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Suprasellar Arachnoid Cyst with Spontaneous Intracystic Hemorrhage: A Rare Complication – Role of MR and Illustration of a Case

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Summary

Background:

Arachnoid cysts are congenital or developmental intra-arachnoidal CSF-filled lesions, which develop probably as a result of splitting or duplication of the arachnoid membrane. Most of them are asymptomatic and are detected as incidental findings on Computed Tomography or Magnetic Resonance Imaging of the head carried out for other reasons. Although complications such as intracystic, subdural, and extradural hematomas are well known after a trauma, spontaneous hemorrhage in an arachnoid cyst is a rare and serious complication with atypical imaging features on cross-sectional imaging and only less than ten cases are documented in the literature till date, with none of them in the suprasellar location.

Case Report:

A 40-year-old female patient presented with history of headache since two months, which was sudden in onset, holocranial.

Conclusions:

Spontaneous intracystic hemorrhage is an uncommon and serious complication of arachnoid cysts, which can give rise to atypical features on imaging. Therefore familiarity with this rare complication is indeed essential.

MeSH Keywords:

Arachnoid Cysts • Intracranial Hemorrhages • Magnetic Resonance Imaging

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Background

Spontaneous intracystic hemorrhage in an arachnoid cyst is a rare complication (although hemorrhagic arachnoid cyst is well known in the clinical setting of trauma), which may present with symptoms of raised intracranial pressure (headache, vomiting, altered sensorium) or focal neurological deficits depending on the location and is an indication for surgery [1,2]. We present a case of suprasellar arachnoid cyst with spontaneous intracystic compartmental haemorrhage in an adult woman. To the best of our knowledge few cases of this rare complication, with none in suprasellar location have been described in the literature so far [2].

Case Report

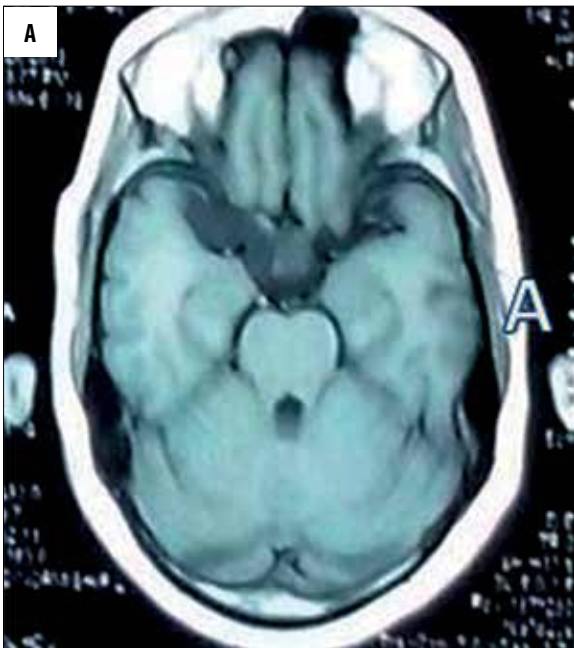
A 40-year-old female patient presented with a two-month history of headache, which was sudden in onset, holocranial and dragging in nature and not associated with vomiting.

There was no history of trauma/fever/seizures/weakness in upper or lower limbs or any memory disturbances. General physical examination and neurological examination were normal. Routine blood investigations including platelet count and coagulation tests were also normal.

