Clinical Brief

Arachnoid Cyst with Growth Hormone Deficiency

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ABSTRACT

On clinical grounds, arachnoid cysts are usually associated with neurological dysfunction. Little is known concerning their involvement in endocrine disorders. A seven-year-old boy was admitted to the hospital for evaluation of an unprovoked afebrile seizure. His neurological examination was normal, however, he had growth retardation. Insulin tolerance and L-dopa growth hormone stimulation tests revealed an inefficient growth hormone response. An MRI of hypophysis and cranium yielded a shift of hypophysis and a large arachnoid cyst. **[Indian J Pediatr 2007; 74 (3) : 294-296]** *E-mail: aycanunalp@mynet.com*

Key words: *Arachnoid cyst; Seizure; Growth retardation; Growth hormone deficiency.*

Arachnoid cysts are usually accompanied by neurological dysfunctions clinically. Little is known about their involvement in endocrine disorders.[1] A suprasellar arachnoid cyst may cause disorders of growth, puberty and hypothalamic-pituitary function, due to the proximity of the cyst to the hypothalamic-pituitary area.[2] The authors recently encountered a rare case of large suprasellar arachnoid cyst in a child who presented to our hospital with afebrile seizures and was incidentally detected to have growth retardation. In this report of a rare suprasellar arachnoid cyst the discussion is focused on clinical manifestations of arachnoid cysts.

CASE REPORTS

A seven-year-old boy was admitted to the emergency service upon an afebrile seizure. In his anamnesis, the patient had a seizure for about 20 min while awakening as focusing his eyes on a particular point and having generalized muscle contractions all over his body. On physical examination he was conscious, in good general condition and he did not have any neurological deficit. The patient weighed (15 Kg) less than -2 SDS and his height (104 cm) was less than -3 SDS. Normal mean and SD values were calculared usind Tanner, Whitehouse and Takaishi Cross-sectional-type Standarts for Supine-Length (up to 2) and Height Attained, boys/girls. Pubertal staging of the patient was Tanner 1. In his history, he was born at home *via* normal vaginal delivery. His family history revealed nothing special but a healthy mother and a healthy father, both of 33 years of age. There was no consanguinal wedding history. Blood biochemistry: glucose, urea, and ions in blood and liver and kidney function tests were in normal ranges. No pathology was detected with electroencephalography (EEG) during sleep or consciousness. The patient received afebrile seizure diagnosis and started receiving 10 mg/Kg/day carbamazepine; one week later the dose was increased to 15 mg/Kg/day.

He was consulted by endocrinologists for growth retardation since his height and weight percentilles were 2 SDS below than normal. His height age and bone age were 4.3/12 years and 6.5/12 years, respectively, where targeted height was 168 ± 6 cm (-0.96 SDS). Both mother's and father's heights were 158 cm (-0.7 SDS) and 165 cm (-1.4 SDS), respectively. In this patient, depending on the aforementioned findings, a height reduction due to a systemic disease was suggested. Thyroid function tests (fT3: 2.8 ng/ml, fT4: 1.9 ng/ml, TSH: 0.8 μIU/mL) were normal, whereas insulin tolerance test (ITT) and L-dopa growth hormone (GH) stimulation tests yielded a peak GH response of 2 ng/ml. Hence, a GH deficiency was diagnosed. Other hypophyseal hormone levels were normal: FSH: 1.7 mlU/ml (1.5-12.4), LH:1.1 mlU/ml (1-11.4), PRL: 21 mlU/ml (3-24), HCG: 0.4 ng/mL (<1), ACTH: 12 pg/ml (10-15), CORT: 24 (10-25) pg/ml. Hypophyseal magnetic resonance imaging (MRI) revealed 'a shift of hypophysis towards brain stem'. Cranial MRI revealed a suprasellar arachnoid cyst, which was hypointense on T1-weighted image and hyperintense on T2-weighted images with mild hydrocephalus accompanying a shift of mesencephalon and pons (Fig. 1, 2).

