Precocious true puberty secondary to a suprasellar arachnoid cyst: A case report and MR characteristics of common suprasellar cystic lesions

Fahad B. Al Badr *, Sarah A. Al Sultan

Department of Radiology, College of Medicine & King Khalid University Hospital, King Saud University, Riyadh, Saudi Arabia

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Abstracts The incidence of suprasellar arachnoid cysts (SSACs), although rare in children, is increasing as a result of the advent of computed tomography (CT) and magnetic resonance imaging (MRI). However, the pathogenesis of precocious true puberty in the presence of SSACs is unclear. In the present paper, we report the case of an 11-year-old boy with precocious true puberty who exhibited a classic SSAC that was identified using CT and MRI imaging. In addition, we highlight the occurrence of SSACs in association with precocious true puberty and discussing the imaging differential diagnosis and the role of imaging technique in reaching the diagnosis.

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1. Introduction

Arachnoid cysts are intra-arachnoid collections of cerebrospinal fluid (CSF)(1) that are often congenital in origin (2). In pediatric patients, arachnoid cysts account for 1% of intracranial mass lesions (2), and 9–11% of these cysts are located in the suprasellar region (1–3). Suprasellar arachnoid cysts (SSACs) may cause disorders involved in growth, puberty and hypothalamic-pituitary function due to the proximity of the cyst to the hypothalamic-pituitary area (4). The exact incidence of true precocious puberty with an SSAC is unknown, and it is an uncommon presentation with only sporadic reports in the literature (1). In the present work, we report a case of an SSAC presented with precocious true puberty and the differential diagnosis of suprasellar cystic lesions including epidermoid cyst, Rathke's cleft cysts and craniopharyngioma as well as the role of recent imaging in reaching the diagnosis.

2. Case report

An 11-year-old boy presented with a history of an abnormal increase in the size of his external genitalia and a change in his voice over a period of 2 years. There was no history of an associated headache, vomiting or a change in visual acuity.
There was no clinical evidence of diabetes insipidus. His height was in the 5th percentile, and his weight was in the 75th percentile for his age and sex. Upon neurological examination, the patient displayed no deficits or visual field defects. He exhibited mild macrocephaly, which manifested primarily as a prominent forehead. His sexual development, which was determined by his pubic hair and testicular volume, was classified as Tanner stage V.

An endocrinological profile revealed the following values: normal levels of luteinizing hormone (2.92 IU/L), follicle-stimulating hormone (1.61 IU/L) and thyroid stimulating hormone (1.71 mL U/L); increased levels of testosterone (32.93 nmol/L); and decreased levels of free thyroxine (8.84 pmol/L).

X-ray images of the patient’s left wrist revealed that the bone development of the patient was consistent with that of a healthy 13-year-old boy. A non-enhanced computed tomography (CT) scan of the patient’s brain (GE 64 slice, 5 mm slice thickness) showed an enlargement of the lateral ventricles and, to a lesser extent, of the 3rd ventricle. There was a large suprasellar cyst carrying CSF density without calcification, septation or an internal structure (Fig. 1).

Non-enhanced magnetic resonance imaging (MRI) of the patient’s brain (GE 1.5 T) revealed a large suprasellar cyst that measured $3 \times 4 \times 4$ cm$^3$. The signal intensity of this cyst was similar to that of the CSF; it appeared as a low signal on the T1-weighted images (Fig. 2), and a high signal on the T2-weighted images (Fig. 3) and was suppressed on the FLAIR (Fig. 4) and diffusion images. The data indicated a mass effect, which involved stretching and anterior displacement of the optic chiasm with superior displacement of the 3rd ventricle. The intrasellar extension of the cyst causing mild displacement of the pituitary gland downward and its stalk anteriorly.

The patient was examined by a neurosurgeon who planned an endoscopic fenestration of the cyst, but the patient’s family refused the operation. The patient was discharged and was prescribed a GnRH analogue to suppress his puberty and thyroxin as replacement therapy. Follow-up monitoring of this patient was not conducted because his parents retracted their consent for any surgical interventions.

3. Discussion

Arachnoid cysts likely develop during the anomalous splitting and duplication of the endomembranous during neural tube folding (1–2). SSACs are thought to develop from an anomaly of the Liliequist membrane, either as a diverticulum or as a result of a split within the membrane, forming a cavity where the CSF is secreted (1,5–7). This diverticulum may increase in size following inflammatory, hemorrhagic or developmental events (1,5,8). SSACs have been classified as communicating or...
noncommunicating cysts using CT cisternography and pneumoencephalography (9–10). Communicating cysts contain a basilar apex and represent an aberrant dilatation of the interpeduncular cistern. Noncommunicating cysts are thought to form within the diencephalic membrane of Liliequist and compress the interpeduncular cistern.

