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Case Report

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Intrasellar arachnoid cyst: a case report

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ABSTRACT

Cystic lesions of the parasellar region are not uncommon and masquerade as a tumor both radiologically and clinically. True intrasellar arachnoid cysts are rare lesions and can easily be confused with intrasellar pituitary neoplasms, craniopharyngiomas, Rathke's cleft cysts, or even empty sella. The preoperative and peroperative distinction is obviously important because of the different treatment strategies and different prognosis. A 44-years-old woman with presenting symptoms of headache and decreased visual acuity with the initial diagnosis of either Rathke's cleft cyst or intrasellar arachnoid cyst is reported. The evaluations for differential diagnosis, commonly encountered pathological entities, treatment modalities and pathogenesis of intrasellar arachnoid cysts were discussed.

Key words: arachnoid cyst, intracranial, MRI

Introduction

True arachnoid cysts of the sellar region are very rare, can easily be confused both clinically and radiologically wih intrasellar cystic pituitary neoplasms such as Rathke's cleft cysts, craniopharyngiomas or even empty sella. Adequate diagnosis is quite important because of different treatment modalities and different prognosis.

The intimate relationship of neural, endocrine, vascular, meningeal and skeletal tissues in the parasellar region provides a myriad of pathological possibilities in this small area. These pathological entities are broadly divided into neoplastic disorders both pituitary and non-pituitary origin and non-neoplastic disorders including non-neoplastic cysts, aneurysms and other vascular malformations, inflammatory disorders.

Non-neoplastic cystic lesions of the parasellar region have a fluid filled compartment and a connective tissue wall and/or epithelial lining but no communication with the subarachnoid space that distiquishes them from an empty sella. These lesions are classified according to the site of origin, tissue of origin and/or pathological features [1-3]. They are broadly classified into primary and secondary cysts. The primary lesions arise within the sella and may be confined to the pituitary fossa including true intrasellar arachnoid cysts, pituitary cysts, Rathke's cleft cysts and other rare types. Secondary non-neoplastic cystic lesions of the sella may arise from the sphenoid sinus or the parasellar region and extend into the fossa, as sphenoid sinus mucocele, arachnoid cysts, Rathke's cleft cysts and other rare pathologies.

Leptomeningeal or arachnoid cysts are lined by a single layer of mesothelial cells, surrounded by collagenous layer. An intrasellar arachnoid cyst represents an important component of the differential diagnosis of sellar region mass. Therefore we report our experience with a case emphasizing diagnosis, pathophysiology, treatment and outcome.

Case Report

A 44-year-old female was admitted to our hospital with presenting symptoms of headache, dizzeness for 5 years and decreased visual aquity lasting for 8 weeks. On admission, her neurological examination was normal. There was non-specific visual field defects in both eyes on perimetry and optic fundi were normal. She had no sign of increased intracranial pressure. Hormonal data including free T3, free T4, T3, T4, TSH, prolactin, FSH, LH, growth hormon, cortisol and ACTH were normal.

Skull X-ray showed balloon dilatation of sella turcica with destruction of sphenoid sinus floor. Computed tomography (CT) showed a water density intrasellar mass with sphenoid sinus extension, which was not enhanced with contrast medium (Figure 1). The lesion showed the same density as the surrounding subarachnoid space on T1-weighted magnetic resonans images (MRI) (Figure 2A-B). The pituitary gland was flattened against the posteroinferior sellar floor. There was no soft tissue component or contrast enhanced lesions on MRI except normal pituitary gland.

The indication for the surgery was the progressive growth of cyst and she was operated by transsphenoidal-rhinoseptal approach. Peroperative, it was found that the sellar floor was destroyed but the dura was intact. The CSF-like watery clear fluid in the cyst was evacuated and after the partial excision of the cyst wall, chiasmapexia

