CASE REPORT

Coexistence of Ectopic Posterior Pituitary and Sellar/Suprasellar Arachnoid Cyst: A Case Report

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Abstract: Background: The ectopic posterior pituitary (EPP) is a rare condition characterized by the ectopic location of the posterior pituitary lobe associated with varying degrees of stalk anomalies. The arachnoid cysts (AC) are benign lesions of the arachnoid, which account for 1% of all intracranial space-occupying lesions. Sellar/suprasellar ACs account for approximately 1% of all ACs. This is the first case of coexistence EPP with sellar/suprasellar AC.

Case Report: A 67-year-old woman presented with 6 months history of fatigue. Her medical history was positive for irregular menstruation. Her endocrine examinations indicated low free thyroxine level with low TSH level, low oestradiol with low gonadotrophin level, slightly elevated prolactin level. Her Insulin-like growth factor-I was below the normal levels. Dynamic contrast hypophys MR image revealed a sellar cystic lesion with a dimension of 18 × 14 × 14 mm, extending from the suprasellar cistern, traversing the diaphragma sellae and reaching the level of the floor of the 3rd ventricle, consistent with sellar/suprasellar AC. There was no wall enhancement. The optic chiasm was compressed. The precontrast T1-weighted magnetic resonance images did not demonstrate the characteristic bright spot of posterior pituitary within the sella, which was higher in position, in the region of the median eminence. The pituitary stalk was not present.

Conclusion: Although speculative, we have a hypothesis to explain how the EPP and sellar/suprasellar AC coexist in this patient. Due to the absence of stalk, CSF may enter the sella through the central aperture of the diaphragma sellae through which normally the stalk passes.

Keywords: Hypothalamo-hypophyseal axis, pituitary gland, pituitary stalk, sellar arachnoid cyst, ectopic posterior pituitary, magnetic resonance imaging.

1. INTRODUCTION

The ectopic posterior pituitary (EPP) is a rare condition characterized by the ectopic location of the posterior pituitary lobe associated with varying degrees of stalk anomalies [1]. The arachnoid cysts (AC) are benign lesions of the arachnoid, which account for 1% of all intracranial space-occupying lesions. ACs are rarely seen in the sellar/suprasellar region, which accounts for only 1% of all ACs [2].

The coexistence of EPP with sellar/suprasellar AC has not been previously described in the English literature. This report describes the first case of the association of EPP with sellar/suprasellar AC.

2. CASE REPORT

A 67-year-old woman presented with 6 months history of fatigue. Her medical history was positive for irregular menstruation. Visual field tests revealed bitemporal hemianopsia. Her blood tests, including complete blood count, routine biochemistry and urinalysis, were within normal limits. Her growth parameters were normal since childhood. Her endocrine examinations indicated low free thyroxine level with low TSH level, low oestradiol with low gonadotrophin level, slightly elevated prolactin level. Her Insulin-like growth factor-I was below the normal levels.

Dynamic contrast hypophys MR image revealed a sellar cystic lesion with a dimension of 18 × 14 × 14 mm, extending from the suprasellar cistern, traversing the diaphragma sellae and reaching the level of the floor of the 3rd ventricle, consistent with sellar/suprasellar AC. There was no wall enhancement. The optic chiasm was compressed. The precontrast T1-weighted magnetic resonance images did not demonstrate the characteristic bright spot of posterior pituitary within the sella, which was higher in position, in the region of the median eminence. The pituitary stalk was not present (Figs. 1 and 2).

3. DISCUSSION

Several recent studies have shown that mutations in gene HESX1 cause both EPP and periventricular heterotopia, and suggested that EPP is included in the spectrum of septo-optic dysplasia (SOD). Apart from SOD, several malforma-
Fig. (1). Coronal T2-WI, sagittal precontrast T1-WI and sagittal postcontrast T1-WI show a sellar-suprasellar cystic lesion having the same signal intensity as CSF, extending from the suprasellar cistern, traversing the diaphragma sellae and reaching the level of the floor of the 3rd ventricle (asterisks). (A higher resolution / colour version of this figure is available in the electronic copy of the article).

