Suprasellar Arachnoid Cyst Presenting with Amenorrhea and Galactorrhea

Amenore ve Galaktore ile Seyreden Suprasellar Araknoid Kist

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Received : 10.2.2000 ⇔ Accepted : 6.7.2000

Abstract: The authors report a case of large suprasellar arachnoid cyst in a 32-year-old woman. The patient presented with amenorrhea and galactorrhea, which were both caused by hyperprolactinemia. These are rare symptoms among the various endocrinologic features that can accompany suprasellar arachnoid cysts. The magnetic resonance imaging findings and surgical options are discussed.

Key words: Amenorrhea, arachnoid cyst, galactorrhea, suprasellar cyst

Özet: Büyük bir suprasellar kisti olan 32 yaşındaki bir kadın bildirilmektedir. Hasta hiperprolaktinemiye bağlı olduğu saptanan amenore ve galaktore şikayeti ile kabul edilmişti. Bunlar, suprasellar araknoid kistlerde görülen değişik endokrinolojik bulgular arasında oldukça nadir olan semptomlardır. Olgunun MRI bulguları ve olguya cerrahi girişim yolları tartışılmıştır.

Anahtar sözcükler: Amenore, araknoid kist, galaktore, suprasellar kist

INTRODUCTION

Infants and young children who develop suprasellar arachnoid cysts (SSAC) usually exhibit abnormally large head circumference due to obstructive hydrocephalus, whereas these lesions in adults tend to cause headaches and visual impairment through compression of the optic chiasm (10). The symptoms of SSAC can include endocrine dysfunction, particularly precocious puberty (2, 6, 7, 12, 14, 15, 17). In this report, we describe a case of SSAC in an adult who presented with the uncommon endocrinologic features of amenorrhea and galactorrhea.

CASE REPORT

A 32-year-old woman presented with a 4-year history of amenorrhea and galactorrhea. She also reported dizziness problems for the past year. The patient had given birth 5 years prior to presentation. One year later, her menstrual periods became irregular and she developed galactorrhea. A work-up showed that her prolactin level was twice the upper-normal limit (40 ng/ml; range 3.0 - 20.0 ng/ml). Bromocriptine was prescribed, but the patient still had endocrinological symptoms upon admission to our hospital. Apart from the prolactin findings, the

patient's medical history was unremarkable. She showed no abnormalities on physical and neurological examination admission. Cranial magnetic resonance imaging (MRI) demonstrated a suprasellar cystic mass. The lesion was similar to cerebro-spinal fluid (CSF) in signal intensity, and appeared to have expanded the suprasellar cistern and displaced the optic chiasm superiorly (Figure 1). There was no solid tissue in the cistern. The hypophyseal stalk could not be identified, but the sella and the pituitary gland were normal. On the basis of the MRI findings, we performed visual evoked potential (VEP) testing and found that amplitude was slightly decreased on the recordings. Cranial computerized tomography (CT) revealed a suprasellar cystic mass with density comparable to that of CSF (Figure 2). Diffusion MRI showed that the apparent diffusion coefficient of the contents of the cystic structure was also similar to that of CSF (Figure 3). All these imaging findings were compatible with the diagnosis of SSAC.

Right pterional craniotomy was performed and sylvian fissure was dissected. Carotid cistern was exposed. Beneath the optic nerve we observed a greyish form that was presumed to be the cyst. We resected a soft grayish-white sample of the cyst membrane for biopsy, and fenestrated the cyst such that it communicated with the basal cisterns. Histological examination of the specimen showed that the cyst had a fibrous wall with a thin nonvascular subepithelial stroma. Lining the cavity