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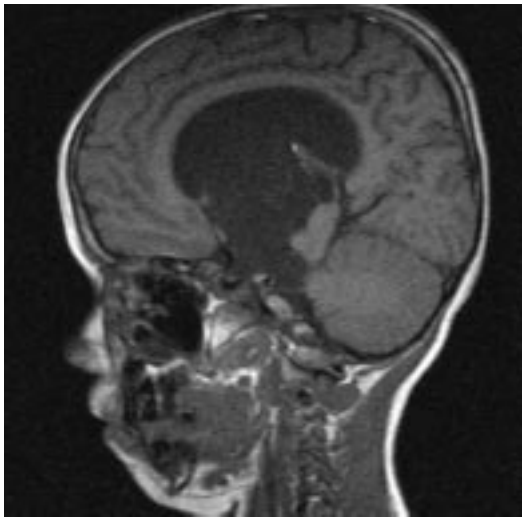

Fig. 1. Suprasellar Arachnoid Cyst, with Mild Hydrocephalus Accompanying a Shift in Mesencephalon and Pons.

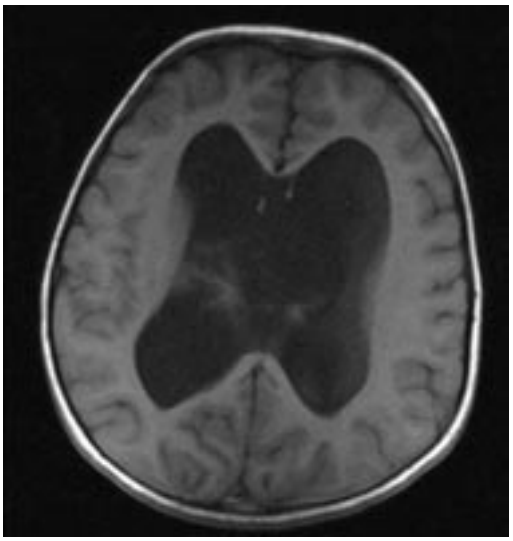

Fig. 2. Suprasellar Arachnoid Cyst, Which was Hypointense on T1-Weighted Image and Hyperintense on T2-weighted Images with Mild Hydrocephalus.

Endoscopic cysto-ventriculostomy and cysto-cisternostomy were conducted by neurosurgeons. No neurological deficits were observed post-operatively. Constructive interference in steady-state (CISS) fusion imaging with MRI revealed the presence of CSF flow. The patient was assigned to GH medication by endocrinologists. He is currently under follow-up.

DISCUSSION

Arachnoid cysts are congenital lesions of unknown etiology and are caused by a reduplication of the arachnoid membrane within which fluid collection results in a cyst.[3] The etiology of the cysts is controversial, and has been attributed to congenital malformation, infection, trauma, or increased intracranial pressure. The cysts are most commonly located at the sylvian or choroidal fissure.[4] Neurological deficits occur following cyst expansion, local compression and obstruction of CSF flow.

Suprasellar arachnoid cysts (SSACs) are uncommon lesions, accounting for 9% of all intracranial arachnoid cysts.[5] In most cases, cysts are not associated with progressive symptoms related to raised intracranial pressure, and frequently are detected incidentally or following hemorrhagic complications due to minor trauma. Clinical manifestations include signs of obstructive hydrocephalus, visual impairment, endocrine dysfunction, gait ataxia and "bobble-head doll" syndrome. Endocrine dysfunction may occur in the form of precocious puberty, amenorrhoea, developmental delay, skeletal growth retardation or hypothalamic disturbances. The frequency of this dysfunction is not well defined.[6] The other hypothalamic-pituitary disorders that have been reported are thyroid and cortisol hormone deficiencies, galactorrhoea or hyperprolactinemia.[7] Expansion of arachnoid cysts is thought to be one of the major causes for the progression of clinical symptoms and signs.[8]

Arachnoid cysts are frequently encountered in association with various neurological disorders, including epilepsy. A number of anecdotal reports have yielded inconsistent conclusions about the relationship of arachnoid cysts and epilepsy. A recent study has shown that epilepsy surgery in selected patients with temporal lobe cysts does not significantly reduce seizure frequency.[9] Although some reports do not directly relate these cysts to occuring epileptic seizures, the hydrocephalus coinciding with cyst progression and lack of recurrence of seizures following neurosurgery suggested that compression by the cyst might have caused the seizure in our patient.

Sexual precociouty is the most common endocrine disorder associated with suprasellar arachnoid cysts.[10] The co-existence of gonadotropin-releasing hormone (GnRH)-dependent sexual precociouty and GH deficiency in patients with arachnoid cysts are rarely reported, and their pathogeneses are not well established.[2, 11] The way in which the SSAC causes disorders of puberty and hypothalamic-pituitary dysfunction is unknown. It causes deficiencies of GH, TSH-releasing hormone and ACTH, however, it also stimulates the hypothalamic-pituitary-gonadal axis leading to central precocious puberty.