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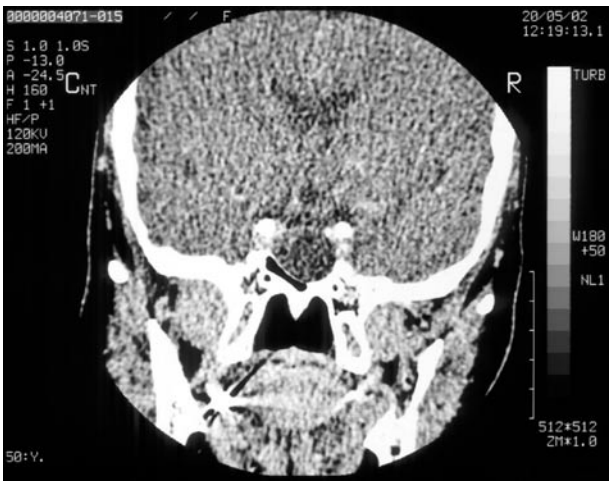

Figure 1 | Contrast enhanced computed tomography showed a water density intrasellar mass, which was not enhanced with contrast medium.

was performed with autologous fascia and fat tissue without any postoperative complication.

The histological examination of the cyst wall was composed of connective tissue containing cells with oval nuclei and a few capillaries. No adenomatous, cuboidal, columnar epithelial cells with cilia or goblet cell were present. Therefore, the histopathology of the cyst was compatible with arachnoid cyst.

The patient was discharged from the hospital on the 3rd post-operative day without any neurological deficit.

Discussion

Intracranial arachnoid cysts have becoming more frequent findings as a result of modern imaging techniques. They occur most commonly in the middle fossa and around the foramen magnum; only 9% of cases have been found to be located in the sellar and parasellar region [4-6].

Current options suggest that intrasellar arachnoid cysts appear to be acquired lesions as evidenced by the enlargement of sella [3, 7]. Almost all series reported that the mean age of presentation was older than other intracranial arachnoid cysts [2, 3]. Also, intrasellar arachnoid cysts are not associated with a normal cerebrospinal fluid (CSF) cistern in contrast to other intracranial cysts which communicates with [3, 4, 8].

An understanding of the mechanism of the development of arachnoids requires a brief review of the embryology of the subarachnoid space. In the early embryo, the neural tube is surrounded by a loose layer of mesenchymal tissue, called, 'perimedullary mesh'. It is tought to be precursor of the pia and arachnoid mater. After the rupture of the rhomboid roof, by the effect of the pulsative force of the choroid plexus, the CSF is flow into the layers of the perimedullary mesh. The differentiation of the perimedullary mesh into a superficial layer (the arachnoid mater) and a deeper layer (the pia mater) is the result of the flow of the CSF. It is thought to be that arachnoid cysts develop because of a minor aberration in the flow of CSF in the primordial stage of the subarachnoid space, resulting in the sequestration of an enclosed chamber within the perimedullary mesh.

Several theories have been proposed to explain the pathogenesis of true intrasellar arachnoid cysts. One of the theories suggest that arachnoid herniates into the pituitary fossa in the presence of incompetent diapragma sella and the communication with the subarachnoid space (SAS) either closes of or behaves as a one-way valve [3, 4, 8, 9].

Other mechanism suggest that arachnoid lined CSF containing space include a true intrasellar origin arising from arachnoid rests below the diaphragma or from duplication of arachnoid which herniated into the fossa through a low attached diagraphma sella [3, 4, 8, 10]. Enlargement believed to occur by a single route process, a ball-valve like phenomenon in which the cerebrospinal fluid gain enterence into the cyst without exit with the effect of pulsative intracranial pressure [3, 8].

Symptoms of arachnoid cyst are similar to that of other intrasellar space occupying lesions. They may enlarge sufficiently to cause parasellar extension; enroachment

