Extra Axial Hyperdense Neurenteric Cyst at Craniocervical Junction: A Diagnostic & Surgical Challenge

Razia Rehmani*
Department of Traumatology, USA
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*Corresponding author: Razia Rehmani, Department of Traumatology, Radiology, Neuroradiology, NYU Langone Medical Center, New York city, USA, Email: razia.rehmani@gmail.com

Abstract
Neurenteric cysts are congenital cysts of the central nervous system. They can present with focal or non-specific neurological signs. Neurenteric cysts can present variably on CT, depending on the intrinsic intracystic mucin content. They can be mistaken for other serious pathologies which can affect patient management. We present an interesting case of a young multi-parous woman who presented with new onset unilateral weakness (3/5). Non-contrast head CT demonstrated a hyper dense lesion anterior/right anterolateral to the brainstem, causing mass effect on the ventral medulla oblongata. Brain MRI demonstrated the lesion to be hyper intense on T1, hypo intense on T2 and FLAIR. There was no diffusion restriction, susceptibility or demonstrable enhancement. The initial diagnosis of hemorrhage based only on non-contrast head CT was quickly eliminated based on MRI. Although only 10% of the neurenteric cysts demonstrate low T2/FLAIR imaging signal and most of them occur in the spine rather than in the brain (3:1), given the location and imaging characteristics, differential diagnosis of neurenteric cyst was entertained and favored over some more common etiologies. A careful attention to imaging features and a location-based approach is required to nail the precise diagnosis.

Introduction
Neurenteric cysts are rare congenital benign lesions of the central nervous system. These are essentially congenital endodermal cysts. Neurenteric cysts are seen more frequently in the spine than the brain (3:1), in particular involving the lower cervical/upper thoracic spine. Neurenteric cysts represent 0.3-1.3% of all intra spinal cystic lesions and are extremely rare intra cranially [1]. In the brain, these are more frequently seen in the infratentorial compartment rather than the supratentorial compartment (70-75%), most commonly in the preponine region. More than 95% of these are extra-axial in location and are usually seen anterior/lateral to the pontomedullary junction, with most of them extending to the midline. While infratentorial lesions are usually less than 2cm, supratentorial lesions can often be quite large [2]. There have been a few reports of these cysts occurring in the medulla or cerebellum [3]. Since these lesions are relatively uncommon and can have variable imaging appearance, they may present as a radiological challenge and may be mistaken for other more ominous diagnoses. Attention to imaging features and location are both essential in formulating the correct diagnosis. Although these lesions can have variable T1 and T2 appearance depending on the intracystic mucinous content, a prepontine anterior/anterolateral lesion, without demonstrable contrast enhancement or diffusion restriction, should steer one towards favoring the diagnosis of a neurenteric cyst. Advanced imaging modalities such as magnetic resonance spectroscopy can serve as an additional diagnostic tool when available, as the neurenteric cysts have been shown to have N-Acetyl Aspartate (NAA)-like peak at 2.02ppm, not seen with other cystic lesions. We illustrate these teaching points via our clinical case report [4].

Case Report
A young 36-year-old Hispanic female G9 P9 L9 presented to the emergency department with dizziness, left sided weakness and blurry vision. She progressed to near syncope and mild left sided ataxia. Physical examination revealed mild left sided motor weakness (muscle power grade 3/5). Initial non-contrast head CT demonstrated an infratentorial, hyper dense, oblong extra-axial lesion ventral to the brainstem, causing mild mass effect. There was no hydrocephalus, midline shift or evidence of suspicious
osseous abnormality. MRI of the brain was performed utilizing 1.5 T scanner and demonstrated this lesion to be hyperintense on T1 and hypointense on T2 and FLAIR series, without abnormal diffusion restriction. There was no demonstrable enhancement on post-contrast T1 series. Susceptibility weighted series failed to demonstrate blooming or evidence of acute or chronic blood breakdown products. Consequently, intracranial hemorrhage and epidermoid cysts were excluded as differentials. In the absence of abnormal enhancement or abnormal calvarial marrow signal, alternative differentials including cystic or metastatic neoplastic processes were also excluded. The patient was managed conservatively since she declined surgical intervention. She showed complete resolution of her left sided weakness on two subsequent six monthly examinations with stability on follow-up brain MRI’s.

**Discussion**

Neurenteric cysts, also known as enterogenous cyst, enteric cyst, gastro enterogenous cyst, endodermal cyst, gastro cytoma, intestinoma and archenteric cyst, are rare congenital endodermal cysts of the central nervous system (like Rathke’s and colloid cysts). Neurenteric cysts are 3 times more common in the spine than the brain. These are extremely rare intracranially, with less than 60 total reported in literature to date [2]. Approximately 70-75% of these cysts are found in the posterior fossa, with more than 95% of these being extra-axial. More than 70% of these extend to the midline. In the infratentorial compartment, the most common location is anterior/lateral to the pontomedullary junction; while supratentorially they are usually seen in the suprasellar and quadrigeminal cisternal region. Most infratentorial cysts are less than 2cm in size, while supratentorially these cysts can often be quite large. Most neurenteric cysts are seen in the spine particularly at lower cervical and upper thoracic spine, and are usually extramedullary intradural in location, ventral to the spinal cord. More than 50% of the spinal neuroepithelial cysts are associated with vertebral anomalies.

