An Unusual Case of Seizure

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ABSTRACT

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Ectopic posterior pituitary is an unusual congenital anomaly of the pituitary and is often associated with hypo plastic or absent pituitary gland and hypopituitarism. We are presenting an unusual case of seizure disorder since childhood, due to hypoglycaemia caused by hypopituitarism due to congenital hypoplastic pituitary gland and associated ectopic posterior pituitary

Keywords: Posterior pituitary Ectopia, Hormone deficiencies.

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INTRODUCTION

Posterior pituitary Ectopia is a rare congenital anomaly which is usually associated with hypo plastic or absent anterior pituitary gland, a completely absent or thin pituitary stalk and associated either isolated growth hormone deficiency or multiple pituitary hormone deficiencies. The ectopic posterior pituitary gland functions normally and the patient does not show features of diabetes insipidus.

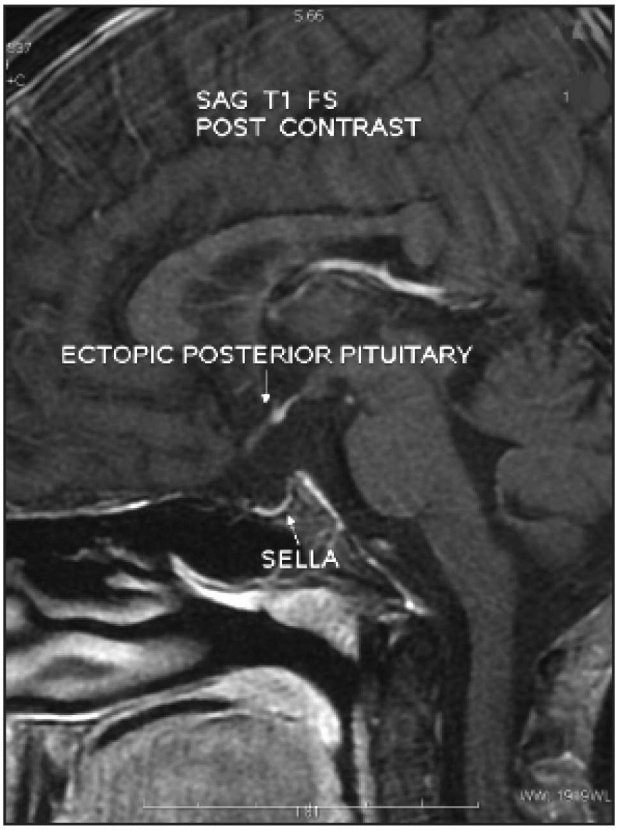

Figure 1. **Sagittal T1-weighted MR image shows the empty sella and ectopic posterior pituitary gland which is seen as an area of high signal intensity**

THE CASE

A 14 year old boy presented with history of recurrent episodes of generalised tonic clonic seizures since birth. He also had growth retardation. His antenatal and natal history was unremarkable. On examination his total height was 109 cm and he weighed 14 kg. He did not have any secondary sexual characters. His other

