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Spontaneously vanishing sellar-suprasellar mass, panhypopituitarism and rashes in an adult with Multisystem Langerhans Cell Histiocytosis:

A Case Report

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INTRODUCTION

Langerhans Cell Histiocytosis (LCH) is a rare disorder mainly affecting children. It has varied manifestations and prognosis. Diabetes Insipidus is the most common endocrinologic abnormality in LCH while panhypopituitarism is rare. Spontaneously vanishing sellar/suprasellar masses associated with LCH has not been reported in literature both locally and internationally. Skin involvement is common in LCH but manifests differently between children and adults.

CASE PRESENTATION

We present case of a Multisystem LCH with unusual manifestations. A 19-year old female had a two-year history of visual loss, polyuria and polydipsia, secondary amenorrhea, generalized maculopapular rashes and a sellar-suprasellar mass on MRI. She presented at the emergency room lethargic, drowsy and hypotensive. Laboratory tests showed hypernatremia, inappropriately low urine osmolarity for a high serum osmolarity, hypocortisolism, hypothyroidism and hypogonadism. A repeat cranial MRI showed a normal pituitary gland without any masses or signs of hydrocephalus. Contrast-enhanced CT-scan of the chest and abdomen showed pulmonary fibrosis, hepatomegaly and lytic lesions with soft tissue components in the iliac crest and mandible. Biopsy of the maculopapular lesions on the skin was positive for \$100 and Cd1a, consistent with the diagnosis of Langerhans Cell Histiocytosis. She was then diagnosed with Multisystem LCH and was given Levothyroxine, Prednisone and Desmopressin. Chemotherapy was being planned for the patient, however, she succumbed to acute pulmonary embolism.

CONCLUSION

This case reports highlights the myriad and variable clinical presentation of LCH that could affect the work-up and management of adult patients with vanishing sellar-suprasellar mass and panhypopituitarism.