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PITUITARY DYSFUNCTION WITH ASSOCIATED LESIONS IN THE HYPOYHALAMO-PITUITARY REGION : HISTIOCYTOSIS OR DYSGERMINOMA?

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Introduction-Aim

Patients who present with clinical and laboratory findings of pituitary dysfunction and whose MRI findings reveal increased pituitary size or thickening of pituitary stalk , posε a diagnostic challenge.
Differential diagnosis mainly includes dysgerminoma, histiocytosis and hypophysitis.
A non-invasive approach is often non-diagnostic
The aim is to present two patients with similar clinical picture and positive MRI findings in whom transphenoidal biopsy of of the pituitary lesion, led to prompt accurate diagnosis and further therapeutic management

Patient- 1

Prepubertal girl 8 years of age, presented with the main complaint of slow growth rate .
PMH Polyuria, Polydipsia, for three years, growth rate < 3 cm/yr
Physical exam: Ht: 122 cm (< 3rd %ile), Wt: 25 Kg (25η %ile.), HC:52 cm
She had no dysmorphic features. Thyroid gland was not palpable
Breasts, Axillae, Pubic Hair : Tanner I.
Laboratory investigations: CBC: Nl, UA. S.g.1005 BUN 33 Cr: 0.5
Nα: 145 K: 3.9 Ca: 9.8 P: 3.9 AP: 180 T4: 85 μg/dl TSH: 3.2 μIU/ml
Prl: 82 IGF- I: 34 ng/ml BA :7 yrs. PEAK GH 2.3 ng/ml
Peak cortisol 8.5 μg/dl At time point 0 cortisol 6.5 μg/dl ACTH: 14
Water deprivatton test12 h plasma Osm 297 mOsm Urine Osm 89 mOsm
Post Minirin Plasma Osm : 307 Urine Osm : 260
Skeletal survey, chest x-ray,abdominal U/S and hematologic indices for histiocytosis were negative. Serum and CSF levels of HCG and α-FP were negative. Cytology of CNS was also negative .

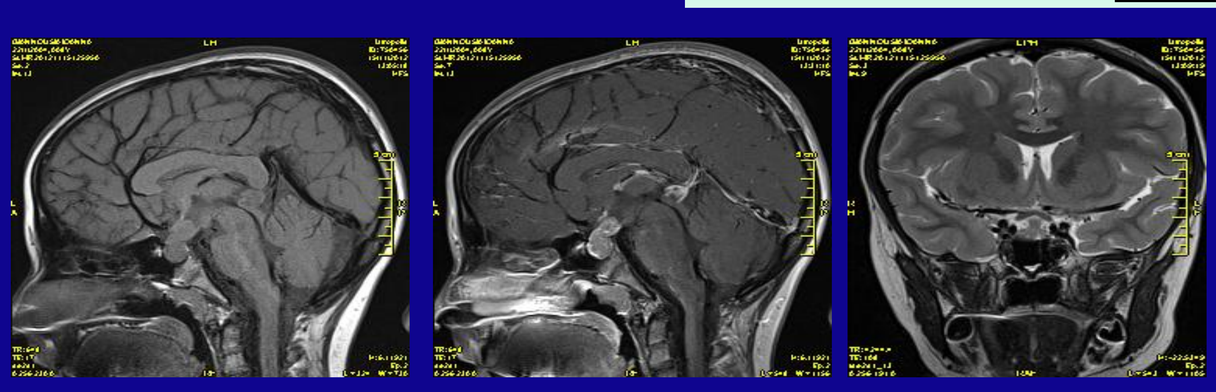
Diabetes insipidus, growth hormone deficiency and cortisol insuficiency

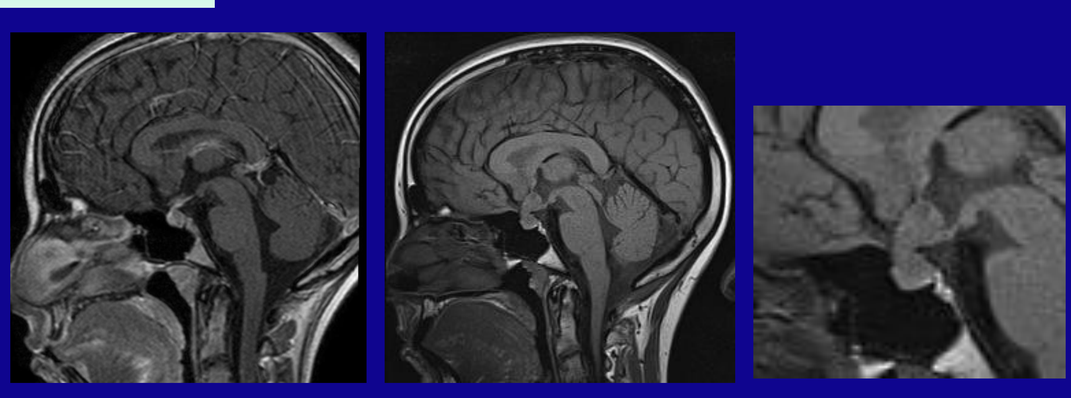
Patient- 2

15. 5 yrs old adolescent girl, presented for investigation of secondary amenorrhea and an episode of fainting.
PMH: Diabetes insipidus, treated with Minirin
Physical exam: Ht 171 cm (75th %ile), Wt: 67.5 Kg (75-90 %le)
She had no dysmorphic features. Thyroid gland was palpable Breasts, Axillae, Pubic Hair : Tanner V.
Laboratory investigations: CBC: Nl., UA s.g. 1005 BUN: 23 Cr: 0.8
Nα: 137 K: 4.0 Plasma osmolality: 290 mOsm Urine osmolality 496 mOsm T4: 70 μg/dl TSH: 0.103.2 μIU/ml Prl: 40.4 ng/ml
PEAK GH 1.57 ng/ml E2 < 12 pg/ml, FSH: 2,4 MIU/ml, LH 1,1 mIU/ml, prl: 40,4 ng/ml Peak cortisol : 21.2 μg/dl At time 0 cortisol 18.35 μg/dl
Skeletal survey chest x-ray,abdominal U/S and hematologic indices for histiocytosis were negative. Serum and CSF levels of HCG and α-FP were negative. Cytology of CNS was also negative .

Hypogonadotrophic hypogoandism, central hypothyroidism, diabetes insipidus, growth hormone deficiency hyperprolactinemia nad normal cortisol secretion

MRI





Management

Transphenoidal biopsy with excision of the lesion revealed dysgerminoma

The patient responded to two cycles of Carboplatin/Etoposide . She also received limited field RT (COG ACNS0232 Protocol).

Transphenoidal biopsy with excision of the lesion revealed dysgerminoma

The patient received chemotherapy consisting of Carboplerapyatin/Etoposide . She also received limited field RT (COG ACNS0232 Protocol).

Conclusions

Tissue biopsy provides the definitive diagnosis and the transphenoidal approach appears to be optimal for certain patients.

Inconclusive imaging appearance, negative markers and slow progression cannot exclude the diagnosis of dysgerminoma

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