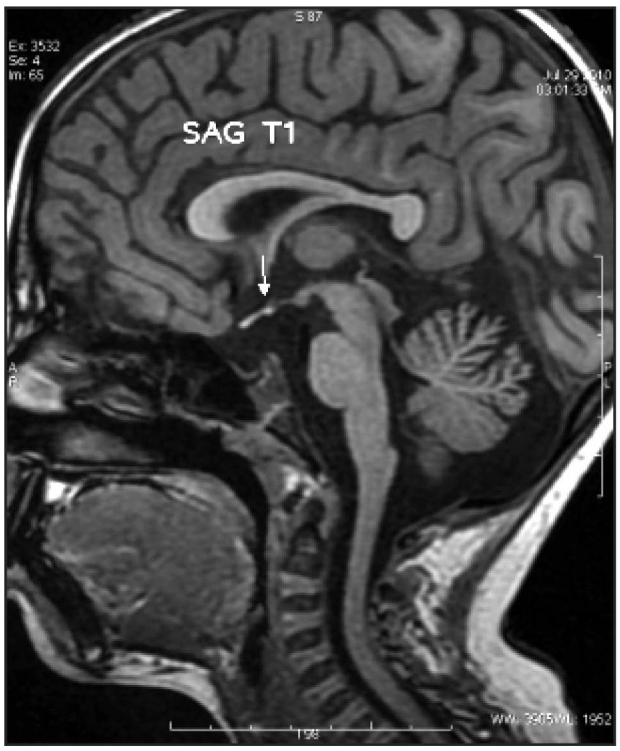

Figure 2. **Ectopic posterior pituitary gland is seen**

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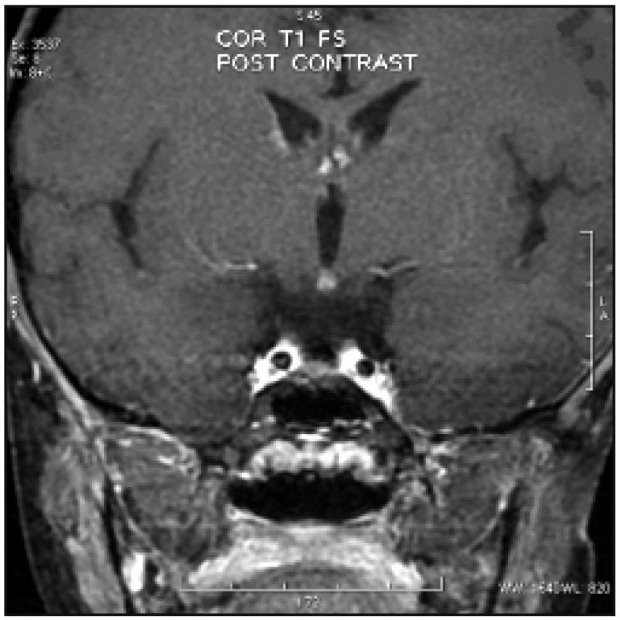

Figure 3. **Coronal view post contrast. The ectopic posterior pituitary gland is visible and the pituitary stalk is not seen**

system examination was unremarkable. His serum testosterone level was low 0.2ng/ml (normal value1.0-3.2), FSH was 0.49mIU/ml (normal 0.95-11.95) LH was 0.29 mIU/ml (normal1.3-12.9) ACTH level was<5pg/ml (normal 0-46 pg/ml) Prolactin level was 24.58ng/ml (normal3.46-19.4). His T3 level was 100ng/ml (normal 80-200) T4 was 5.6 microgram/ml (normal 4.5-12.5) and TSH was 0.67IU/ml (normal 0.35-5) His urine specific gravity was normal. MRI revealed absent anterior pituitary gland, absent pituitary stalk and a focal nodular iso to intermediate signals on T1 noted just posterior to the optic chiasma in the midline likely to be ectopic posterior pituitary gland.

The patient's history, examination findings, laboratory values in combination with MR findings, was diagnostic for pituitary stalk transection syndrome with an ectopic posterior pituitary gland. The seizures he was having since childhood was most likely to be due to recurrent episodes of hypoglycaemia. He was started on pituitary hormone replacement therapy.

DISCUSSION

Posterior pituitary ectopia refers to an absent normal posterior pituitary bright spot within the sella with ectopic bright signal at another site such as the median eminence on a weighted magnetic resonance. On imaging the lesion appears as a retro-chiasma mass which is bright on T1 and may demonstrate some degree of enhancement. Fat saturation fails to decrease the signal associated with the posterior pituitary. The differential diagnosis of suprasellar lesions with increased T1 signal includes craniopharyngioma; Rathke cleft cyst, hemorrhage, dermoid, lipoma, and ectopic posterior pituitary. The fact that the lesion is stable over time, mildly enhances and does not contain fat and in addition, the presence of a hypo plastic or absent sella suggests the diagnosis of an ectopic posterior pituitary gland. Ectopic posterior pituitary tissue could result from defective neuronal migration during embryogenesis. Another possible pathophysiologic mechanism for ectopic posterior pituitary tissue is transection of the pituitary stalk followed by hypertrophy of the proximal axons with subsequent reorganization. Transection of the pituitary stalk may be due to vascular accidents, anoxia or compression injuries. Breech deliveries and perinatal anoxia have been implicated as causes of transection of the pituitary stalk, with ensuing neuronal reorganisation of the proximal stump of the infundibulum.[1]However, many patients with hypopituitarism with ectopic posterior pituitary hyper intense signal have been reported to have uncomplicated perinatal courses. In our patient there was no history of breech delivery or perinatal anoxia. G Binder, et al has suggested that MRI pituitary morphology may have some correlation with the aetiology. Normal morphology suggests the presence of gene mutations, while severe hypoplasia with malformation might suggest traumatic insults.[2]A posterior pituitary hormone deficiency in the presence of posterior pituitary ectopia appears to be uncommon suggesting that this ectopic tissue functions normally to produce antidiuretic hormone.[3]Non-visualisation of the stalk is associated with lack of anterior pituitary function while visualization of the stalk is associated with preservation of some anterior pituitary function. The majority of isolated growth hormone deficiency (IGHD) patients had a truncated or thin stalk and a normal or small adenohypophysis, while an absent stalk and adenohypophysis are characteristic of multiple pituitary hormone deficiencies (MPHD). MR imaging can contribute to the prediction of the pattern and severity of hypopituitarism in patients with growth hormonedeficiency.[4]

END NOTE

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Conflict of Interest: None declared

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