Current Research in MRI

Case Report

Sphenoidal Ectopic Adenohypophysis: A Rare Case Report and Literature Review

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Abstract

The ectopic located pituitary gland is extremely rare, as in other congenital pituitary anomalies. This term refers to the location in the pharyngeal wall or sphenoid bone extending along the persistent craniopharyngeal canal. It occurs as a result of migration disorder of the adenohypophysis and may present with pituitary dysfunction or often accompany midline anomalies-related symptoms. Correct diagnosis of this rare ectopic location is important for the prevention of unnecessary surgery and surgical-induced panhypopituarism. Here, we report a 41-year-old female patient with a pituitary gland located in the nasopharynx, which was detected incidentally in magnetic resonance imaging performed for neck pain.

Keywords: Craniopharyngeal canal, ectopic pituitary, magnetic resonance imaging

INTRODUCTION

Congenital anomalies of the pituitary gland are rare. They may be accompanied by midline cranio-fascial anomalies. Intrasphenoidal development of the anterior pituitary gland is also a very rare anomaly and is often asymptomatic. The reason for this anomaly is not clear.[1] In the literature, there are less than 10 cases related to this rare anomaly.[2–4] We aimed to present the adenohypophysis located in the nasopharynx through the persistent craniopharyngeal canal and the accompanying corpus callosum dysgenesis in this article.

CASE PRESENTATION

A 41-year-old female patient was admitted due to increased neck pain at 4-month intervals. There was no medical history other than a major depressive disorder. Physical examination revealed cervical axis flattening and paravertebral muscle spasm. Neurological deficits and muscle weakness were not detected. Then, cervical magnetic resonance imaging (MRI) examination revealed no pathology except flattening of cervical lordosis. But the cross-section of the image area revealed a linear structure going from the sella turcica to the nasopharynx. Subsequently, in the pituitary MRI study performed later, it was seen that the anterior pituitary gland extended to the superior pharyngeal wall from the persistent craniopharyngeal canal (Figure 1). Ectopic localization of the pituitary gland was also seen on computed tomography (Figure 2).

Since there was no hormonal abnormality in the patient, imaging studies were considered to be most compatible with ectopic adenohypophysis owing to arrest of normal gland migration. In addition, dysgenesis of the corpus callosum was observed in the brain MRI study for additional anomaly screening.

DISCUSSION

The pituitary gland is an organ of dual origin. The anterior lobe (adenohypophysis) is derived from oral ectoderm and is epithelial in origin, whereas the posterior lobe (neurohypophysis) derives from the neural ectoderm.[5] Adenohypophysis development in human begins with the formation of Rathke pouch in the fourth week. Rathke pouch (adenohypophyseal pouch) is formed by the funnel-shaped upward extension of the primitive oral roof lined with ectoderm. As the developmental steps progress, the adenohypophyseal pouch thickens, becomes elongated and eventually loses its connection with the primitive oral cavity in the seventh week. The 5 principal specialized endocrine cell types that form the pars distalis differ from the anterior wall of the pouch by the effect of various differentiation factors.[6]

Migration anomalies are congenital anomalies caused by genetic, vascular, or environmental insults to migrating neuroblasts. Exposure to ionizing radiation, excessive levels of alcohol (fetal alcohol syndrome), anticonvulsants (phenytoin and barbiturates), or toxic materials (methyl mercury poisoning) may be associated with abnormal neural cell migration.[7]

The persistence of the craniopharyngeal canal (CPC) is a corticated osseos canal that prolongs from the roof of the nasopharynx to the sellar region.[7] This canal, which traverses the sphenoid bone corpus, has also been named the transsphenoidal canal in the literature.[8] The incidence of

