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Seller Mass in a Young Man with Diabetes Insipidus and Panhypopituitarism—What is the Diagnosis?

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Abstract Pituitary and suprasellar lesions can cause diabetes insipidus due to interruption of pathways carrying vasopressin and oxytocin to pituitary. Central diabetes insipidus is characterized by polyuria and polydipsia and is a direct result of deficiency of vasopressin. Midline mass lesions in brain and body warrant further workup for possible diagnosis of germ cell tumors. We present an interesting case of a young man with suprasellar brain lesion and diabetes insipidus with pan hypopituitarism where further workup revealed germinoma. Patient had a favorable outcome with surgery followed by radiation therapy.

Keywords: *diabetes insipidus, germ cell tumor, panhypopituitarism*

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1. The Case

A 28-year-old Hispanic male presented with gradually worsening dizziness, headache and blurry vision for 2 weeks. His headache was bifrontal, throbbing and was associated with photophobia. Past medical history included mild intermittent asthma and suprasellar brain tumor. Patient had polyuria and polydipsia. He also had cold intolerance and decreased libido.

Review of systems was negative for nausea vomiting diarrhea, chest pain, dyspnea or abdominal pain. He denied any weight loss or focal weakness.

There was no past surgical history, he is allergic to penicillin, NSAIDS and IV contrast dye. Home medications included albuterol inhaler and Claritin. He lives with family and works as a waiter. Family history was noncontributory. He denied substance abuse and quit smoking 1 year ago.

The vitals showed pulse 73, blood pressure was 112 x 74, saturating 99% on room air and daily urine output of 10 Liter. Head was normocephalus, pupils equal and extraocular muscles were intact. Peripheral vision by confrontation method was normal. Neck is supple, no lymph node or JVD appreciated. No thyromegaly. Lungs clear and heart sounds normal, abdomen nontender with no hepatosplenomegaly, no pedal edema. CNS examination revealed normal cranial nerves, gait, reflexes and cerebellar signs, no motor or sensory deficits.

Routine labs were as follows--WBC count 8.11, hemoglobin 13.3, platelets 297 sodium was 139 potassium 4.0 bicarb was 30 BUN was 8 creatinine 0.9 glucose 92. LFTs normal, serum osmolality calculated was 276, measured serum osmolality 299 and urine osmolality less than 100. Toxicology screen, HIV, hepatitis-B and C panel and urine analysis negative. Prolactin level is 39 [2.5–17], beta hcg elevated 6.6 [normal less than 5], alpha -fetoprotein 1.9 [0 – 8], am cortisol low at 1.2 [3 to 22]. Thyroid studies showed free T4 0.49 [0.5-1.8] with inappropriately low TSH 2.3 for the T4 value. Testosterone was 18 [270 – 1730], with inappropriately low LH less than 0.07 [0.8 – 7.6], and FSH 0.69 [0.7 – 11].

Chest x-ray was negative. CT head showed at 2.6 cm enhancing suprasellar mass showing interval enlargement. MRI confirmed the mass lesion, now 3 x 2 x 2 cm with extension to the posterior aspect of sella. MRA of the head and MRI of spine was negative. CT chest abdomen pelvis showed mild bronchiectasis and scrotal ultrasound was normal.

The patient was admitted with suprasellar mass with central diabetes insipidus and panhypopituitarism. Elevated beta HCG in a male patient with midline brain tumor made us entertain germ cell tumor as prime differential diagnosis, along with others as lymphocytic hypophysitis, pituitary macroadenoma or neurosarcoidosis.

Ophthalmology examined our patient and patient had bitemporal hemianopsia. The patient was seen by neuro surgery and craniotomy was done with removal of the mass.

Prednisone was started for secondary hypocortisolism in higher doses, 15 mg qam and 5 mg qpm to cover for peri-operative stress. For hypothyroidism, Levoxyl was started and decision was made to pursue gonadotropin supplement as outpatient. For central diabetes insipidus Desmopressin was started 10 mcg per day and later on increased to 10 mcg b.i.d. patient's urine output decreased from 10 L per day to 3 L per day and urine osmolality increased from 110 to 650.

The tumor pathology returned as germ-cell tumor - germinoma with malignant cells, also confirmed by a 2nd opinion from Mayo Clinic. Keppra was started for seizure prophylaxis.

The patient recovered well and was discharged but did not immediately follow up outpatient with oncology secondary to financial constraints. Patient was readmitted about 5 months later with tumor recurrence as shown on several intracranial sites in repeat MRI. The patient then followed up with radiation oncology and received radiation therapy with complete resolution of tumor lesions. He continues to follow in our clinic with no recurrence of tumor noted.

2. Discussion

Pituitary stalk lesions can be congenital, inflammatory, infectious or neoplastic. Pituitary stalk lesions are discovered incidentally on MRI or during the work up of diabetes insipidus [1]. Central diabetes insipidus is a diverse condition characterized by polydipsia and polyuria due to deficiency of vasopressin [2]. There is a clear female preponderance in pituitary stalk lesions, but incidence of intracranial germ cell tumors is significantly higher in males, with 85% occurring prior to second decade of life. [3]

Interruption of pathway carrying vasopressin and oxytocin to pituitary can cause DI and loss of hypothalamic stimulation of pituitary gland can cause panhypopituitarism. The most common clinical presentations include DI, hypopituitarism hyperprolactinemia or visual disturbance [1]. Germ cell tumors of pituitary can secrete HCG [human chorionic gonadotropin] and AFP [alpha feto protein] which can assist in diagnosis. At high levels, these tumor markers can be measured in serum, although CSF levels are more reliable. [3] If they are only mildly elevated, tumor biopsy is strongly recommended to confirm the diagnosis.

A study in 1997 reported low positive hcg concentrations in pure germ cell tumors, as was the case in our patient [4]. A pituitary tumor can be either primary or metastatic and imaging alone cannot reliably separate these entities [5]. There is a report of pulmonary carcinoid with metastasis to pituitary gland causing hypopituitarism. [6]

Radiation therapy remains very effective treatment and has a cure rate of 90% [7], explaining significant response in our patient.

There are reports of resolution of DI with time, especially if surgeon preserves the stalk or if stalk dissection is as distal as possible. [8] There is also a report of successful twin pregnancy in a patient treated for pan hypopituitarism caused by supracellar germinoma [9].

3. The Takeaway

The take home message in our case is to consider germ cell tumor as a differential in young patients with midline tumors in chest, abdomen or brain, especially with elevated HCG or alpha feto protein. Also, post op follow up is important for further treatment with radiation for complete resolution and remission. Fortunately, our patient had a favorable outcome with radiotherapy.

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