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Case Report
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Spontaneous Resolution of Neuroenteric Cyst Following Rupture and Chemical Meningitis: MR Findings

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Abstract

Neuroenteric cysts are congenital cysts commonly seen in the spine. It can however be seen intra cranially predominantly in posterior fossa. We present a rare case neuroenteric cyst which showed resolution following spontaneous rupture and chemical aseptic meningitis during the hospital stay awaiting surgery.

Introduction

Neuroenteric cysts are rare cystic tumor like lesions which embryo logically harbor heterotopic endodermal tissue and commonly seen in brain and spine [1,2]. These lesions may present with compressive symptoms and rarely with aseptic chemical meningitis due to leak or rupture [3,4]. We present a case of intracranial neuroenteric cyst, which spontaneously ruptured during hospital course, developed chemical aseptic meningitis and resolved on follow up imaging.

Case Presentation

A young lady of 35 years presented to us with complaints of persistent head ache since 10 days with episodes of vomiting, occipital headache worsening on cough. Clinical examination did not reveal any significant findings like raised intracranial pressure etc. Her all routine blood investigations were within normal limits.

Patient underwent MRI scan (SIEMENS AVANTA) with following sequences T1W axial and sagittal (TR, TE), T2W axial and coronal (TR and TE), FLAIR axial (TR, TE, TI), DWI axial (TR, TE, ETL), gradient sequence SWI (TR, TE, ETL), MPR sagittal (TR, TE) with contrast study (Magnevist 0.1 mmol/kg body weight dose).

MRI showed a well-defined cystic lesion in the pre medullary cistern extending inferiorly to the craniovertebral junction. It was seen to extend more to the left side with mass effect on the left side of medulla. The lesion appeared hyper intense on T2W and FLAIR sequence with subtle hyper intensity on T1 weighted images. No obvious contrast enhancement was noted. In view of the location and benign nature a diagnosis of Neuroenteric cyst was made (Figure 1).

Subsequently, patient was planned for surgical intervention. However during the hospital course, awaiting surgery, patient developed sudden onset of severe headache with neck rigidity.

Immediately, repeat MRI was done which revealed mild reduction in size of the cystic lesion with FLAIR hyper intensities in prepontine, perimesencephalic cisterns and also in frontoparietal sulci bilaterally. Post contrast there was significant diffuse meningeal enhancement. These findings suggested rupture of the cyst causing aseptic chemical meningitis (Figure 2). Patient was treated with steroids and following treatment she showed significant relief of symptoms.

Follow-up MRI brain done after one week revealed almost complete resolution of the Neuroenteric cyst with thin floating membranes in CSF space (Figure 3). Patient was clinically asymptomatic and discharged. Follow-up MRI could not be done since patient informed us on telephone that she was absolutely fine and not willing for repeat imaging.

Discussion

Neuroenteric cysts are rare tumor like cystic lesions seen in spine and brain harboring heterotopic endodermal tissue. In 1928, Kubie [1] first described it as teratomatous cysts and later as intestinomas by Puuseep [2] in 1934. In 1954, the current name of Neuroenteric cyst was given

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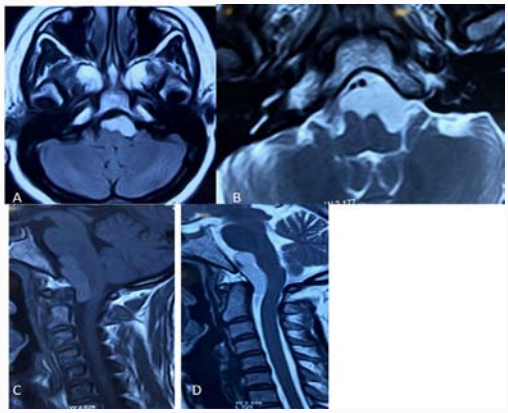

Figure 1: FLAIR image in A, shows hyper intensity of the cyst anterior to medulla, CISS 3D shows mild hypo intense cyst compared to CSF With well appreciated wall in B, T1W in sagittal image in C Shows mild hyper intensity and T2W sagittal image in D Shows mild hypo intensity compared to surrounding CSF.

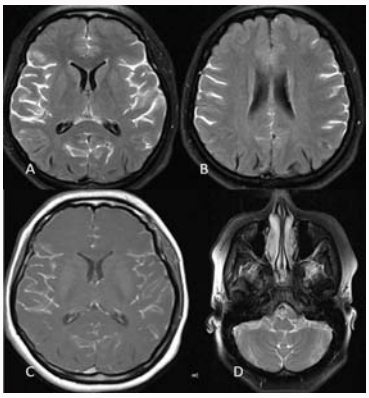

Figure 2: A and B shows sulcal FLAIR hyper intensity, C shows T1W post contrast with intense meningeal enhancement and T2W image in D shows reduced size of the cyst.

by Matson et al. [3].

These are benign cysts of foregut origin with endodermal lining from the incomplete dissolution of the neuroenteric canal. There are two theories which describe the origin of these cysts. First, at the time of notochordal development, failure of separation of notochord and foregut during the process of excalation leads to incorporation of endodermal cells with notochord which later becomes Neuroenteric cysts [4-6]. Second theory is of an endodermal diverticulum behind the oropharyngeal membrane called as Seesel pouch which may be a progenitor for midline supratentorial cysts [7,8].