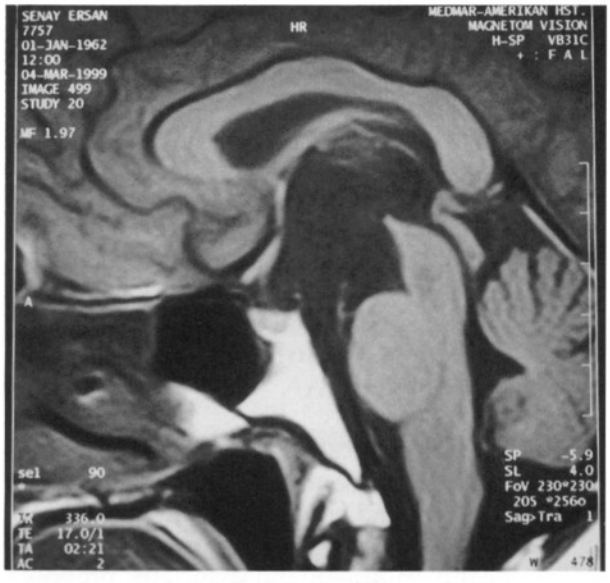

Figure 1: A sagittal T1-weighted image (TR/TE=460/15 ms) shows enlargement of the suprasellar cistern and displacement of the optic chiasm.

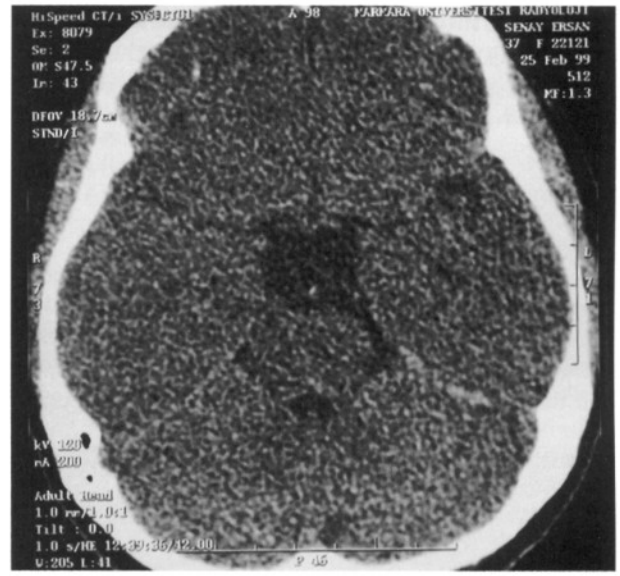

Figure 2: Axial CT shows a cystic lesion with density similar to that of CSF causing expansion of the suprasellar cistern.

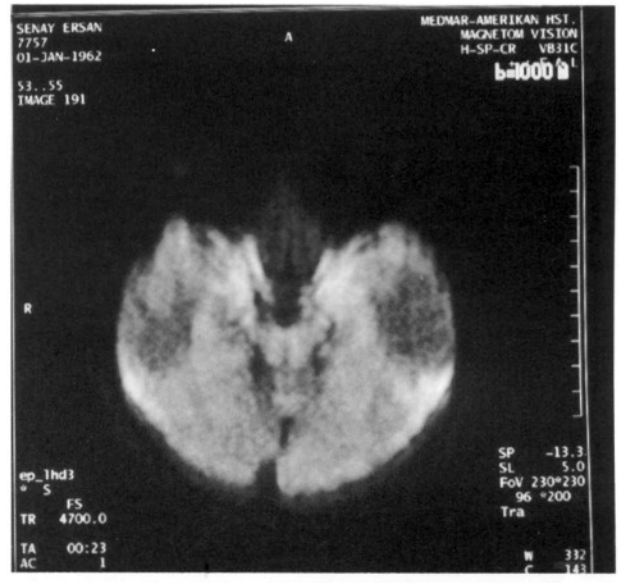

Figure 3: Diffusion MRI revealed that the apparent diffusion coefficient of the lesion was similar to that of CSF, indicating the cystic nature of the mass. The findings confirmed the diagnosis of arachnoid cyst.

was a single layer of flattened meningoepithelial "cap" cells. The pathological diagnosis was arachnoid cyst (Figure 4). There were no postoperative complications. Two months after the surgery, menstrual cycle turned normal again and galactorrhea disappeared. 2 months after the operation prolactin level dropped significantly, to

21 ng/ml. 5 months after the surgery she was in good health. Follow-up MRI at 12 months after the surgery showed that the suprasellar cistern decreased in size, and the displacement of the optic chiasm was less noticeable (Figure 5).

