LCH treated with surgical excision and local radiotherapy was referred to our endocrinology department upon detection of hypermetabolic nodular lesions in the left lateral lobe of thyroid gland on Positron Emission Tomography - Computed Tomography(PET/CT) scan in the 9th month of follow up.
- Current evaluation revealed euthyroid status, a hypoechoic solid lesion of 13x9mm in size with irregular borders in the left thyroid lobe on thyroid USG and diagnoses of suspected oncocytic lesion, Hashimoto thyroiditis or LCH based on cytologic assessment of thyroid nodule fine needle aspiration of biopsy.
- The patient underwent total thyroidectomy and pathological assessment confirmed the diagnosis of Langerhans cell histiocytosis. Assessments in the 6th month of postoperative follow up revealed euthyroid status with no thyroid tissue remnants or pathological lymph node on thyroid USG.
- In view of the multifocal lesions indicating multi-system disease,a systemic chemotherapy protocol with combination of prednisone and vinblastine has been planned by the hematology department.

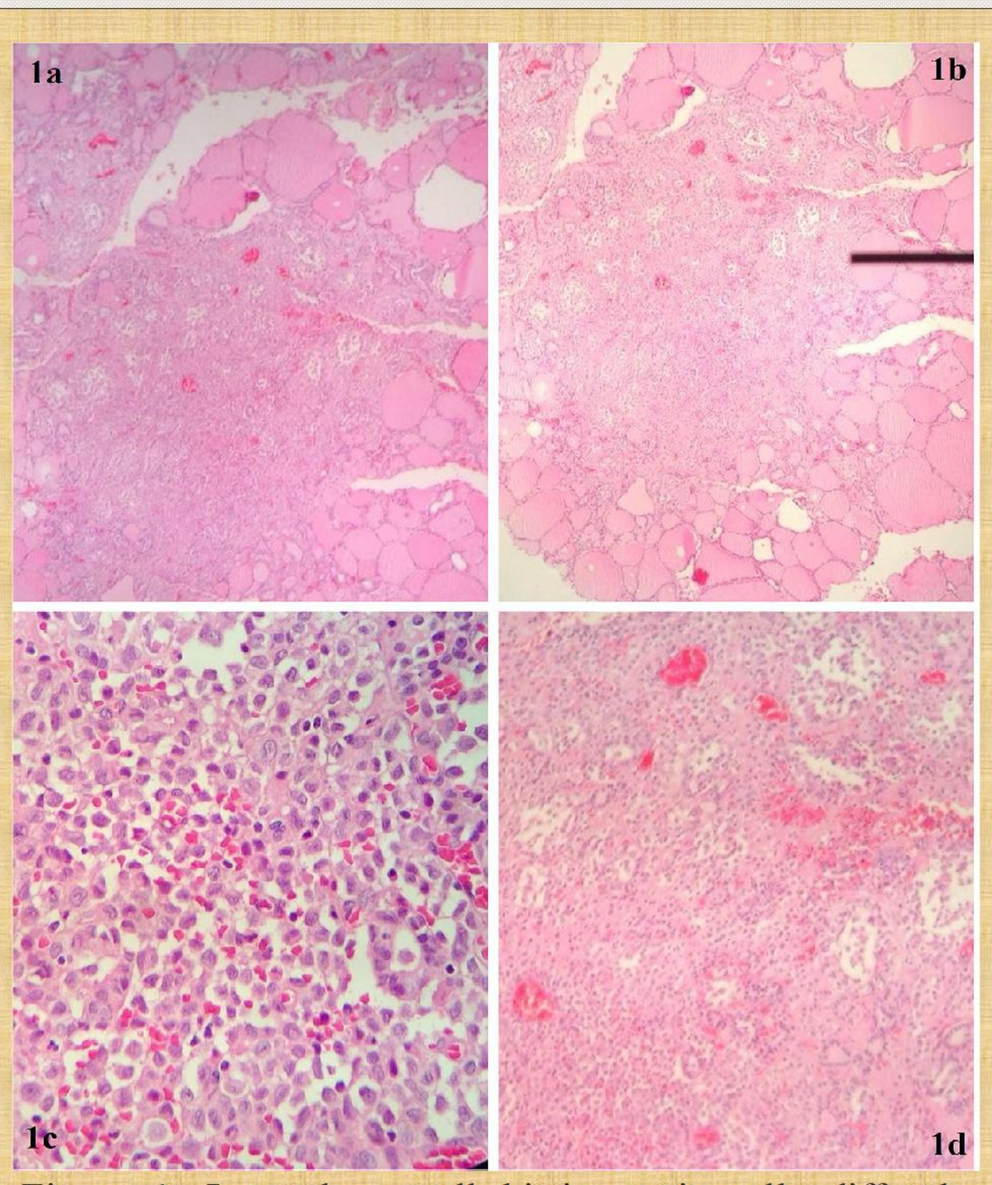

Figure 1. Langerhans cell histiocytosis cells diffusely infiltrating the thyroid gland

Conclusion

- Incidental detection of thyroid involvement in our patient with history of perianal LCH supports the likelihood of cutaneous involvement to precede multi-systemic disease, but also the rarity of solitary thyroid gland involvement of LCH, presenting as part of multisystem disease in most cases. Nonetheless, given that involvement of the thyroid gland in LCH is extremely rare, even in the setting of multifocal disease, our findings indicate a very rare and unique presentation of adult onset multi-systemic LCH with involvement of perianal mucosa and thyroid but no other systemic foci.
- In conclusion, we report the clinical experience and management of an adult onset perianal LCH with an incidental finding of thyroid gland involvement. Our findings underscore the importance of awareness of this rare condition associated with low index of suspicion among practitioners for LCH as well as the significance of ensuring proper follow-up in adult onset LCH to rule out systemic disease. Along with implementing a treatment plan matching the prognostic stratification of the patient, multidisciplinary approach and long-term close follow up with PET/CT scan in the management of adult onset LCH seem crucial given its rarity and varied presentation.

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