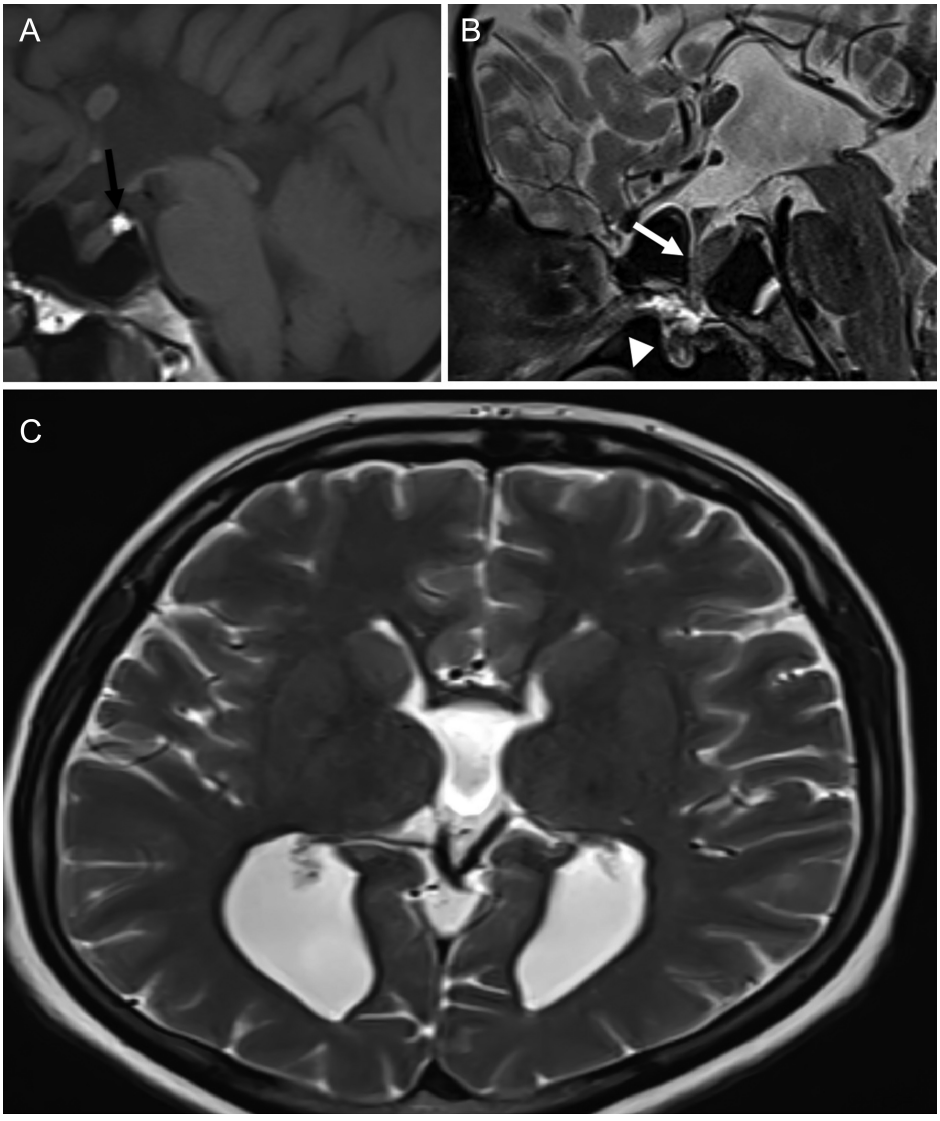

Figure 1. (A) Sagittal T1W, normally located neurohypophysis (black arrow). (B) The anterior pituitary gland extends from the persistent craniopharyngeal canal (white arrow) to the nasopharyngeal cavity (white arrowhead) and corpus callosum agenesis. In the sagittal T2W 3D-SPACE sequence (C) axial T2W, parallel configuration of the lateral ventricles and dilatation of the occipital horns secondary to corpus callosum agenesis. T1W, T1-weighted; T2W, T2-weighted.

CPC was reported as 0.42% in the population.[9] It is also stated that CPC may accompany many congenital anomalies. Parantez içerisindeki üstü çizili ifade metin içindeki atıfı belirtmekte olup buraya 8 ve 10 numaralı referansların "8,10" şeklinde eklenmesi uygun olacaktır. In our case, there was ectopic adenohypophysis and corpus callosum dysgenesis accompanying CPC, similar to the literature.

MAIN POINTS

- Persistent craniopharyngeal canal is a rare midline anomaly that may be accompanied by adenohypophysis malformations.
- Ectopic adenohypophysis may cause hypopituitarism.
- Accurate diagnosis of ectopic adenohypophysis and description of anatomical structures can prevent important complications that may occur during surgical procedures.
- In patients with persistent craniopharyngeal canal and ectopic pituitary anomalies, other midline anomalies in the head and neck region should also be investigated.

