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Transventricular Endoscopic Management of a Suprasellar Arachnoid Cyst Causing Precocious Puberty

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Suprasellar cysts arachnoid cysts (SSAC) are relatively uncommon making up less than 2% of all arachnoid cysts. Suprasellar arachnoid cyst causing precocious puberty is infrequent. The pathogenesis remains unknown in the majority of patients with precocious puberty. Endoscopic ventriculocystocisternostomy procedure is an effective and minimally invasive method in the treatment of these cysts.
This report describes a 7-year old male who had a huge intracranial arachnoid cyst with initial symptoms and signs of sexual precocity. Magnetic resonance image (MRI) of the brain revealed the presence of large suprasellar cystic lesion.

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1. INTRODUCTION

Arachnoid cysts are developmental anomalies of the arachnoid membrane collecting cerebrospinal fluid (CSF) [1]. Suprasellar region is an uncommon location for arachnoid cysts. Suprasellar arachnoid cysts (SSAS) represent less than 2% of all arachnoid cysts [2].

Intracranial hypertension due to hydrocephalus and visual changes are the most common symptoms of SSAS. Precocious puberty is a particular clinical presentation of SSAS. Few cases have been reported in the medical literature [3-11].

Several therapeutic strategies for the management of SSAC are proposed such as cyst fenestration, stereotactic drainage or resection through craniotomy, or cysto-peritoneal shunting [1,12-15].

Intracranial endoscopy is currently the treatment of choice of SSAC which allows cyst marsupialization without the invasiveness of open craniotomy [2,12,16-21].

We report an interesting case of precocious puberty caused by suprasellar arachnoid cyst in an infant male treated successfully by endoscopy.

2. CASE DESCRIPTION

A 7-year old male child, was referred with a two-weeks history of symptoms of progressively worsening increased intracranial pressure. No history of epileptic seizures has been reported by child's parents.

Neurological examination featured a conscious right handed child without motor deficit. Aggressiveness was observed during child examination. Ophthalmologic examination showed decreased visual acuity estimated to 5/10 with papillary edema in both eyes.

Signs of precocious puberty were observed on clinical examination. Indeed, axillary and pubic hair was well developed. Voice deepening and testicles enlargement were also observed on clinical examination.

Blood tests revealed higher levels of Follicle-Stimulating Hormone (FSH) and Luteinizing Hormone (LH) than normal for age. The plasma testosterone level was 4 nmol/l. Bone age on X-rays hand was more advanced than chronological age, confirming early puberty.

Brain magnetic resonance image (MRI), showed a large suprasellar cystic expanding intracranial lesion in the third ventricle. The cystic mass appeared hypointense on T1 weighted-images (WI), hyperintense on T2-WI, similar to cerebro-spinal fluid (CSF) signal, without enhancement after Gadolinium administration. The mass showed notable mass effect on surrounding structures and biventricular hydrocephalus was associated (Fig. 1).

Endocopic ventriculocystocisternostomy was performed. Through a right pre-coronal burr hole, the endoscope was introduced into the frontal horn of the lateral ventricle. The cyst bombs in the third ventricle and blocs the foramen of Monro. The superior wall of the cyst was coagulated and opened. The endoscope was then advanced into the cyst and ventriculostomy between the third ventricle and the basal prepontine cistern was made (Fig. 2).

The postoperative course was uneventful. Signs of increased cranial pressure disappeared two days postoperatively. The patient was discharged on the 5th postoperative day.

Physical examination six months later noticed regression of aggressive behavior and pubertal signs. MRI control showed a decrease of the size of the arachnoid cyst (Fig. 3).

3. DISCUSSION

Bright first described arachnoid cyst as "serous cyst in the arachnoid" in 1831 [22]. Suprasellar cysts arachnoid cysts (SSAC) are relatively uncommon making up less than 2% of all arachnoid cysts [2].

SSAC are thought to develop from an anomaly of the Liliequist's membrane [23], either as a diverticulum or from a split within the membrane and secretion of CSF within the cavity. This diverticulum could increase in size following inflammatory, haemorrhagic or developmental events [4].