Neurenteric cysts were first described by Puuseep in 1934 in a 27-year-old woman with quadriplegia. They can present at any age from the newborn to the fifth decade of life [3]. Review of prior literature, suggests unclear gender preference, with some articles favoring male predominance, while others favoring vice versa [2]. Hence the actual occurrence may be independent of gender.

The exact etiology of neurenteric cyst is controversial. The popularly presumed theory suggests that these cysts arise from a persistent neurenteric canal, due to a residual connection between the embryonic foregut and the developing notochordal plate. However, this theory fails to explain approximately 25-30% of the supratentorial neurenteric cysts, since any lesion cranial to the clivus would not participate in the notochordal development [2]. An alternative theory of Seesel pouch suggests that these cysts probably arise from the endodermal diverticulum found behind the oropharyngeal membrane. However, this only accounts for the midline supratentorial cysts and fails to explain the off midline supratentorial cysts.

Neurenteric cysts have a broad spectrum of imaging presentations. A systematic and simplified imaging and location-based approach can direct one towards the right diagnosis. Intracranial neurenteric cysts are most commonly seen infratentorially and usually present in front of the brainstem in the preoptic cistern region. They tend to be well-demarcated lobulated cystic lesions. Neurenteric cysts can have a variable appearance on non-contrast CT ranging from hypo to iso to hyperdense appearance. They may rarely demonstrate calcifications and maybe associated with bony anomalies. 90% of these are hyperintense to CSF on T2-weighted series, with only 10% being hypointense, which may be attributed to the protein concentration within the cyst [5]. On proton-density and FLAIR series, these lesions are usually hyperintense to CSF, demonstrate no blooming on susceptibility series or abnormal restriction on diffusion series. They usually do not enhance. Minimal rim enhancement has been described and may be secondary to reactive changes [2]. The literature review demonstrates a role for MR spectroscopy in noninvasively confirming this diagnosis at this critical inaccessible skull base location. MR spectroscopy describes the classic finding of a large NAA peak at 2.02ppm, despite the absence of neuronal elements, which may be attributed to the presence of a NAA-like compound in the cyst contents, likely secreted by the goblet/ciliated columnar cells in the cyst wall. Although MR spectroscopy appears to have a role in narrowing the differential diagnosis, it is not always readily available, limiting its utility in everyday practice.

Just like imaging, the pathological appearance of neurenteric cysts on light microscopy can be inconsistent. However, two main histological patterns are found in the cyst walls- one simple columnar non-ciliated epithelial cells (rich in mucin producing cells) and second pseudo-stratified dilated columnar epithelium (poor in mucin-producing cells) [2]. There is a lack of significant correlation between the imaging characteristics and mucin content of the cyst wall as well as between the cyst attenuation on non-contrast CT and cyst pathology. No intracystic hemorrhage is demonstrated pathologically.

Interestingly, T2 hypointensity on brain MRI has been shown to be consistent with elevated protein concentration [5] and has been found to demonstrate squamous metaplasia and voluminous keratinous debris [2]. Although malignant transformation is reported it is extremely rare [6].

The differential diagnosis for neurenteric cysts includes other cystic lesions such as an arachnoid cyst, dermoid cyst, epidermoid cyst, other Endodermal cysts (Rathke and colloid cyst) as well as cystic neoplasms such as schwannomas. While epidermoid cysts typically demonstrate diffusion restriction, the characteristic feature of dermoid cyst is T1 hyperintensity reflective of the underlying lipid components. Endodermal cysts like Rathke’s and colloid cyst are predominantly seen in the
supratentorial region and can be differentiated from neurenteric cysts based on location [2]. Lack of enhancement or minimal rim enhancement differentiates these lesions from alternative cystic neoplasms and nerve sheath tumors such as schwannomas, which demonstrate avid enhancement.

Neurenteric cysts are extremely rare with only 75 reported cases to date. The prognosis is usually stable versus extremely slow growth. Therefore, management is controversial between conservative and surgical management, depending on the clinical presentation. Alternatively, in the presence of incapacitating neurological presentation, surgical resection may be considered [1]. Many different surgical approaches both anteriorly and posteriorly have been described in the literature, including the more recent Far Lateral Transcondylar (FLT) approach. Neurenteric cysts may often time be closely adherent to critical structures such as the brainstem and the spinal cord which may make complete surgical resection challenging.

Intracranial neurenteric cysts have a wide array of imaging appearances and can be mistaken for other more sinister pathologies. We suggest a comprehensive approach including both imaging characteristics and location. Although these are exceedingly rare congenital intracranial lesions, they should be considered in the differential diagnosis of any sharply marginated, extra-axial lesions both supra and infratentorially, regardless of imaging characteristics on non-enhanced CT, T1, and T2 series, since the imaging spectrum is broader than previously described and can vary with the cyst protein content.

Learning Points

a. Just like all that Glitters is not Gold; all that is extra-axial and Hyper dense is not Hemorrhage! Imaging features and attention to location clinch the diagnosis. A well-defined lobulated extra-axial hyper dense lesion anterior to the brainstem is characteristic of an incidental neurenteric cyst.

b. Depending on the protein content in the cyst, these can have variable appearance on CT and T1/T2 MRI and can also demonstrate mild cyst wall enhancement. Look for bony abnormalities.

c. Malignant transformation is extremely rare.

d. Conservative treatment is the preferred treatment. Complete surgical resection may be an option when clinically indicated, but is not always possible or complete due to proximity to critical structures.

References


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