Adult E-Poster

In 2022, investigations confirmed elevated vasoactive intestinal peptide (VIP) levels of 276 pg/mL (normal reference: <75 pg/mL) and a pancreatic body tumor with diffuse pancreatic infiltration, as well as liver and nodal metastases were identified on multimodal imaging. Extensive debulking surgery was recommended, but the patient declined.

In March 2023, she presented to our hospital with hypovolemic shock resulting from profuse diarrhea and severe hypokalemia. Intramuscular octreotide long-acting release (LAR) 30 mg was initiated, leading to prompt symptom control, and she continued to receive monthly injections. Genetic studies confirmed multiple endocrine neoplasia type 1 (MEN 1), prompting subsequent screening of family members.

By March 2024, the patient was asymptomatic, had experienced significant weight gain (from 50 to 65 kg), and her VIP levels had normalized to 66 pg/mL. A Gallium-68 (Ga-68) DOTATATE PET/CT scan in March 2024 demonstrated stable pancreatic and nodal disease but progressive liver metastases. The neuroendocrine multidisciplinary team offered the option of peptide receptor radionuclide therapy (PRRT), but she declined this treatment.

CONCLUSION

This case highlights the effectiveness of medical management with Octreotide LAR for control of symptoms and tumor-related hormonal hypersecretion in inoperable metastatic VIPoma regardless of disease progression. A personalized multidisciplinary approach is a pre-requisite for achieving optimal outcomes.

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THE SHRINKING HYPOTHALAMIC LESION: SERIAL MRI-GUIDED MANAGEMENT OF REFRACTORY XANTHOMATOUS HYOPHYSITIS

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INTRODUCTION/BACKGROUND

Xanthomatous hypophysitis is a rare inflammatory condition affecting the pituitary gland, characterized by the presence of lipid-laden macrophages (xanthoma cells) and chronic inflammation.

CASE

We describe a 26-year-old female with a 12-year history of xanthomatous hypophysitis complicated by panhypopituitarism and diabetes insipidus. Initial presentation at age 13 years with growth failure led to transcranial tumor debulking in 2016, with histopathology confirming chronic xanthogranulomatous inflammation. Due to recurrent disease activity, she received two courses of high-dose prednisolone (2018 and 2019-2020), followed by maintenance azathioprine (initiated 2020, currently 25 mg daily).

Serial MRI surveillance showed a gradual regression of the lesion over time. In July 2023, the hypothalamic lesion measured 1.3 × 0.8 × 0.8 cm, a reduction from its previous size of 1.5 × 1.3 × 1.1 cm in 2021. This regression was accompanied by decreased compression on the optic chiasm. The pituitary gland remained thinned, and the posterior pituitary bright spot is absent, consistent with long-standing structural damage. These findings suggest ongoing inflammatory control under azathioprine.

Hormonal management included levothyroxine (37.5 mcg/day), sublingual desmopressin (60 mcg/day for DI), and cyclic estrogen-progestin (Progyluton). Adrenal function recovered post-steroid withdrawal, confirmed by a normal Synacthen test (peak cortisol 624 nmol/L, 2022). Complications included secondary osteoporosis (spine T-score -1.8, hip -2.5) managed with calcium/vitamin D, and microcytic anemia (Hb 9.4 g/dL, MCV 29.8 fL), likely due to iron deficiency, now treated with oral iron and folate.

CONCLUSION

This case highlights the chronic relapsing-remitting nature of xanthomatous hypophysitis, necessitating long-term immunosuppression and meticulous endocrine surveillance. The latest MRI findings (2023) confirm disease stability under azathioprine, supporting its role in preventing progression. However, residual hypothalamic involvement underscores the need for continued monitoring. A multidisciplinary approach (endocrinology, neurosurgery, rheumatology) remains essential to manage hormonal deficits, bone health, and potential disease recurrence. Future considerations include azathioprine tapering if imaging remains stable, emphasizing the importance of serial MRI in guiding therapy.