DISCUSSION

The prevalence of SSAC among all types of arachnoid cyst is approximately 10% (16). These lesions develop within the chiasmatic cistern. As they increase in size, they compress the hypothalamus and third ventricle from below, encroach on the optic chiasm, and stretch the optic nerves and the pituitary stalk (4). Affected individuals typically present with symptoms resulting from mass effects on neighbouring structures (1).

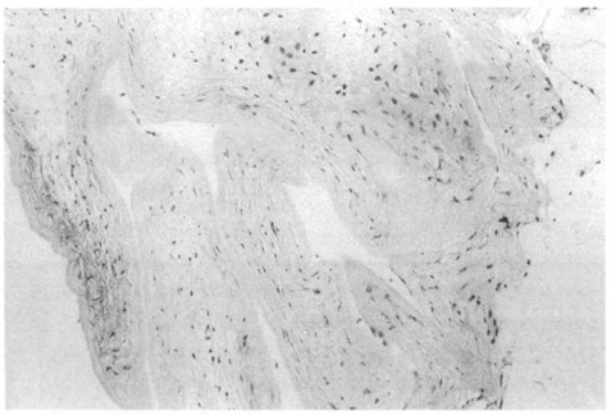

Figure 4: A photomicrograph of the cyst wall shows flattened epithelium of arachnoid cap cells composing the cavity lining.

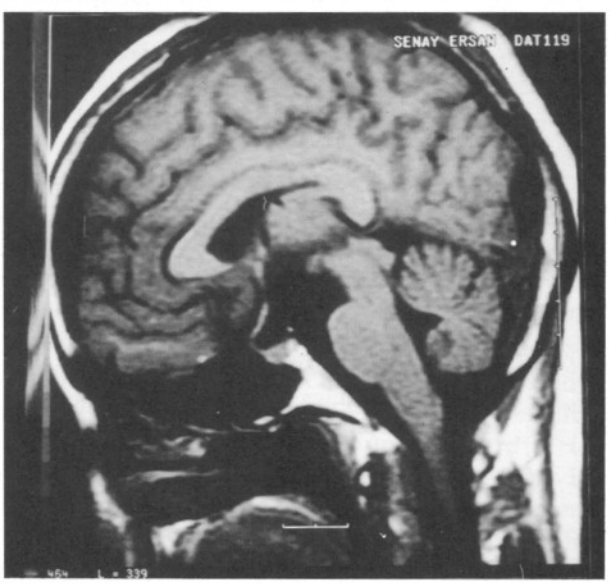

Figure 5: A postoperative sagittal T1-weighted image (TR/TE=460/15 ms) confirms reduction in the size of the suprasellar cistern and less obvious displacement of the optic chiasm.

In reviewing 54 cases, Hoffman et al (7) reported hydrocephalus as the most common finding in SSAC. Related symptoms of headache and increased head circumference were often noted as well. These authors labelled hypopituitarism as a rare manifestation that was usually seen in older children. Hydrocephalus was not found in our patient and headache was not her significant complaint, but she did have symptoms that suggested possible hypothalamic involvement.

Amenorrhea is a common symptom in hypothalamic diseases (3); however, it is hardly ever seen in cases of SSAC. This symptom is caused by either hypopituitarism or hyperprolactinemia. In their investigation of diseases of the hypothalamus, Ferrari et al. (5) reported only one case of SSAC that presented with amenorrhea due to hypopituitarism. That patient's final diagnosis was isolated gonadotrophin deficiency.

In our case, hyperprolactinemia was the sole hormonal abnormality, and was causing the amenorrhea and galactorrhea symptoms. Her symptoms persisted during bromocriptine therapy, however those symptoms resolved 2 months after the surgery. London and colleagues (11) described a similar case, but their case had severe frontal headaches in addition to amenorrhea-galactorrhea syndrome.

Few reports in the literature indicate optic nerve or optic chiasma involvement in SSAC, and visual field defects have been noted in these cases(9,16). Our patient had no history of visual disturbance, but her MRI results led us to test her visual function. A perimetry test was normal, but the VEP recordings showed slightly reduced amplitude to indicate optic nerve dysfunction. Still, the patient was asymptomatic.