Computed Tomography (CT) of the head was done which showed a well-defined extraaxial mixed density lesion in the suprasellar region (Figure 1). Further Magnetic Resonance Imaging (MRI) of the head was carried out for better characterization of the lesion and to know its exact extent. MRI showed a well-defined, lobulated lesion in the sellar and suprasellar region extending along the anterior skull base (greater wing of sphenoid) towards the right side. The lesion showed mixed signal on T1-weighted sequence (Figure 2A, 2B) and it was hyperintense on T2-weighted (Figure 3) and FLAIR sequences. There was no evidence of restricted diffusion (Figure 4). It showed evidence of a thin septum, and a part of the lesion on left side of the septum



Figure 1. Axial section from a non-contrast CT of the head showing a well-defined, extra-axial lesion of attenuation of CSF in the suprasellar region extending towards the anterior skull base on the right side. A part of the lesion on the left side shows evidence of increased density.



was hyperintense on T1-weighted images (Figure 2A, 2B). That area also showed smooth, thin, peripheral enhancement, although the rest of the lesion was non-enhancing (Figure 2C). Those features were suggestive of hemorrhage in a compartment of the cyst. The lesion was causing mass effect in the form of elevation of optic nerves and chiasma, and displacement of the infundibulum towards the right side. The pituitary gland was normal. Differential diagnosis of arachnoid cyst was given.

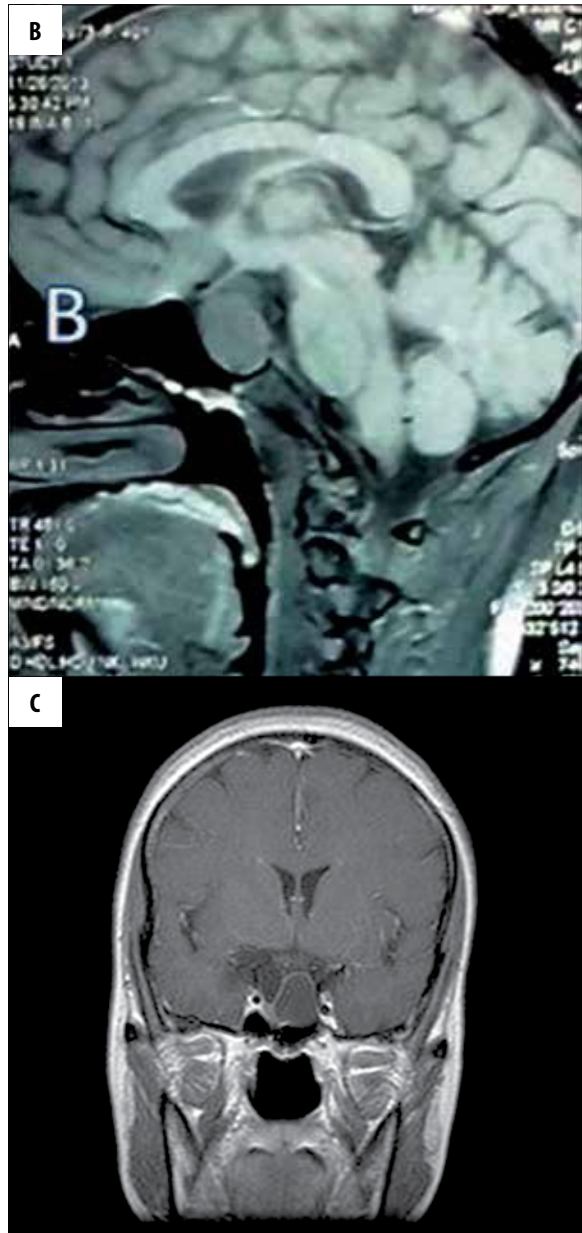


Figure 2. Axial (A) and sagittal (B) T1-weighted MR images of the brain showing a hypointense signal similar to CSF in major portion of the sellar and suprasellar lesion with a thin septum. A part of the lesion on the left side of the septum shows a hyperintense signal. Coronal post-contrast T1-weighted image (C) of the brain showing non-enhancement in major portion of the lesion with smooth, thin rim enhancement in the left-sided locule.

Via right sub-frontal approach cyst wall was near totally excised leaving behind the part attached to pituitary stalk and tuberculum sellae. Cyst was also punctured at the time of surgery and light straw colored fluid was aspirated (Figure 5A, 5B).

