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Case Report

Prepontine arachnoid cyst presenting with headache and diplopia: A case report study

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Abstract

Background: Arachnoid cysts are found everywhere in cerebrospinal axis, most often in the middle cranial fossa. They are very rare in prepontine location.

Case Description: In this study, we report a 26-year-old female presenting with a 3-month history of headache and diplopia. On physical examination, she had clinical manifestations of sixth cranial nerve palsy. Magnetic resonance imaging revealed a prepontine arachnoid cyst with extension into interpeduncular and suprasellar cisterns. Computed tomography scan demonstrated no evidence of hydrocephalus. The patient was treated surgically by endoscopic fenestration of the cyst with endonasal transsphenoidal approach. The cyst was opened to prepontine, interpeduncular, and suprasellar cisterns.

Conclusion: Endoscopic endonasal fenestration of the cyst to adjacent cistern may be safe in prepontine arachnoid cysts with sellar and suprasellar extension; it may be effective and less invasive compare to transcranial approach.

Key Words: Arachnoid cysts, diplopia, prepontine arachnoid cyst, retroclival arachnoid cyst, sixth cranial nerve palsy

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INTRODUCTION

Arachnoid cysts are benign, fluid-filled, cyst-liked malformations related to the arachnoid mater.[1,27] They can be formed everywhere of cerebrospinal axis with preference in the middle cranial fossa.[12] Arachnoid cysts involving the posterior fossa are less common and are most often found in the cerebellopontine angle (CPA);[17,29] however, they are very rare in retroclival or prepontine location.[20,22] Extending superiorly to the interpeduncular cistern and laterally to the CPA is usually seen in retroclival or prepontine arachnoid cysts, and variable cranial nerve involvement may be found depending on their extensions.[20]

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CASE REPORT

A 26-year-old female presented with a 3-month history of headache and diplopia. Gait problems, different types of seizures, and endocrine dysfunctions were not found. She had no history of any disease in the past. On physical examination, she had clinical manifestations of sixth cranial nerve palsy. Other neurological examinations were normal.

Magnetic resonance imaging (MRI) revealed a prepontine arachnoid cyst extending into the interpeduncular and suprasellar cisterns along with brainstem compression [Figure 1]. No evidence of hydrocephalus on computed tomography (CT) scan was visible [Figure 2].

The patient was treated surgically by endoscopic fenestration of the cyst with endonasal transsphenoidal approach. The cyst was opened into the prepontine, interpeduncular, and suprasellar cisterns [Figure 3]. The incidence of postoperative cerebrospinal fluid (CSF) leak and meningitis after endoscopic endonasal surgery has been estimated to be 11% and 10%, respectively.[16] To prevent these complications, the base of the skull was reconstructed by fat, fascia lata, and mucosal flap, and a lumbar drain was placed for 3 days. It is likely that the risk and the rate of recurrence of the cyst in both endoscopic and transcranial methods are the same. However, it needs to be assessed in future studies.

Postoperatively, she had impressive improvement in the function of sixth cranial nerve, and was discharged with no complications. Follow-up MRI showed a decrease in the size of the cyst and decompression of the adjacent tissues [Figure 4].

We reported this case due to rare occurrence of prepontine arachnoid cyst, nonclassic clinical manifestation in this case, and the different surgical approach that we adopted to treat the cyst. We discuss these concerns below.

DISCUSSION

Arachnoid cysts are benign, fluid-filled, cyst-liked malformations related to the arachnoid mater.[1,27] They can be formed everywhere on the cerebrospinal axis with preference in the middle cranial fossa.[12] Several mechanisms have been considered in the formation of arachnoid cysts such as splitting or duplication of the arachnoid membrane,[21] trauma,[8] and genetic factors.[4] Most are developmental anomalies. Acquired form of arachnoid cysts are associated with neoplasms, leptomeningitis, hemorrhage, or surgery.[25] It has been estimated that intracranial arachnoid cysts account for approximately 1% of intracranial space occupying lesions.[9] The prevalence of these lesions has been estimated 1.4% in adults[1] and 2.6% in children.[2] They are thought to be more common in males than in females.[1] Arachnoid cysts are most often found in the middle cranial fossa, retrocerebellar, and cerebral convexity, respectively. Cysts in middle fossa and retrocerebellar locations are less likely to be considered symptomatic despite their more common prevalence.[1] They often result in nonspecific symptoms such as headache, impaired cognition, or dizziness. Nevertheless, they may present with specific sensorimotor symptoms.[13,28] Surgical treatments of these lesions include fenestration of the cysts wall to create a communication between the cyst and normal subarachnoid space which can be achieved by open or endoscopic surgery.[19,26] Furthermore, stereotactic puncture, cystoperitoneal shunting, and radical resection of the cyst are some other surgical methods to treat arachnoid cysts.[5-10] Decompression of arachnoid cysts by surgical treatments yields significant clinical benefit associated with a low risk of severe complications.[14,18]