There are also few cases of growth hormone deficiency reported in this situation.[2, 12] Isolated GH deficiency was also observed in some patients with chronic hydrocephalus, the reason of which was suggested as the compression of the hypothalamus or pituitary by increased pressure.[13] Only isolated GH deficiency, but not precocious puberty was detected in the present case. Such

a large cyst caused growth retardation without signs of neurological deficits may be attributable to its slow rate of expansion. The hypothesis that GH deficiency in their patient might be caused by the pressure effect of expanding arachnoid cyst on hypothalamus and pituitary stalk is also in good agreement with the cases guested in the literature.[13]

Hence, evaluation of growth and sexual development in patients with arachnoid cysts during follow-up visits is recommended.[2] Bone age study and GnRH test are indicated when an abnormal progression of puberty is suspected. On the other hand, it is equally important to examine the GH status in these patients despite normal growth so that they can receive appropriate therapy as soon as possible to prevent compromise of their final height.[11]

In conclusion, the authors decided to publish their data from a patient in whom growth retardation and arachnoid cyst were determined while being hospitalized for evaluation of an afebrile seizure because arachnoid cysts are rare causes of growth retardation. They wanted to attract attention for the importance of systemic physical examination simultaneously with growth progression during the follow-up of patients having seizures accompanied by arachnoid cysts.

REFERENCES

1. Mohn A, Schoof E, Fahlbusch R, Wenzel D, Dorr HG. The endocrine spectrum of arachnoid cysts in childhood. *J Pediatr Neurosurg* 1999; 31 : 316-312.
2. Adan L, Bussieres L, Dinand V, Zerah M, Pierre-Kahn A, Brauner R. Growth, puberty and hypothalamic-pituitary function in children with suprasellar arachnoid cyst. *Eur J Pediatr* 2000; 159 : 348-355.
3. Krawchenko J, Collins GH. Pathology of an arachnoid cyst: Case report. *J Neurosurg* 1979; 50 : 224-228.
4. Passero S, Filosomi G, Cioni R, Venturi C, Volpini B. Arachnoid cysts of the middle cranial fossa: A clinical, radiological and follow-up study. *Acta Neurol Scand* 1990; 82: 94-100.
5. Harsh NGR, Edwards MSB, Wilson CB. Intracranial arachnoid cysts in children. *J Neurosurg* 1986; 64: 835-842.
6. Gupta SK, Gupta VK, Khosla VK, Dash RJ, Bhansali A, Kak VK, Vasishta RK. Suprasellar arachnoid cyst presenting with precocious puberty : report of two cases. *Neurol India* 1999: 2; 148-151.
7. Baskin DS, Wilson CB. Transsphenodal treatment of non-neoplastic intrasellar cysts. *J Nerosurg* 1984; 60: 8-13.
8. Mohn A, Schoof E, Fahlbusch R, Wenzel D, Dorr HG. The endocrine spectrum of arachnoid cysts in childhood. *Pediatr Neurosurg* 1999; 31: 316–321.
9. Koch CA, Voth D, Kraemer G, Schwarz M. Arachnoid cysts: Does surgery improve epileptic seizures and headaches*?* *Neurosurg Rev* 1995; 18: 173-181.
10. Sugawara A, Ebina K, Ohi H, Sawataishi J, Akabane M, Itoh T, Kikuchi K, Sakamoto T. Suprasellar arachnoid cyst associated with precocious puberty: report of an operated case and review of the literature. *No Shinkei Geka* 1992; 20 : 1107-1112.
11. Huang HP, Tung YC, Tsai WY, Kuo MF, Peng SF. Arachnoid cyst with GnRH-dependent sexual precocity and growth hormone deficiency. *Pediatr Neurol* 2004; 30 : 143-145.
12. Pierre-Kahn A, Capelle L, Brauner R, Sainte-Rose C, Renier D, Rappaport R *et al*. Presentation and management of suprasellar arachnoid cysts. Review of 20 cases. .*J Neurosurg* 1990; 73 : 355–359.
13. Hochhaus F, Butenandt O, Schwarz HP, Ring-Mrozik E. Auxological and endocrinological evaluation of children with hydrocephalus and / or meningomyelocele. *Eur J Pediatr* 1997; 156 : 597-601.