

An unusual location and presentation of a usual pathology-Colloid cyst of fourth ventricle presenting as spontaneous rhinorrhea

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Abstract

Colloid cysts are relatively rare benign intracranial lesions preferentially located within the third ventricle. There are only a few reports in which they have been found to be ectopic, such as in the fourth ventricle. A young female presented on with spontaneous non-traumatic cerebrospinal fluid (CSF) rhinorrhoea for three months which was positional in nature, relieved temporarily by neck flexion. Magnetic resonance imaging (MRI) scan showed a focal well-defined rounded cystic lesion along the fourth ventricle, showing subtle peripheral rim enhancement. Significant hydrocephalus was also noted. A suboccipital craniotomy and total excision of the lesion was done. Postoperatively, the patient recovered quickly with no neurological deficits. Her rhinorrhoea was completely cured. Histopathology was consistent with a colloid cyst. Colloid cyst is rarely found in infratentorial location. However, such a rare diagnosis has to be considered in the differential diagnosis in patients who present with an infratentorial cystic lesion associated with spontaneous CSF rhinorrhoea.

Keywords: Colloid cyst, Fourth ventricle cyst, Infratentorialcyst.

Introduction

Colloid cysts are relatively rare benign intracranial lesions that are preferentially located within the rostral aspect of the third ventricle. There are only a few reports in which colloid cysts are described in other locations such as the fourth ventricle, pituitary gland, leptomeninges, cranial vault, brain-stem, cerebellum and the suprasellar region.¹ A report described a case of a colloid cyst of the velum interpositum in a young woman.² These cysts account for 0.2% to 2% of all intracranial neoplasms. They are slow-growing benign tumours, the majority of which are located in the third ventricle.³ In the present report, we describe the radiological features of a case of infratentorial colloid cyst.

Case Report

A 29-year-old female presented to the Radiology

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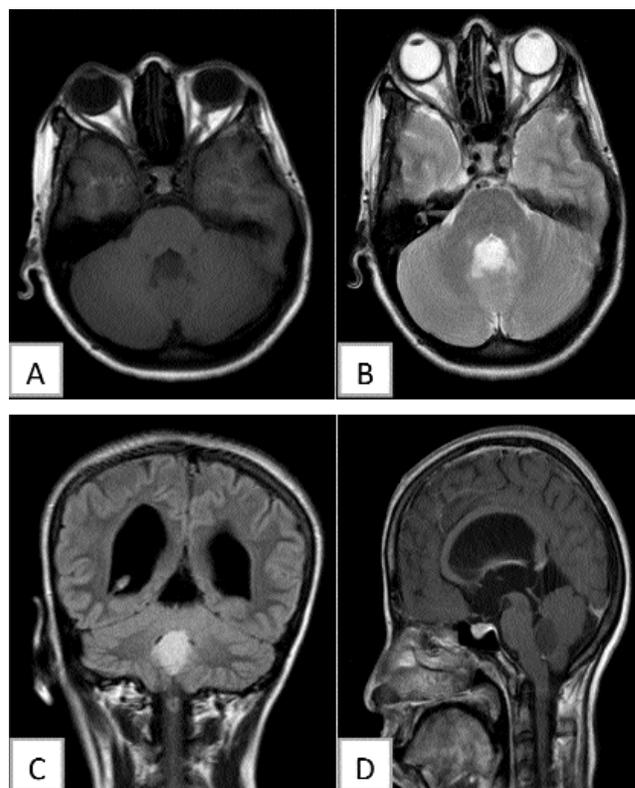


Figure: 29-year-old female with spontaneous rhinorrhoea. Unenhanced T1-weighted (a), T2-weighted (b) axial, FLAIR coronal (c) and contrast enhanced T1-weighted sagittal (d) magnetic resonance image of brain dated April 2013 show well-defined cystic lesion in the fourth ventricle with peripheral enhancement.