Furthermore, there have been some case reports regarding the atypical presentations of arachnoid cyst. In 1992, Bourekas *et al.* reported a case of a 54-year-old woman who presented with a 6-month history of ataxia,

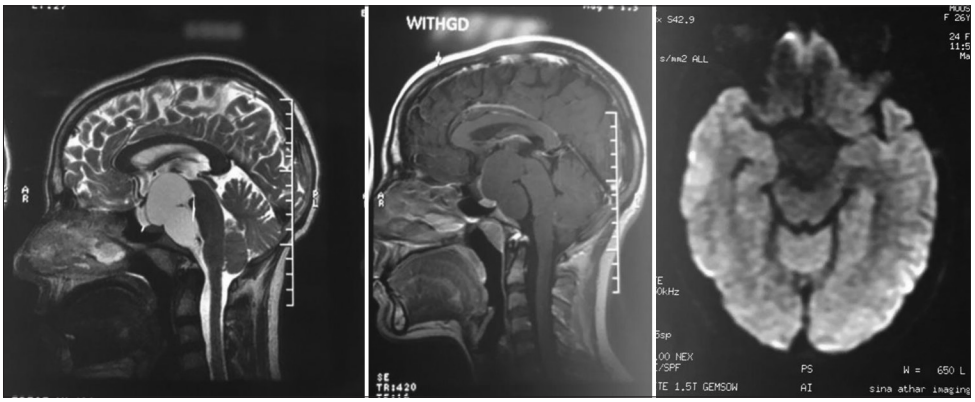

Figure 1: Preoperative magnetic resonance imaging. The T2-weighted sagittal image (left) shows a large hyperintense retroclival cystic mass extending into interpeduncular and suprasellar cisterns, causing compression and posterior displacement of brain stem. The T1-weighted postgadolinium sagittal image (middle) shows a nonenhancing mass. Diffusion weighted image (right) demonstrates a mass lesion in interpeduncular cistern without restricted pattern

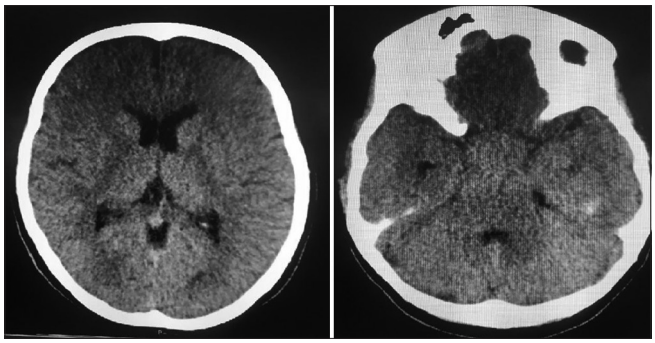

Figure 2: Computed tomography scan demonstrates no evidence of hydrocephalus

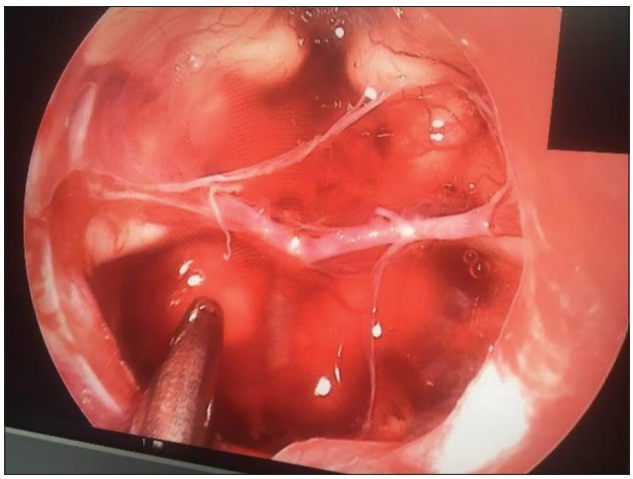

Figure 3: Intraoperative transnasal endoscopic view after fenestration of the cyst into adjacent cisterns