A sample was sent for histopathological examination. Histopathology showed the cyst wall lined by meningotheelial cells along with fibroconnective tissue (Figure 6A, 6B). Immunohistochemistry showed positive staining with EMA (Figure 6C) and vimentin (Figure 6D). Those features were



Figure 3. Axial T2-weighted sequence of MRI showing uniform hyperintense signal within the lesion with a thin septum.



Figure 4. Axial diffusion-weighted image showing evidence of increased diffusivity within the lesion.

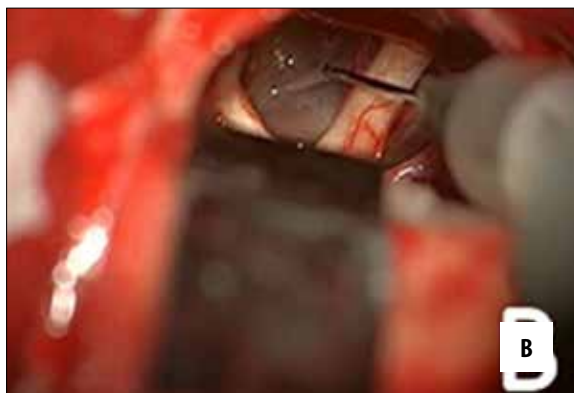


Figure 5. Peroperative photograph showing cyst visible in the inter optic and optico carotid space (A) and straw colored fluid being aspirated (B).

suggestive of arachnoid cyst. Postoperatively the patient's general condition was fair, vitals were stable with no neurological deficits. Postoperative CT showed a craniotomy defect in the right frontal bone with no evidence of residual lesion. She was continued on maintenance steroids and was advised on a follow-up.

Discussion

Arachnoid cysts are developmental, intraarachnoidal CSF-filled lesions, which comprise almost 1% of all non-traumatic intracranial mass lesions. Although they can be found in the suprasellar region (as in our case), the cerebral convexity, cerebellopontine angles, cisterna magna, quadrigeminal plate cistern and interhemispheric fissure, their most common location is Sylvian fissure in the middle cranial fossa [1,3].

Many theories have been postulated for etiopathogenesis of arachnoid cysts. Various hypotheses are: entrapment of CSF in a diverticulum; CSF flow changes leading to arachnoid

cell layer tears during the formation of various cisternae; during embryological separation (at around 15th week of gestation) of arachnoid from the dura mater [2].

Usually they remain stable in size and are asymptomatic, however, a few cysts contain remnants of the choroid plexus or arachnoid granulations leading to secretion of CSF resulting in an increase in size with time. These cases may present with features of compression of adjacent structures (Kallman syndrome, precocious puberty, bitemporal hemianopia in suprasellar lesions, cranial nerve palsies etc.) and/or raised intracranial pressure due to their large size or hemorrhage. Spontaneous hemorrhage is supposed to be due to a minor trauma with rupture of intracystic or bridging vessels [1,2]. We propose intracompartmental rupture of a bridging vessel over the left locule of arachnoid cyst as the cause of bleed in our case.

On imaging, uncomplicated arachnoid cysts are well-defined, extra-axial, non-calcified and non-enhancing