Department of Jinnah Postgraduate Medical Centre (JPMC), Karachi, on April 7, 2013, with history of intermittent spontaneous cerebrospinal fluid (CSF) rhinorrhoea for three months which was postural in nature, triggered by flexing her neck and relieved by changing the neck position. On examination, she had no neurological deficits. Contrast-enhanced magnetic resonance imaging (MRI) scan of brain showed a focal well-defined abnormal signal intensity area along the distal half of the fourth ventricle. It appeared iso-intense on both T1-weighted and T2-weighted MRIs and hyperintense on fluid-attenuated inversion recovery (FLAIR) images (Figure-A-D). It showed subtle peripheral rim

enhancement on post-contrast MRI sequences. No restriction was noted on diffusion-weighted scans. No signal dropout was seen on venographic blood oxygenation level-dependent (VENBOLD) sequences. Significant obstruction to the CSF outflow was noted resulting in marked hydrocephalus. Marked flattening of the tectal plate was noted due to cerebral aqueductal dilatation and dilated perimesencephalic cisterns. The cyst was removed endoscopically and its microscopic examination revealed amorphous eosinophilic material lined by a single layer of cuboidal and ciliated columnar cells. These were strongly positive for periodic acid-Schiff (PAS), epithelial membrane antigen (EMA) and cytokeratin. The patient improved symptomatically with no neurological deficits. At the third-month follow-up visit, there was no complaint of rhinorrhoea and the patient did not show any signs of raised intracranial pressure.

Discussion

Colloid cysts are benign tumours of the central nervous system (CNS) that can be found anywhere throughout the neuroaxis, but are preferentially located within the rostral aspect of the third ventricle between the columns of the fornices at the foramina of Monro. These are frequently attached to the stroma of the choroid. Cysts lined by a single layer of epithelium are a rare occurrence within the posterior cranial fossa. Colloid cyst belongs to the same group and its wall is built up of a collagenous connective tissue stroma, lined with a single layer of epithelium. Almost all varieties of cells have been reported in a colloid cyst may it be columnar, ciliated, non-ciliated, cuboidal, pseudo-stratified or squamous.⁵ Exact germline derivation of a colloid cyst has remained a debate although it has been suggested that these cysts are derived from the non-degenerated rudiments of the embryonic paraphysis. Neuroepithelial, endodermal and primitive foregut endoderm origin have also been considered for the pathogenesis due to the ultra-structural similarities. When these cysts are encountered outside the ventricular system and in the cortical areas their origin from primitive ectopic glial tissue in the subarachnoid space has been proposed. Similarities in histological features with the Rathke' cleft cysts and enterogenous cysts have also been found in some of the studies.⁶ Therefore, histopathological analysis remains the gold standard diagnostic method for these lesions. These cysts consist of amorphous gelatinous material that reacts positively to PAS staining. They stain positively for cytokeratin and EMA. They are negative for GFAP and Prealbumin stains. Expression of CEA, Vimentin and S-100 varies.⁷ Most reported cases occurred in the third to

fifth decades of life with paucity of cases in early years. In majority of patients, colloid cysts are an incidental finding in an otherwise asymptomatic patient, coming in for neuroimaging of an unrelated reason. However, brief episodes of paroxysmal headache are a common finding due to intermittent obstruction of CSF flow by the pedunculated lesion suspended from the roof of the ventricle. These headaches are usually the first presenting symptom. They last for a few seconds to minutes and are basically positional in nature. Obstruction at the foramina of Monro can lead to acute hydrocephalus and tonsillar herniation, resulting in sudden death. Other less frequently encountered symptoms include progressive dementia, drop attacks, and spells of altered levels of consciousness. Children usually present with signs of raised intracranial pressure, including headache, nausea, vomiting, papilloedema, and diplopia.⁸ Spontaneous CSF rhinorrhoea is another rare presentation of a colloid cyst. In our patient, the only presenting symptom was positional spontaneous CSF rhinorrhoea lasting for 10-15 minutes and relieved by flexing the neck. A case has been reported of spontaneous rhinorrhoea in a patient with known 3rd ventricle colloid cyst.⁹ Three cases of non-traumatic CSF rhinorrhoea have been reported secondary to colloid cysts.¹⁰ Colloid cysts can markedly expand in size due to variety of causes e.g. by the secretion of an amorphous and protein-rich fluid, fluid entry into the cyst due to the osmotic effect of the high intracystic protein level, water diffusion from the surrounding tissues into the cyst and by active haemorrhage into the cyst. Any of these phenomena can cause rapid increase in the size of a colloid cyst resulting in acute obstruction of CSF flow at the foramen of Monro. Haemorrhage into a colloid cyst is another extremely rare phenomenon. One study has reported incidence of at least 9 haemorrhagic colloid cysts.^{11,12} On the contrary spontaneous regression of a colloid cyst is another possibility that has been reported. Altered levels of hydration have been implicated as a cause of this rare occurrence rather than frank rupture of the cyst wall.¹³ For asymptomatic patients with no hydrocephalus, observation with serial imaging is usually recommended. However, for symptomatic patients, the risk of sudden death and sudden neurological deterioration due to acute obstruction of CSF flow is significantly higher. Therefore, in these patients surgical options have to be considered. These usually include bilateral or unilateral shunting with simultaneous fenestration of the septum pellucidum, endoscopic fenestration, or removal of the cyst and suboccipital craniotomy for direct removal of the cyst by either a transfrontal or transcallosal approach.¹⁴ In our patient craniotomy with total excision of the cyst was done.