Congenital anomalies of the adenohypophysis may be associated with other midline anomalies such as Chiari type 1 malformation, dysgenesis of the corpus callosum, and agenesis of the internal carotid artery.[2,11] Our patient having dysgenesis of the corpus callosum supported the hypothesis that this malformation may be associated with midline anomalies. It has been hypothesized that the relationship between congenital anomalies of adenohypophysis and other midline anomalies is related to fetal pituitary hormone deficiency.[1]

Ectopic intrasphenoidal pituitary gland is an extremely rare migration anomaly and refers to the ectopic location of the adenohypophysis in the nasopharyngeal wall or sphenoid bone. In both possibilities, CPC allows this ectopic location. Well-developed neurohypophysis can be in its normal position.[1] In our case, it is seen that the neurohypophysis is in its normal location. Therefore, persistent CPC plays

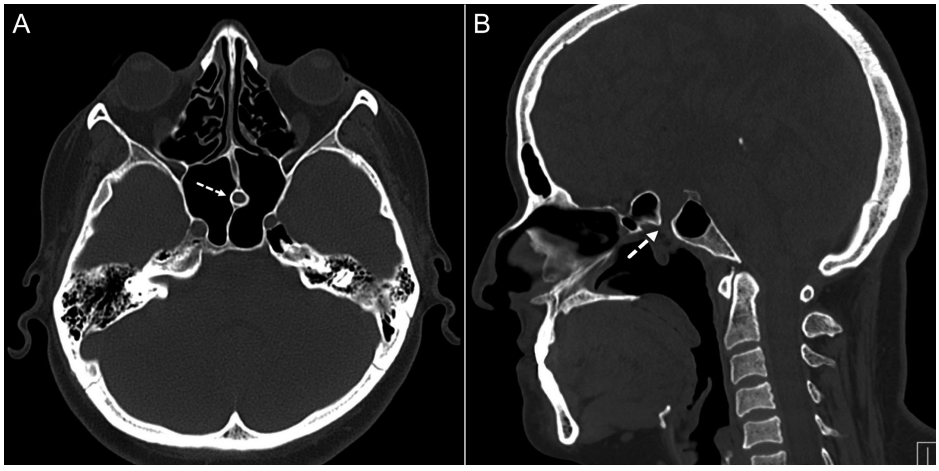

Figure 2. (A, B) CT (axial–sagittal), Persistent craniopharyngeal canal (dashed arrow) and protrusion of the adenohypophysis into the nasopharyngeal space. CT, computed tomography.

a key role in the occurrence of this anomaly. The CPC is believed to be due to the defective fusion in the postsphenoid cartilage and the non-obliteration of the adenohypophyseal stalk, which connects the adenohypophyseal pouch and the stomodeum.[3,12] In our case, the ectopic adenohypophysis was detected incidentally in the pharyngeal wall at the far end of the CPC and the neurohypophysis was in the intrasellar position. Abele et al[3] defined this as type 2 CPC which is characterized by the association of mid-sized CPC with ectopic pituitary tissue.

Various symptoms may be detected in cases of the ectopically located pituitary gland (type 2 persistent CPC). Pituitary displacement or surgical interventions may cause pituitary dysfunction.[3,12,13] Airway obstruction, accompanying symptoms related to hypothalamic hamartoma (hyperprolactinemia and precocious puberty), may be observed.[12,14,15] Also, it may present with symptoms and clinical findings related to other accompanying anomalies.[1,3] In our case, the diagnosis was made incidentally in an asymptomatic patient with significant pituitary displacement, and there was no hormonal abnormality. Therefore, a detailed evaluation in terms of pituitary function and other anomalies that may accompany these cases is important.[3] In addition, with the correct diagnosis, operation-related panhypopituitarism can be prevented by avoiding unnecessary surgery.[4,13] The causes of persistent CPC and ectopic hypophysis are still unclear and further studies are needed to find out the reasons that may prevent this malformation in the embryonic period. Clinical suspicion and early radiological evaluation (MRI) are required for accurate diagnosis and detection of other accompanying anomalies.

In conclusion, persistent craniopharyngeal canal and ectopic adenohypophysis are very rare variations and pathologies. Radiological evaluation is essential in the diagnosis and guiding treatment. The identification of these pathologies is of great importance in terms of preventing unnecessary surgery and possible complications.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

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Declaration of Interests: The authors declare that they have no competing interest.

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