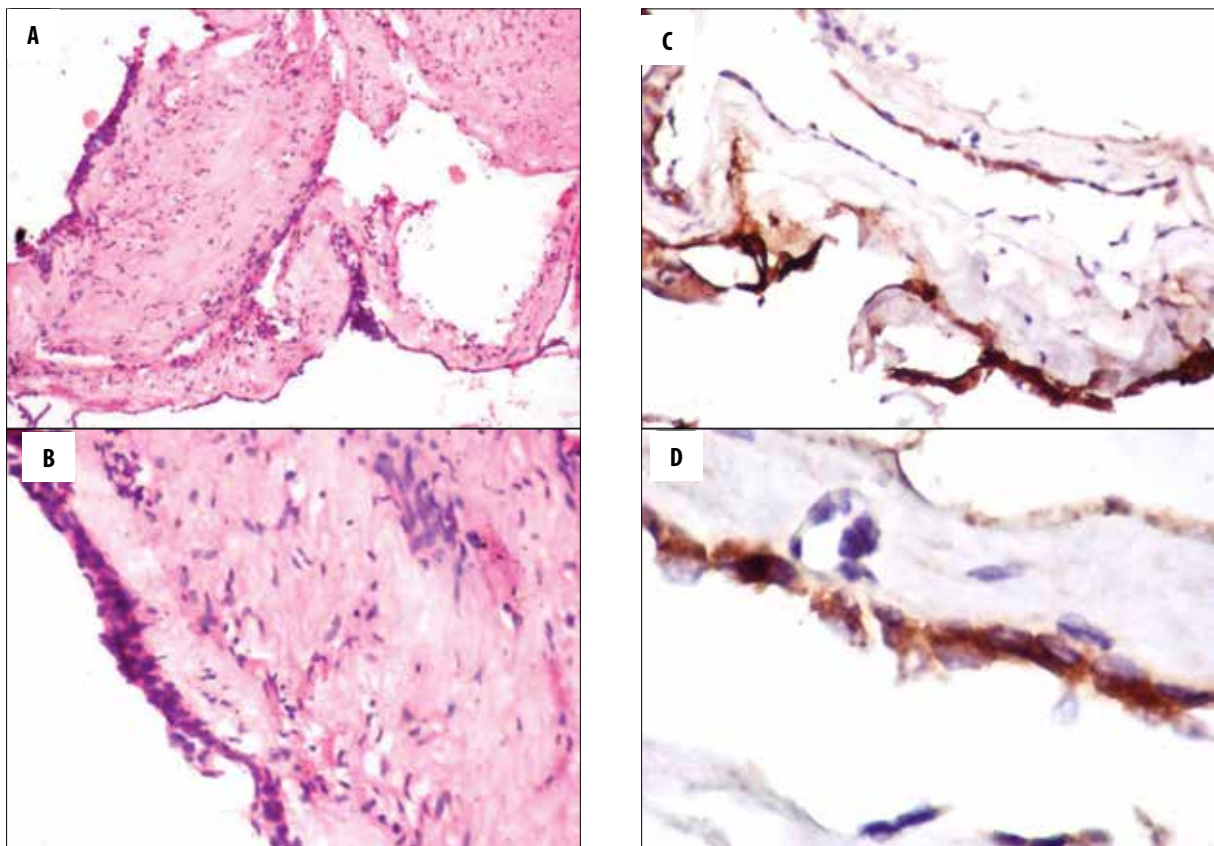


Figure 6. Low power view (100×) (A) and high power view (400×) (B) showing evidence of meningotheial cells within the cyst wall along with fibro-connective tissue. Immunohistochemistry showed positive staining with EMA (epithelial membrane antigen) in meningotheial cells (C) and with Vimentin (D).

lesions with CSF-like attenuation and signal intensity on CT and MRI (in all sequences), respectively. However, hemorrhagic arachnoid cysts show varied signal intensity on MR depending on the stage of bleeding. MR is superior to CT in detecting hemorrhage except in acute cases, where it can present as hyperdensity in such cysts [4,5].

Sometimes a fluid-blood level can also be visualized in these lesions (as in our case) due to sedimentation of red blood cells into a CSF-filled cyst [1].

The most common differentials of arachnoid cysts include epidermoid and cystic neoplasms. Epidermoid can be differentiated on the basis of characteristic diffusion

restriction and cystic tumors due to enhancing solid component. Other uncommon differentials are: parasitic cysts, Rathke cleft cysts etc. but they don't show evidence of hemorrhage [1].

Conclusions

Spontaneous intracystic hemorrhage is an uncommon and serious complication of arachnoid cysts, which can give rise to atypical features on imaging. Therefore, familiarity with this complication and its imaging features is essential and this entity should be excluded in all cases of symptomatic arachnoid cysts.

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