Conclusion

We reported a case of an infratentorial colloid cyst located within the fourth ventricle with a rare presenting symptom of spontaneous non-traumatic CSF rhinorrhoea. It was diagnosed by contrast-enhanced MRI of brain. Successful removal of the cyst was done. Histopathology well correlated with the features of a colloid cyst. Though rarely found in this location, such a rare diagnosis has to be considered in the differential diagnosis in patients who present with an infratentorial cystic lesion associated with spontaneous CSF rhinorrhoea.

References

1. Bladowska J, Bednarek-Tupikowska G, Biel A, S?siadek M. Colloid cyst of the pituitary gland: Case report and literature review. *Pol J Radiol* 2010; 75: 88-93
2. Hingwala DR, Sanghvi DA, Shenoy AS, Dange NN, Goel AH. Colloid cyst of the velum interpositum: a common lesion at an uncommon site. *Surg Neurol* 2009; 72: 182-4.
3. Müller A, Büttner A, Weis S. Rare occurrence of intracerebellar colloid cyst. Case report. *J Neurosurg* 1999; 91: 128-31.
4. Parkinson D, Childe AE. Colloid cyst of the fourth ventricle. Report of a case of two colloid cysts of the fourth ventricle. *J Neurosurg* 1952; 9: 404-9.
5. Elmadbouh H, Halpin SF, Neal J, Hatfield RH, Hourihan MD. Posterior Fossa Epithelial Cyst: Case Report and Review of the Literature. *AJNR Am J Neuroradiol* 1999; 20: 681-5.
6. Nomikos P, Buchfelder M, Fahlbusch R. Intra- and suprasellar colloid cysts. *Pituitary* 1999; 2: 123-6.
7. Paniraj GL, Panigrahi M, Reddy AK, Immaneni S. Suprasellar Colloid Cyst: An Unusual Location. *World Neurosurg* 2014; 81:1, 202.e15-202.e18.
8. Armao D, Castillo M, Chen H, Kwock L. Colloid Cyst of the Third Ventricle: Imaging-pathologic Correlation. *Am J Neuroradiol* 2000; 21: 1470-7.
9. Deepak KS, Kesavadas C, Kapilamoorthy TR, Menon G. Cerebrospinal fluid rhinorrhea and acquired anterior basal encephalocele in a patient with colloid cyst of the third ventricle. *Neurol India* 2010; 58: 156-8.
10. Kane PJ, Mendelow AD, Keogh AJ, Symon L. Cerebrospinal fluid rhinorrhoea associated with colloid cyst. *Br J Neurosurg* 1991; 5: 317-20.
11. Tamura Y, Uesugi T, Tucker A, Ukita T, Tsuji M, Miyake H, et al. Hemorrhagic colloid cyst with intraventricular extension. *J Neurosurg* 2013; 118: 498-501.
12. Ogbodo E, Kaliaperumal C, Bermingham N, O'Sullivan M. Spontaneous haemorrhage and rupture of third ventricular colloid cyst. *Br Med J* 2012; 3: 2012
13. Duncan G, Kanodia AK, Jamel SE. Spontaneous Enlargement then Regression of a Colloid Cyst of the III Ventricle. *Int Neuropsychiatr Dis J* 2014; 2: 28-33.
14. Hadjipanayis CG, Schuette AJ, Boulis N, Hao C, Barrow DL, Teo C. Full Scope of Options. *Neurosurg* 2010; 67: 197-203.