These cysts are mostly seen in the spine but can be rarely seen intra cranially. They are commonly seen in posterior fossa, anterior to brainstem, frequently in midline and sometimes in cerebellopontine angle [9].

Histologically the intracranial Neuroenteric cysts are classified as mucin poor and mucin rich based on the content of mucin within the cyst. The mucin poor cyst shows pseudostratified ciliated columnar epithelium while the mucin rich cyst shows simple columnar epithelium [10].

Few important imaging features of neuroenteric cyst have been described. Also, the imaging characteristics vary as per the content of the cyst like the mucin, a proteinaceous secretion. On Computed tomography, Mucin rich cysts appear hyperdense and mucin poor appear mildly hypodense. In MRI imaging, the protein content tends to vary the signal characteristics on T1 and T2. The lesions appears hyper intense on FLAIR sequence, predominantly hyper intense on T2W and sometimes hypo intense and on T1W it appears hyper to isointense compared to CSF. Contrast study shows no obvious contrast enhancement except at the site where the cyst is adherent with brain parenchyma [10].

Neuroenteric cyst can be circular or lobulated and significantly hypo intense on T2W images due to presence of high protein content, squamous metaplasia and keratin debris. Presence of NAA like peak at 2.02 ppm on MR spectroscopy was considered specific for these cysts [11]. This NAA like peak is supposed to be secreted by cell lining of the cyst wall and contains perchloric acid or sialic acid bound to macromolecules resonating at 2.02 ppm [12,13].

The main differential diagnosis of neuroenteric cyst are arachnoid cyst, epidermoid cyst, dermoid cyst rarely Rathke and colloid cyst. However imaging features can differentiate between these cysts. Arachnoid cyst is of CSF intensity on all the MRI sequences, epidermoid cyst shows restricted diffusion, Rathke and colloid cyst have different site of location. The presence of N-Acetyl Aspartate (NAA) like peak at 2.02 ppm is also considered as specific finding in neuroenteric cyst which is absent in other cysts [11,14,15].

Similar imaging features were noted in our case. In addition we found heavily T2W sequence like CISS 3D very useful in differentiating these cysts. IN CISS sequence we can clearly define the wall of the cyst and it appears hypo intense compared to surrounding CSF with smooth wall. The wall of epidermoid cyst however can appear irregular or frond like on CISS sequence [16]. As a rare complication, these cysts can spontaneously leak slowly or rupture following trauma leading to aseptic chemical meningitis [17-19].

The recurrent aseptic meningitis is called as Mollaret

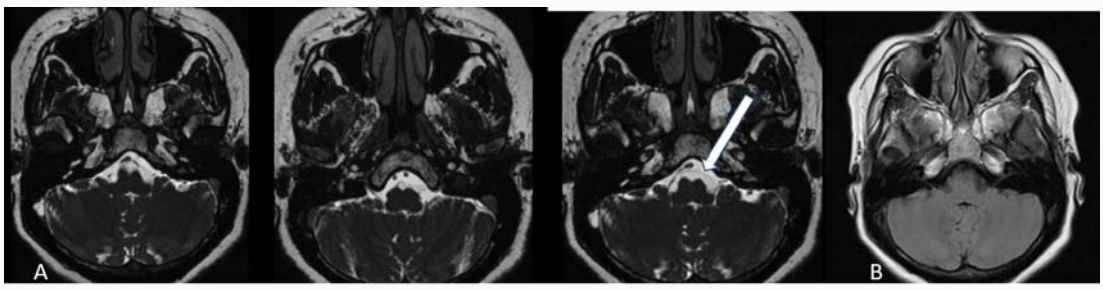

Figure 3: CISS sequence in A shows disappearance of the cystic lesion with thin floating membrane (arrow) and FLAIR in B shows no hyper intensity as was seen in Figure 1.

meningitis associated with CSF pleocytosis predominantly with polymorphonuclear cells and caused by virus however Mollaret meningitis due to slow leakage of epidermoid cyst are reported [20]. Similarly Mollaret meningitis is also reported due to slow leakage from neuroenteric cyst [21].

In a ruptured dermoid cyst, fat globules can be seen in the subarachnoid space as hyper intensities on T1 weighted images, subtle sulcal FLAIR hyper intensities and leptomeningeal enhancement on contrast study due to chemical meningitis [22]. Similar imaging picture can be seen in ruptured neuroenteric cyst. The chemical meningitis in neuroenteric cyst could be due to its enteric mucin contents [17,19,20].

In our case, patient initially presented with headache and vomiting probably suggesting leakage of neuroenteric cyst and aseptic meningitis however there was no obvious contrast enhancement on imaging. Subsequently, the neuroenteric cyst ruptured spontaneously during the hospital course causing chemical aseptic meningitis with sulcal and cisternal FLAIR hyperintensity and intense leptomeningeal enhancement. Following treatment with steroids, patient recovered completely within two days. Follow-up imaging showed near total resolution of the neuroenteric cyst. In our knowledge this is first case, showing near total resolution of the neuroenteric cyst following spontaneous rupture without any obvious trauma. We could not get the imaging follow up of the patient but telephonic follow-up suggested that patient was asymptomatic.

Conclusion

Clinical signs of headache and vomiting in a known case of neuroenteric cyst could be the warning sign of impending rupture and spontaneous rupture can occur leading to partial or complete resolution. Also CISS 3D sequence (heavily T2W) can be used additionally to further characterize the cystic lesions.

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