Many arachnoid cysts are clinically silent and are identified only incidentally [24]. Expansion of

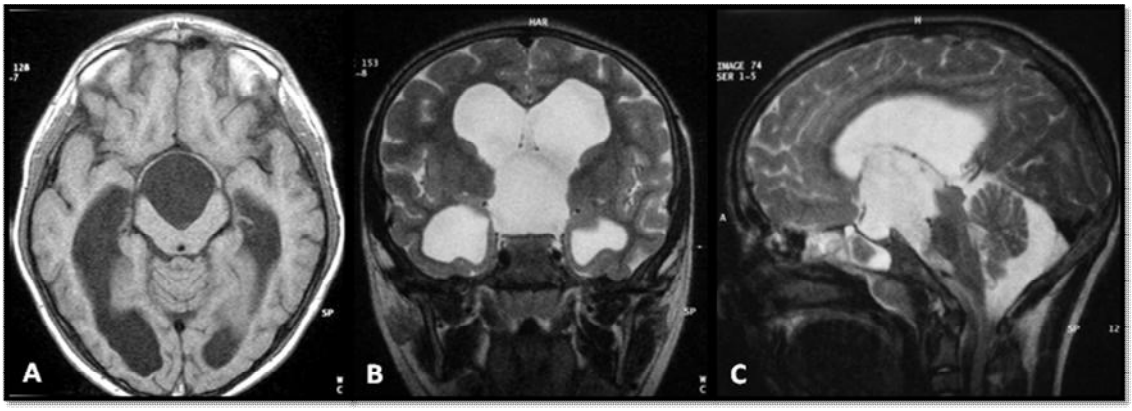

Fig. 1. A, axial T1-WI showing a homogeneous suprasellar cystic mass exerting a mass effect on the midbrain. C & D, coronal T2-WI and sagittal T2-WI displaying hyperintense suprasellar mass with obstructive hydrocephalus

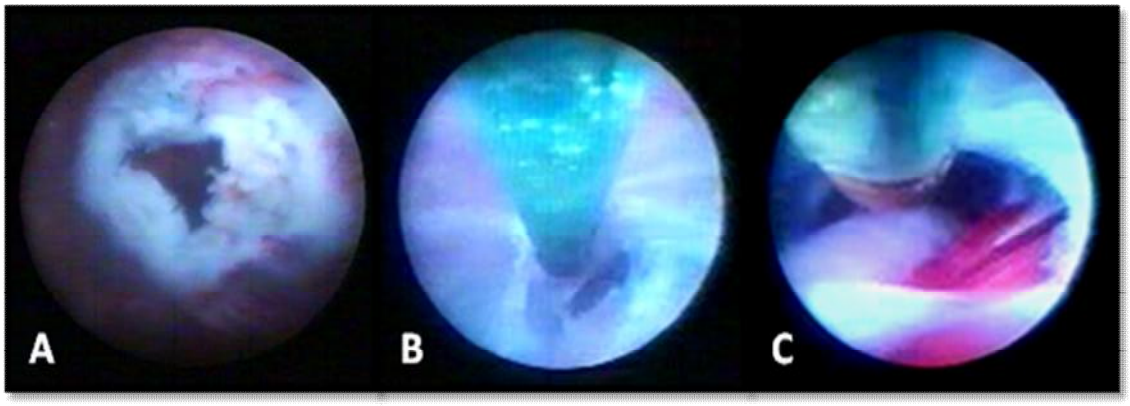

Fig. 2. Endoscopic operative view showing: A, fenestration of the superior wall of the suprasellar arachnoid cyst. B, opening of the floor of the third ventricle. C, visualization of neurovascular structures in the interpeduncular cistern

