A RARE CAUSE OF HYPOPITUITARISM IN A DOWN SYNDROME PATIENT

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Disclosures: None

Background:

Sellar masses arise from intra-pituitary or para-sellar tissues. Up to 80% are pituitary adenomas and the most common non-adenomatous lesions are rathke's cleft cysts, craniopharyngiomas, and meningiomas. Meningiomas account for about 10-15% of non-adenomatous sellar masses. Brain tumors are rare in individuals with Down syndrome.

Case Presentation:

A 28-year-old male with Down syndrome presented to his PCP's office with 45 lb. weight loss over a year, fatigue, blurry vision, chronic diarrhea, and hypotension. Prior labs were interpreted as normal but significant for normal TSH with a low FT4; repeat labs demonstrated low FT4 and a mildly elevated TSH. Patient was started on levothyroxine and developed worsening symptoms including hypotension.

He was then referred to endocrinology for possible central hypothyroidism. Further investigation revealed pan-hypopituitarism The patient was started on prednisone 5 mg for secondary adrenal insufficiency. MRI of pituitary showed a 2.1 x 2.6 x 2.8 cm sellar and supra-sellar mass with extension into the right cavernous sinus concerning for pituitary macroadenoma with mass effect on the optic chiasm. A trans-sphenoidal resection was attempted, however, due to the fibrous nature of the tumor the procedure was aborted. Biopsy showed a grade 1 meningioma with a ki67 proliferation index of 5.5%. Subsequently, the patient underwent craniotomy for the debulking of the tumor with a slight improvement in vision. Post-operatively, labs show persistent pan-hypopituitarism.

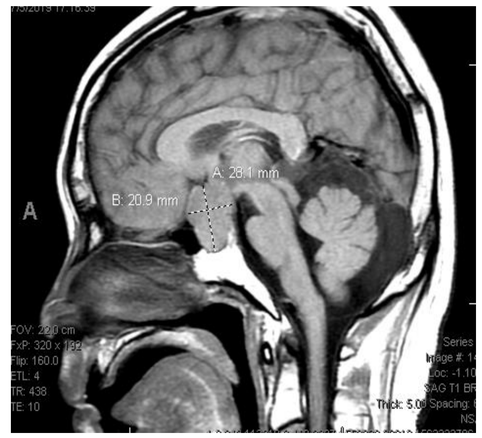
Follow up:

He has done remarkably well and completed a course of radiotherapy despite the COVID-19 pandemic. He continues steroid and thyroid replacement and was recently started on testosterone as well.

Labs:

TSH	5.160 (N: 0.45 – 4.5 uIu/ml)
FT4	0.43 (N: 0.82 – 1.77 ng/dl)
TPO AB	10 (N:0 - 34 IU/ml)
Cortisol	0.7 (N: 6.2 – 19.4 mcg/ml)
Prolactin	19.6 (N: 4 - 15.2 ng/ml)
ACTH	16.1 (N: 7.2 – 63 pg/ml)
IGF-1	212 (N: 98 - 282 ng/ml)
Total Testosterone	<3 (N: 264 - 916 ng/ml)
FSH	0.9 (N: 1.5 – 12.4 mIU/ml)
LH	0.7 (N: 1.7 – 8.6 mIU/ml)

MRI Brain:



Discussion:

Intra-sellar meningiomas arising from the dura surrounding the pituitary fossa are rare and can mimic non-functioning adenomas. Differentiating meningioma from non-functioning adenoma can be challenging given the similar clinical presentations. MRI shows homogenous enhancement in >90% of sellar meningiomas, consistent with the imaging in our patient. Imaging can also reveal a "dural tail sign", although this is non-specific. Suspicion of an intra-sellar meningioma could influence the surgical approach. To the best of our knowledge, this is the first reported case of an intra-sellar meningioma in a Down syndrome patient.

References: *1.* ***Meningioma in Down Syndrome patient.***
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