LANGERHANS CELL HISTOCYTOSIS PRESENTING WITH DIABETES INSIPIDIS

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INTRODUCTION

A 14-year-old male was diagnosed with diabetes insipidus and ultimately found to have Langerhans Cell Histiocytosis (LCH). LCH is an exceptionally rare disease, particularly in the pediatric population, with an annual incidence of 1-2 cases per million newborns and 4-5 cases per million children under age 15. Despite its rarity, the etiology of LCH remains poorly understood, encompassing various presentations, with rash as the most common initial finding.

METHOD

Workup During Admission

CBC: WBC 10.2, Platelet 527
CMP: NA 153
Urine osmolality: 301
ACTH: 28
Cortisol: 22.1
Beta hcg < 2
AFP 1.8
Copeptin: 2
H.pylori antigen: negative
FSH: 0.6
LH: 0.8
Uric acid: WNL
Angiotensin converting enzyme: 36
Xray bone age: WNL
MRI brain wo contrast: Abnormal appearance of the pituitary hypoplastic with some thickening of the pituitary stalk, suspicion for absent posterior pituitary favoring diagnosis of lymphocytic hypophysitis

Sacral bone biopsy conducted a month later: pathology confirmed Langerhans Histiocytosis

RESULTS & DISCUSSION

Patient was initially seen in the ED due to persistent episodes of nausea and vomiting for 6-8 months. Patient had previously followed with GI and been treated with omeprazole with little improvement. Patient also reported polydipsia and polyuria for 2 years. Endocrinology work up for diabetes was performed; however A1c was 5.3. Additional workup showed CMP unremarkable, serum osmolality 301, normal calcium, normal beta hcg, unremarkable AFP level. MRI of brain showed “Abnormal appearance of the pituitary. Favor diagnosis of lymphocytic hypophysitis”. Endocrinology was consulted and felt that Langerhans Cell Histiocytosis or Germinoma was a more likely diagnosis in a 14-year old. Pediatric oncology also followed the patient for the undifferentiated diagnosis. Water deprivation test was performed and confirmed diabetes insipidus. Patient was also found to have sacral manifestation, and biopsy ultimately confirmed Langerhans Cell Histiocytosis.

CONCLUSION

This patient's initial presentation consisted of diabetes insipidus and pituitary infiltration, which is quite uncommon in LCH. The prognosis of LCH varies significantly, ranging between 50% mortality among children under 2 with disseminated LCH and a potentially prolonged life-span for those with localized disease. This unique case contributes valuable insights for approaching similar cases, ultimately enhancing the overall understanding and outcomes in the realm of Langerhans Cell Histiocytosis. The significance of this case for the osteopathic perspective lies in this patient’s unique presentation, emphasizing how important the holistic view is in determining a patient’s diagnosis.

REFERENCES

1.Tillotson CV, Anjum F, Patel BC. Langerhans Cell Histiocytosis. 2023 Jul 17. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan–. PMID: 28613635.
2.Aruna DR, Pushpalatha G, Galgali S, Prashanthy. Langerhans cell histiocytosis. J Indian Soc Periodontol. 2011 Jul;15(3):276-9. doi: 10.4103/0972- 124X.85675. PMID: 22028518; PMCID: PMC3200027.
3.Jezierska M, Stefanowicz J, Romanowicz G, Kosiak W, Lange M. Langerhans cell histiocytosis in children - a disease with many faces. Recent advances in pathogenesis, diagnostic examinations and treatment. Postepy Dermatol Alergol. 2018 Feb;35(1):6-17. doi: 10.5114/pdia.2017.67095. Epub 2018 Feb 20. PMID: 29599667; PMCID: PMC5872238.
4.Milen Minkov (2018) An update on the treatment of pediatric-onset Langerhans cell histiocytosis through pharmacotherapy, Expert Opinion on Pharmacotherapy, 19:3, 233-242, DOI: 10.1080/14656566.2018.1429405
5.Morimoto, A., Oh, Y., Shioda, Y., Kudo, K., & Imamura, T. (2014). Recent advances in Langerhans cell histiocytosis. Pediatrics International, 56(4), 451-461.