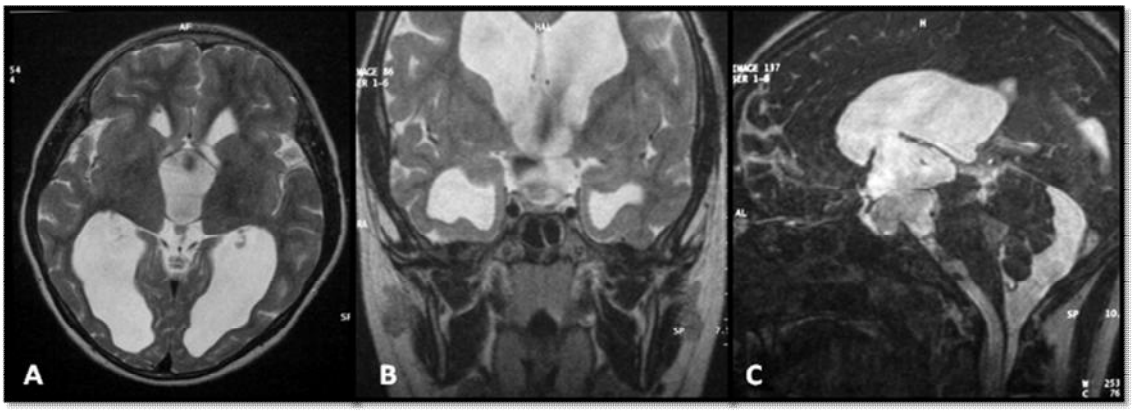

Fig. 3. A & B, postoperative axial T2-WI and coronal T2-WI showing a reduction of the size of the suprasellar cyst. C, sagittal CISS-3D MRI displaying CSF flow void sign of the ventriculocisternostomy

SSAC is one of the major causes for the progression of clinical symptoms and signs. Presenting symptoms range between obstructive hydrocephalus, visual changes, endocrine abnormalities and head bobbing [2,25]. Among the endocrine disorders which are reported to be associated with up to 60% of cases of SSAS [25], sexual precocity is frequent.

The pathogenesis of precocious puberty in SSAC is debatable. It is known that hypothalamus is sensitive to pressure and SSAC involving the anterior portion of the third ventricle may destroy the posterior hypothalamus leaving the anterior hypothalamus intact [4,26]. The intact anterior hypothalamus, in the absence of inhibitory influences, leads to increased pituitary function [4].

Thus, premature activation of the hypothalamic-pituitary-gonad axis is a result of the disruption of the neural pathway that normally inhibits the GnRH pulse generator and has been postulated as a causative mechanism [27]. Other mechanisms such as hyperinsulinism and/or overweight may also be partly responsible for the precocious puberty [7].

Hydrocephalus and dilatation of the third ventricle were present in certain patients with GnRH-dependent sexual precocity and was thought to cause damage to the hypothalamus [7]. In our case, we could not confirm whether the etiology of the patient's precocious puberty was the arachnoid cyst or/and hydrocephalus, because both were found on the brain MRI.

A computerized tomography (CT) scans and especially MRI should be performed in all patients. Neuro-imaging features obtained by CT and/or MRI are helpful to eliminate other differential diagnosis of lesions that may develop in the suprasellar area with precocious puberty [26,28-30] and in deciding about therapeutic management. CT scans depict arachnoid cysts as non-enhancing extra-axial hypodense lesions with sharp borders [12]. On MRI the cysts have low signal intensity on T1-WI and a high signal on T2-WI with the cyst fluid having the same signal characteristics as CSF. There is no enhancement of any part of the lesion after contrast agent administration [8].

Patients with asymptomatic suprasellar arachnoid cysts should be routinely followed with serial MRIs, endocrine laboratories and comprehensive ophthalmological evaluations [6,31]. Symptomatic suprasellar arachnoid cysts are managed surgically. There is no consensus as to operative procedure of choice in SSAC. Endoscopy, by communicating the cyst with the ventricular system and/or basal cisterns, is currently the best available treatment for SSAC. This communication should be as large as possible to prevent secondary closing [32].

Endocrinological disorders may frequently persist following treatment despite the satisfactory decrease in volume of the cyst [25]. Then, gonadotropin releasing hormone (GnRH) analogue may be an effective adjuvant treatment in suppressing the hypothalamic-pituitary-gonadal axis in children with precocious puberty. Several researchers have documented a regression of clinical signs of development of puberty in response to treatment. This Therapy should be prescribed continuously until the age for the onset of natural puberty [10].

An earlier treatment seems to be more profitable in regression of signs and development of puberty and can minimize their physical and psychosocial problems and enhance their quality of life [33]. Due to non availability of this treatment, clinical, biological and radiological monitoring was performed in our patient after endoscopic approach. Despite that, regression of pubertal signs, which is exceptional, was observed during the clinical follow-up.

4. CONCLUSION

Early detection of precocious puberty in children is important and should lead to neuro-radiologic investigations in order to eliminate suprasellar intracranial tumors. SSAC is one of the uncommon etiologies of precocious puberty. Endoscopic intervention is the treatment of choice of these cysts with few complications. It allows resolutions of clinical symptoms and stabilization of endocrinological problems.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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