Fig. (2). Sagittal precontrast T1-WI does not reveal the characteristic bright spot of posterior pituitary within the sella, which is higher in position, in the region of the median eminence (arrow). (A higher resolution / colour version of this figure is available in the electronic copy of the article).

ACs are fluid accumulations within the leaves of arachnoid. Sellar/suprasellar ACs account for approximately 1% of all ACs [2]. The pathogenesis of the development of sellar/suprasellar ACs remains unclear. It has been hypothesized that sellar/suprasellar ACs result from a congenital defect of diaphragma sellae, which allows arachnoid and cerebrospinal fluid (CSF) to enter the sella turcica via a ball--valve effect. This defect may close as a result of meningitis, bleeding, or inflammatory events forming a sequestered (noncommunicating) cyst with an arachnoid lining [5]. Headache and visual impairment are the most common symptoms related to sellar/suprasellar ACs. Endocrine symptoms are less common, usually involving gonadotrophic axis [2]. Patients may also exhibit deficiencies of growth hormone and thyrotropin. Hyperprolactinemia due to stalk effect is a common laboratory finding [6].

Although speculative, we have a hypothesis to explain how the EPP and sellar/suprasellar AC coexist in this patient. Due to the absence of stalk, CSF may enter the sella turcica from the central aperture of the diaphragma sellae...
Coexistence of Ectopic Posterior Pituitary Current Medical Imaging, calcifications or solid areas are identified [2, 5]. RCC is usually bounded and compressed against the pituitary gland and stalk seemed to be stretched over the cyst walls. The thin wall does not enhance. The pituitary anomalies, including both sellar/suprasellar ACs and EPP. Sellar/suprasellar ACs are seen as a well-delineated, balloon-shaped, homogenous, cystic sellar lesion with a typical CSF-like signal behavior, that is a hypointense T1-weighted signal and a hyperintense T2-weighted signal [2]. Expansion of the sella turcica is frequently observed. No invasion to cavernous sinus may indicate the diagnosis of sellar/suprasellar AC and EPP. MRI is the essential imaging modality of choice to detect the entire spectrum of pituitary anomalies, including both sellar/suprasellar ACs and EPP. Sellar/suprasellar ACs and RCC are usually detected. The thin wall does not enhance. The presence of solid components and calcifications favors a diagnosis of CP over RCC and AC. CP is usually hyperintense on T1-weighted images [7]. On MRI, RCC also presents as absence or hypoplasia of septum pellucidum resulting in a box-like appearance of the frontal horns, optic tract hypoplasia and hypothalamo-pituitary axis abnormalities. Abnormalities of the hypothalamic-pituitary axis include anterior pituitary hypoplasia, ectopic posterior pituitary, and empty sella [9]. Normally, the posterior pituitary is seen as a characteristically bright spot on unenhanced T1-weighted MRI. Combining a lack of the normal posterior lobe enhancement signal in the sella turcica and the presence of a hyperintense nodule in an ectopic location is classified as EPP [1, 3].

Sellar/suprasellar ACs should be monitored with serial imaging due to their growth risk. If the lesions increase, the surrounding tissues are oppressed, and the patient may experience headache, visual disturbance, or endocrine dysfunction, which require surgical intervention. The basic strategy is to remove the wall of the capsule and to allow the cyst to communicate with the subarachnoid space. The most commonly used methods are microsurgical fenestration via craniotomy, neuroendoscopic fenestration and cystoperitoneal shunting [8, 10]. Intraoperative ultrasound (IoUS) integrated with preoperative MRI can offer real-time visualization of the ACs itself, all its surrounding structures, both neural and vascular, as well as ventricular system and may be used when approaching those lesions from either via a transcranial route or via endonasal endoscopic approach. Thus, greater safety would be achieved [11].

CONCLUSION

In conclusion, this is the first report of the coexistence of sellar/suprasellar AC and EPP. MRI is the essential imaging of choice to confirm and demonstrate the extent of congenital abnormalities of hypothalamic-hypophyseal axis.

CONSENT FOR PUBLICATION

Not applicable.

STANDARDS OF REPORTING

CARE guidelines were followed in this study.

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CONFLICT OF INTEREST

The authors declare no conflict of interest, financial or otherwise.

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REFERENCES