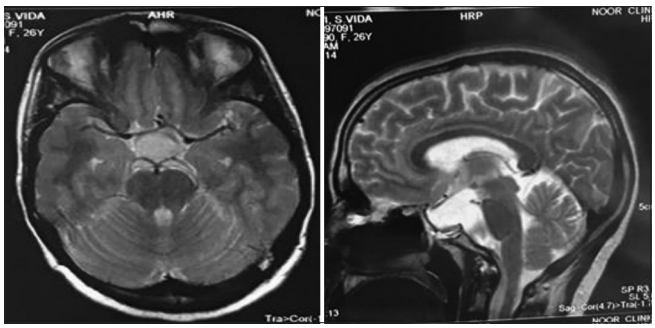

Figure 4: Postoperative magnetic resonance imaging. The T2-weighted axial (left) and sagittal (right) images show cyst shrinkage and decompression of brain stem

quadraparesis, headaches, intermittent slurred speech, and urinary incontinence. She was also suffering from a progressive 10-year history of decreasing hearing in the right ear. MRI revealed a large extra-axial retroclival mass extending from the posterior suprasellar region into the upper cervical spinal canal and into both CPAs, right being greater than left. Compression and posterior displacement of the brainstem was noted. She was treated surgically by fenestration of the cyst through a right retromastoid craniectomy and C1 laminectomy.[6]

In 2008, De Bonis *et al*. reported a case of a 64-year-old man with a clinical history of gait disturbance and urge incontinence. Radiologic studies showed evidence of a supratentorial normal pressure hydrocephalus, with enlarged pericephalic spaces caused by a 4-cm prepontine arachnoid cyst.[11]

In 2008, Bonde *et al*. presented a 26-year-old woman with a 10-year history of right hemifacial spasm controlled by botulinum injections. On examination, she had right sensorineural hearing loss, right hemifacial spasm and ataxia. MRI showed a retroclival cyst vertically extending all the way from the suprasellar region, over the clivus up to the level of the body of the axis. It also extended into the both cerebellopontine angles, more on the right side. She underwent retrosigmoid craniectomy in the sitting position and wide marsupialization of the cyst was performed.[7]

In 2009, Akyuz *et al*. reported a case of a 40-year-old woman who had a sudden onset of occipitocervical pain and vomiting for a week. MRI revealed a retroclival cystic lesion complicated with intracystic hemorrhage. The cystic lesion was treated surgically by a right lateral suboccipital approach.[3]

In 2014, Rao *et al*. reported a case of 9-year-old boy with a history of precocious puberty of several months. MRI revealed a giant 51 × 52 × 52-mm well-circumscribed suprasellar arachnoid cyst. They concluded that suprasellar arachnoid cysts may present with endocrinopathies months to years prior to development of visual symptoms.[23]

In 2015, Ramesh *et al*. presented three cases with bobble-head doll syndrome (anteroposterior bobbling of the head and neck on the trunk every 2–3 seconds) associated with a large suprasellar arachnoid cyst and obstructive hydrocephalus, which were treated with endoscopic cystoventriculocisternostomy and marsupialization of the cyst.[24]

Our patient presented with unilateral (left) abducens nerve palsy reported for the first time as a clinical manifestation of arachnoid cysts. Although arachnoid cysts are often described as cystic masses with a MRI signal intensity similar to CSF on all sequences, such as T1-weighted, T2- weighted, and diffusion-weighted images, the cyst in our patient was isointense on T1-weighted, hyperintense on T2-weighted and nonrestricted on diffusion-weighted sequences. We could not locate any such case in the literature. The fluid of an uncomplicated arachnoid cyst without intracystic hemorrhage or infection is indistinguishable from CSF (low protein), thus the intensity of the cyst fluid is similar to CSF on all MRI sequences. Neoplastic, inflammatory, and hemorrhagic cysts (complicated cysts) have a greater amount of protein and cellular debris resulting in different intensities compared to CSF on

at least one sequence.[15] The report of pathology of the cyst in our patient was arachnoid cyst. However, we assessed neither intracystic hemorrhage nor intracystic inflammation.

CONCLUSION

For patients presenting with headache or other neurological symptoms who have occupying lesions in suprasellar and prepontine or retroclival locations in further evaluation, arachnoid cysts should be considered as a differential diagnosis which can be only followed up in asymptomatic cases because of their benign nature; whereas in symptomatic cases, different surgical methods can be considered as the most effective treatment. In prepontine arachnoid cysts with sellar and suprasellar extension, endoscopic endonasal fenestration of the cyst to adjacent cistern may be safe, effective, and less invasive compared to the transcranial approach.

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Conflicts of interest
There are no conflicts of interest.

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