The goal in surgical management of SSAC is to open the membrane and achive communication between the cyst and either the ventricular system or the basal cisterns (12). In the absence of hydrocephalus, Hoffman and associates proposed that the subfrontal approach was the only surgical option that would permit the surgeon to achieve adequate communication between the cyst and the

basal cistern. However, some reports have highlighted the risk of damage to the olfactory nerve by this technique (8), in addition to the potential dangers of open surgery (13). Using a pterional craniotomy approach, we were able to fenestrate the cyst such that it communicated effectively with the basal cisterns. There were no complications, and the follow-up results were satisfactory.

The clinical presentations of SSAC and the treatment options available are well documented in the literature (6,13). The purpose of this report is to point out our observation of an unusual presentation of SSAC and to present MRI findings in this case. We also indicate that cyst fenestration through a pterional craniotomy is a viable treatment option for SSAC.

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REFERENCES

1. Amagasa M, Sato H, Niizuma H, Suzuki J : Trigeminal neurinoma associated with suprasellar arachnoid cyst: case report. Neurosurgery 19:454-457, 1986
2. Brauner R, Pierre-Kahn A, Nemedy-Sandor E, Rappaport R, Hirsch JF: Precocious puberty caused by a suprasellar arachnoid cyst.Analysis of 6 cases. Arch Fr Pediatr 44:489-493, 1987
3. Christy NP, Warren MP: Disease syndromes of the hypothalamus and anterior pituitary, in De Groot LJ, Cahill Jr GF, Odell WD (eds), Endocrinology, New York: Grune and Stratton, 1979: 215-252
4. Di Rocco C: Arachnoid cysts, in Youmans JR (ed), Neurological Surgery, volume 2, third edition, Philadelphia: WB Saunders, 1990: 1299-1325
5. Ferrari C, Crosignani PG: Hypothalamic disease presenting as amenorrhea: value of computed tomography. Br J Obstet Gynaecol 92:1251-1257, 1985
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 47:148-151 , 1999
7. Hoffman HJ, Hendrick EB, Humpreys RP, Armstrong EA : Investigation and management of suprasellar arachnoid cysts. J Neurosurg 57:597-602, 1982
8. Jones RFC, Warnock TH, Nayanar V, Gupta JM: Suprasellar arachnoid cysts: management by cyst wall resection. Neurosurgery 25:554-561, 1989
9. Kaisho Y, Miyazaki S, Shimo-oku M, Hayashi T, Tani E: A case of suprasellar arachnoid cyst followed up for a long time. Nippon Ganka Gakkai Zasshi 99:109-114, 1995
10. Leo JS, Pinto RS, Hulvat GF, Epstein F, Kricheff II: Computed tomography of arachnoid cysts. Radiology 130:675-680, 1979
11. London SN, Olive DL: Arachnoid cyst as a cause of the amenorrhea-galactorrhea syndrome. Am J Obstet Gynecol 149:87-88, 1984
12. Ozek MM, Pamir MN, Erzen C: A combined surgical approach to a suprasellar arachnoid cyst with precocious puberty. Turkish Neurosurgery 2:187-189, 1992
13. Pierre-Kahn A, Capelle L, Brauner R, Sainte-Rose C, Reiner D, Rappaport R, Hirsch JF: Presentation and management of suprasellar arachnoid cyst. J Neurosurg 73:355-359, 1990
14. Schroeder HW, Gaab MR: Endoscopic observation of a slit-valve mechanism in a suprasellar prepontine arachnoid cyst :case report. Neurosurgery 40:198-200, 1997
15. 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 20:1107-1112, 1992
16. Takahashi T, Kawai S, Kaminoh T, Hiramatsu K, Maekawa M, Yuasa T, Miyamoto N, Hattori Y: Suprasellar arachnoid cyst, report of a case. Neurol Surg (Tokyo) 10:435-441, 1982
17. Turgut M, Ozcan OE: Suprasellar arachnoid cyst as a cause of precocious puberty and bobble-head doll phenomenon. Eur J Pediatr 151:76, 1992

Arachnoid cysts and neuroepithelial cysts constitute the second most frequent type of mass lesion able to compromise cerebrospinal fluid flow. They are most often encountered in the suprasellar region, the foramen magnum and the posterior fossa, where it is important that they must be distinguished from Dandy-Walker malformations.