

Primary empty sella: an unusual presentation

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Citation

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Abstract

Empty sella although reported to occur in 5.5% of the population,1 uncommonly presents with endocrine abnormalities. When they do present, endocrine disturbances are usually mild and usually represent a small increase in prolactin levels in adults. We present here an unusual case who presented clinically with primary amenorrhea where the cause remained undiagnosed for more than a decade by which time she had progressed to panhypopituitarism.

CASE HISTORY

A 25 year old female presented to the outpatient department of internal medicine with a previous diagnosis of primary amenorrhea and now increasing fatigue and lack of energy for the past year. She had also felt sleepier than before over this same period and would nap several times a day. She had been investigated previously for her primary amenorrhea at the age of 18 years. However, an incomplete hormonal profile done due to lack of available healthcare resources had revealed nothing abnormal. By the time she came to us she seemed resigned to her diagnosis of primary amenorrhea of unknown cause and appeared more concerned with her fatigue. She denied fever, cough, change in bowel habits, blood with stools, rash, jaundice, joint pains or swelling, snoring, headache, visual disturbances, or any nipple discharge. Her past history other than her amenorrhea was unremarkable. There was no family history of a similar illness. She was not on any medications.

On physical examination she was slightly built. Her pulse was 62/min and regular in rhythm. Her blood pressure was 80/60 mm Hg. Her secondary sexual characters appeared normal. She had no visual field deficits. The examination of her fundus failed to reveal any abnormalities. The rest of her systemic examination was also unremarkable.

Her blood counts and biochemical profile was normal. An x-ray chest showed no abnormalities. An ultrasound of the abdomen revealed a normal uterus and ovaries.

DISCUSSION

Considering her previous history of primary amenorrhea, present symptoms suggestive of probable hypothyroidism

and a low resting blood pressure, we thought primarily in terms of pituitary hypofunction secondary to a slow growing non functioning pituitary adenoma. A pituitary adenoma causing no signs of compression of the surrounding structures, however, seemed odd. Another possibility seemed pituitary dysfunction limited to gonadotropin secretion with coincident adrenal loss of function. A low accompanying blood pressure also made a probable primary adrenal disturbance likely. With these possibilities in mind, we next proceeded to resolve our doubts by determining the patient's LH and FSH levels, thyroid function, prolactin levels and morning cortisol levels. Her profile was revealing; both FSH and LH were very low, 7.4 U/L and 2.4 U/L respectively. FT3 & FT4 were undetectable, while TSH (4.8 mU/L) although within normal range was inappropriately low for the FT3 and FT4 levels. Cortisol was low normal at 12.8 µg/dL (12-25 µg/dL) and prolactin was mildly elevated at 29.4 ng/mL. This pointed to our first suspicion of hypopituitarism and it remained to be determined how extensive it was and what was its underlying cause. So far, our hypothesis of an underlying pituitary adenoma seemed justified; especially with elevated prolactin levels pointing towards stalk compression. However, accompanying hypothyroidism could also account for the rise in prolactin levels. On the other hand, the lack of clinical signs of compression of surrounding structures and absence of evidence of a raised intracranial pressure put the presence of an adenoma large enough to cause extensive pituitary hormonal deficiency in doubt. We decided, therefore, to first maintain our pursuit of determining the extent and level of pituitary hypofunction. This we completed by stressing the patient with insulin induced hypoglycemia and measuring

her consequent hormonal response. Both cortisol and growth hormone levels remained inappropriately low during the test (15.0 µg/dL & <1.00 µg/L respectively at the time of the hypoglycemia), confirming the presence of a panhypopituitarism. With extensive pituitary hypofunction determined the only thing that remained to be evaluated was its cause; which we expected to be a pituitary adenoma. An obvious choice now in this case was to do a magnetic resonance imaging of the brain. To our surprise imaging revealed an empty sella and not a pituitary adenoma (Fig 1, 2 & 3). No apparent cause for the empty sella could be determined. This led us to the diagnosis of a primary empty sella, presenting unusually first as primary amenorrhea and later going on to cause extensive pituitary hypofunction. Possibility of an initial pituitary adenoma which later involuted to leave an empty sella was also considered but seemed remote.