The majority of patients with suprasellar arachnoid cysts that have been described in the literature are children (9–12). The clinical manifestations of suprasellar arachnoid cysts include signs of obstructive hydrocephalus, visual impairments, endocrine dysfunction, gait ataxia and ‘bobble-head doll’ syndrome (2–9). SSACs may be asymptomatic and therefore may be discovered incidentally (1). Endocrine dysfunction may occur in the form of precocious puberty, amenorrhea, developmental delay, skeletal growth retardation and hypothalamic disturbances (2–3). Of the 54 cases of SSACs that were reviewed by Hoffman et al. (9) in 1982, only eight cases exhibited features of precocious puberty (14.8%). In another large case series of SSAC patients, four cases (out of 20) exhibited precocious puberty (13).

The pathogenesis of precocious puberty with an SSAC is controversial. These lesions may destroy the posterior hypothalamus while leaving the anterior hypothalamus intact. The intact anterior hypothalamus, in the absence of inhibitory influences, leads to an increased level of pituitary function. The preoptic region may play an important role in regulating the release of gonadotropic hormones from the anterior lobe of the hypophysis. According to Styme, the inhibitory tone within the central nervous system develops after birth, which reduces the levels of gonadotropin and prevents early pubertal development (1).

Other lesions that have been reported to present with precocious puberty are suprasellar epidermoid cyst and Rathke’s cleft cysts. A suprasellar germ cell tumor involving the anterior portion of the third ventricle presenting with both precocious puberty and diencephalic syndrome has been reported (14–15).

On CT scans, SSAC appears as a noncalcific low-density extra-axial lesion that is of CSF density and does not show enhancement after the injection of a contrast material. The lesion’s borders are smooth and clearly defined which may potentially fill the anterior third ventricle (12). CSF flow studies using radionuclide or iodinated contrast agents have been used to augment the diagnostic power of CT scans. These studies are also commonly used to differentiate between communicating and noncommunicating cysts or to differentiate between SSACs and cases of a dilated 3rd ventricle (9). MRI scanning has eliminated the need for invasive neuroradiological interventions (1). These scans can discriminate between the CSF of a true arachnoid cyst and the fluid of neoplastic cysts (2).

The MRI of arachnoid cysts shows a smoothly marginated mass that is hypointense on T1-weighted images and hyperintense on T2-weighted images. The MR signal intensity of this type of lesion is similar to that of CSF on all imaging sequences (1).
Other differential diagnoses should be considered in suprasellar cystic lesions including epidermoid cyst, Rathke’s cleft cyst (RCC) and craniopharyngioma.

**Epidermoid tumors** have CSF signal intensity on T2 and are slightly hyperintense on T1; however, they need to be distinguished from CSF-containing arachnoid cysts that also occur in the suprasellar region. Epidermoids have a higher CSF signal on fluid-attenuated inversion recovery and diffusion weighted imaging due to restricted diffusion, whereas the arachnoid cyst is dark on both sequences. Another distinguishing feature is the tendency for epidermoids to insinuate between adjacent structures, whereas the arachnoid cysts displace them. No enhancement is seen in the epidermoid or arachnoid cysts.

**RCC** either contains low protein content, which appears isointense with CSF on all sequences, or high protein content that becomes hyperintense on T1WI. RCC does not calcify. Although they rarely show enhancement, enhancing rims and nodules can sometimes be seen.

**Craniopharyngioma** has a heterogeneous appearance with solid and cystic elements. Cysts frequently contain high protein, cholesterol, or blood products, which are hyperintense on unenhanced T1-weighted images. The solid portions and cyst wall enhance heterogeneously. MRI with its multiplanar capability is essential for defining the tumor extension and is the most important imaging method used to plan surgical approaches. CT scan is the most sensitive method to demonstrate characteristic calcification that occurs in these tumors (16).

4. **Conclusion**

The present report describes a case involving a suprasellar arachnoid cyst in a child who presented with precocious true puberty. A male child presenting with precocious puberty showed be investigated for an intracranial tumor. Although the most common tumor in precocious puberty is a hamartoma, an arachnoid cyst should be considered in the differential diagnosis. Patients with suprasellar arachnoid cysts should be observed over long periods of time to monitor their growth and pubertal development (4).

5. **Consent**

Written informed consent was obtained from the parent for the publication of this case report, as the child was a minor. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**References**