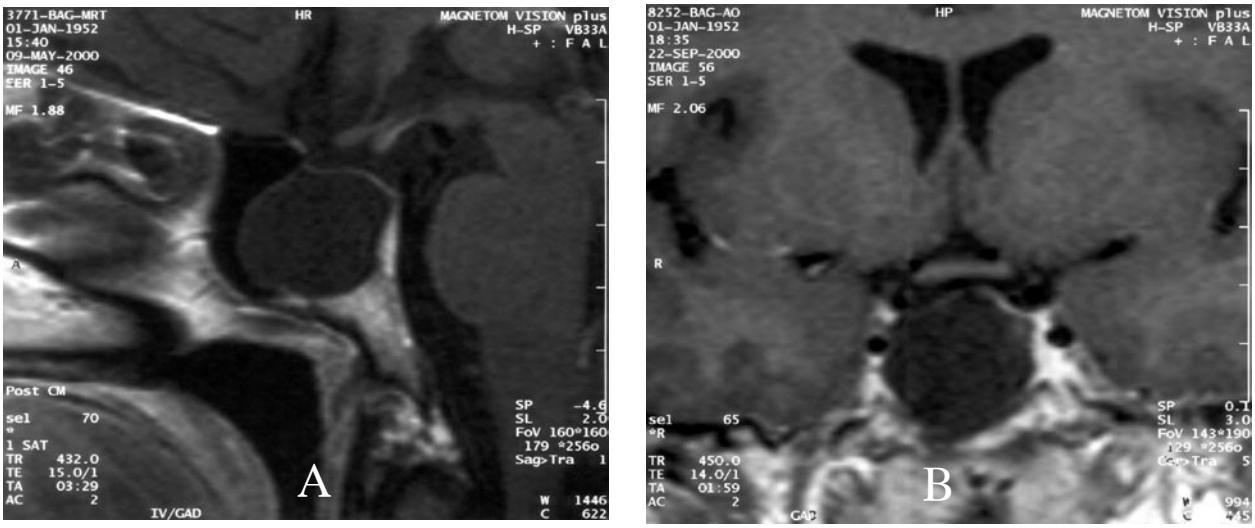

Figure 2 | The lesion showed the same density as the surrounding subarachnoid space on contrast-enhanced T1–weighted magnetic resonans images (A-B).

into the cavernous sinus and the suprasellar located optic chiasm, hypothalamus or foramen of Monro. Most of the patients presented with headache whereas only 70% of symptomatic patients have visual field defects, decreased visual acuity or hypopituitarism symptoms [2, 11-13].

Radiological features of an arachnoid cyst are very similar to that of other cystic neoplasms of sellar region whether benign or malignant it may not be easy to distinguish the type on various modalities preoperatively. The typical appearence of a cystic lesion on skull X-ray is enlarged sella with or without erosion and displacement of the dorsum sella. CT scans typically show a cystic lesion within the sella. The arachnoid cyst has the density in Hounsfield units idetical to CSF without any enhancement, shows often suprasellar extension [2, 3].

As the development of MRI, differential diagnosis has become easier depending on intensity of cyst content and cyst wall enhancement pattern. However, because of the similar radiological features, differential diagnosis between arachnoid cyst and Rathke's cleft cyst is not easy even on MRI. Rathke's cleft cyst with CSF intensity is common up to 44% [3, 5, 14]. Cyst walls are often not enhanced neither in arachnoid cysts nor Rathke's cleft cysts. The position of pituitary gland varies in cases of Rathke's cleft cyst, however, so far it was reported that in arachnoid cyst cases the flattened pituitary gland was compressed and displaced posteroinferiorly [3, 15-17]. Intrasellar arachnoid cyst must be strongly considered in the differential diagnosis of intrasellar cystic lesions that showing CSF pattern and compress the pituitary gland posteroinferiorly.

Significant percentages of patients having a low-density parasellar lesion are asymptomatic (20%) at the time of diagnosis and may not reqire surgery [3]. In symptomatic patients, the aim of surgery in non-neoplastic parasellar cystic lesion is to obtain the histological diagnosis and releive the mass effect upon intrasellar and parasellar structures.

The choice of surgical approach is either transsphenoidal surgery or craniotomy [18]. Most of the non-neoplastic cystic lesions can be dealt with by transsphenoidal surgery with drainage and biopsy of cyst wall. Lesions with huge suprasellar extension may require craniotomy; in most of them simple drainage and biopsy is again sufficient [1-3].

As the outcome of the surgery, headache becomes less severe and rare in most of the patients whereas the endocrinologic outcome does not fare well patients with arachnoid cyst [1, 13]. The long-standing pulsative pressure in the arachnoid cyst perhaps causes more intrinsic arachnoid damage than does the simple expansive pressure produced by other groups of cysts [1].

The major complication of the transsphenoidal surgery is CSF rhinorhea that usually follows an accidental arachnoid tear or intentional opening of the arachnoid membrane at the chiasmatic cistern for suprasellar procedures [1, 16]. Refilling of the cystic lesion is another postoperative problem and recurrence rate was reported as 20% for arachnoid cysts [13].

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