Figure 1

Figure 1

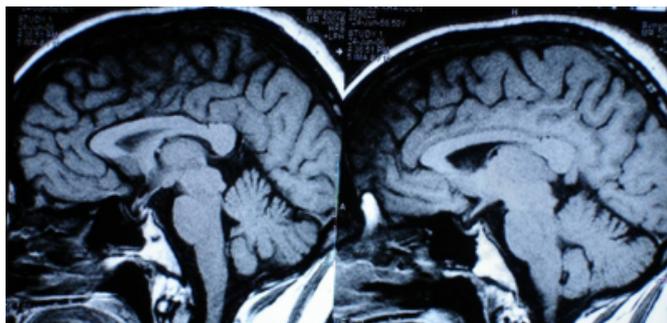


Figure 2

Figure 2

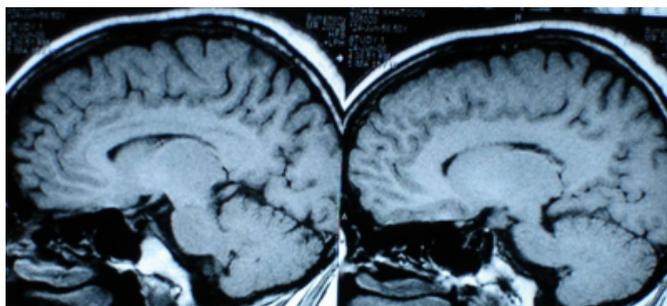
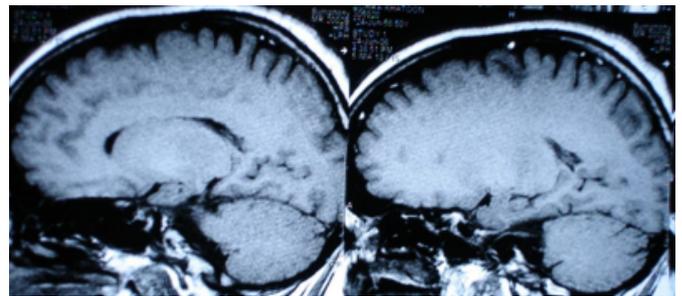


Figure 3

Figure 3



Primary empty sella, which by definition is intrasellar herniation of suprasellar subarachnoid space in the absence of an inciting cause rarely leads to a widespread pituitary hormonal deficiency. It is an even rarer cause of an extensive pituitary disturbance. ² An incomplete diaphragma sellae is essential for the development of an empty sella. ³ Most cases are asymptomatic and usually found incidentally. When present, hormonal deficiency generally manifests as a growth disturbance in children, ⁴ and usually a mild hyperprolactinemia in adults. ² The typical symptomatic patient is an obese multiparous female with occasional accompanying pseudotumor cerebri and CSF rhinorrhea. ⁵ Symptoms may also include headache and visual disturbances in occasional cases. Growth hormone reserve is usually affected in these patients. Although uncommon, sometimes thyrotropin and gonadotropin deficiency may be observed in symptomatic cases ⁵. Because hormonal deficiency is present in a minority of cases replacement therapy is not usually required.

On the other hand, secondary empty sella due to surgery, radiation, trauma, or a tumor, commonly leads to hormonal deficiency. Adhesions post surgery may require chiasmectomy. ⁶ Hormonal replacement is required in all symptomatic cases.

CONCLUSION

Clinical presentation of primary empty sella as primary amenorrhea is virtually unknown and represents a unique case. Although apparently rare as a cause it may need to be pursued in cases where amenorrhea secondary to gonadotropin deficiency remains unexplained, especially in third world countries where facilities for complete hormonal evaluation and advanced imaging remain marginalized. Also with its potential to convert into full scale pituitary hypofunction, early recognition of deficiency in relation to a confirmed primary empty sella will allow for timely recognition of progression and consequent replacement therapy.

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