INTRODUCTION

Neurenteric Cysts (NECs), also known as enterogenous cysts, are congenital cysts of the CNS that are believed to be of endodermal origin with the majority reported to occur in spinal canal.\(^1\) They are extremely rare in supratentorial location and giant cysts are even rarer. Less than 17 cases of giant supratentorial neuroenteric cysts have been reported in world literature so far.\(^2\)

Enterogenous cysts have been rarely reported in the mediastinum and thoraco-abdominal region and associated with axial skeletal anomalies, and malrotation.\(^3,4\) Supratentorial NECs may arise from remnants of Seesel's pouch, a transient endodermally derived diverticulum of the cranial end of the embryonic foregut.\(^5\)

Neurenteric cysts that lie ventral to the brainstem are rare and should be included in the differential for intracranial extra axial cystic lesions.\(^6\) Recently, there is an evidence regarding the malignant transformation of these benign lesions into an invasive mucinous papillary cystadenocarcinoma.\(^7\)

CASE REPORT

We report a 35 years old right handed man who presented with one-year history of progressive left hemiparesis, headache, personality changes and seizures which had progressively increased in frequency and were simple partial in character. Clinical examination revealed a confused patient with left hemiparesis (power 4/5 in both left upper and lower limbs); up going plantar reflex and exaggerated deep tendon reflexes at 3+, left facial weakness of upper motor neuron type. CT scan head with contrast revealed a non-enhancing spherical cystic lesion in the frontotemporoparietal region with minimal to moderate mass effect. The cyst was removed using a combination of hydrodissection and excision. Recovery was complete with no evidence of recurrence or residual disease at 3 months.

Key Words: Neurenteric cyst. Seizures. Supratentorial. Extra axial.

ABSTRACT

Intracranial neurenteric cysts are rare congenital lesions that may be mistaken for other cystic neoplastic and non-neoplastic lesions. A 35 years old right handed man presented with a one year history of progressive left hemiparesis, headache, personality changes and seizures. Clinical examination revealed a confused patient with left hemiparesis (power 4/5 in both left upper and lower limbs), upper motor neuron type signs in left half of the body (up going plantar reflex and exaggerated deep tendon reflexes at 3+), left facial weakness of upper motor neuron type. CT scan head with contrast revealed a non-enhancing spherical cystic lesion in the frontotemporoparietal region with minimal to moderate mass effect. The cyst was removed using a combination of hydrodissection and excision. Recovery was complete with no evidence of recurrence or residual disease at 3 months.

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for Glial Fibrillary Acidic Protein (GFAP) immunohistochemical stains. No evidence of malignancy was noted. His postoperative CT scan at 3 months (Figure 1D) shows no evidence of recurrence or residual lesion.

**DISCUSSION**

Neurenteric Cysts (NECs) are manifestations of a rare congenital dysplasia probably resulting from a separation of the notochord and the upper gastrointestinal tract. They are most likely of endodermal origin and lined by ciliated + non-ciliated, low cuboidal + columnar and contains a few mucin secreting goblet cells. The cysts mostly contain a clear fluid of varying viscosity and protein content. NECs represent 0.01% of CNS tumors. They frequently arise in an intradural-extra axial location anterior to the cervical-thoracic spinal cord while intracranial cysts have a tendency to be infratentorial. Although they are decidely known to be benign, there have been four reported cases of malignant transformation in cysts which were intracranial which emphasizes the need for gross total resection of the cyst as well as the wall.7

Giant supratentorial neuroenteric cysts are rare. Less than 17 cases have been reported in the world literature, a majority of which have been reported in the anterior fossa or in deep midline structures.8 There is no set definition as to what size constitutes a ‘giant’ cyst, however, the authors believe that dimensions larger than 5 x 5 cm should be considered to be in the ‘giant’ territory. Supratentorial NECs located far from the location of primitive endoderm suggest the possibility that NECs could arise from ectopic endodermal tissue through an undetermined process of anomalous differentiation.2 These cysts also have the propensity to undergo repetitive spontaneous intracystic hemorrhage which makes it liable to be confused with a brain abscess, aspiration of which may yield a brown viscous pus like material.9

The presentation in this case was headache, seizures, personality changes and hemiparesis as variously described in other case reports.10 The imaging modality of choice is MRI but the authors relied on CT findings as MRI would not have changed the treatment strategy. There was no calcification of the cyst wall but it may be present in other cases. Supratentorial NECs are rare in adult patients and giant cysts are even rarer. Total removal seems to be curative, and is best treatment for symptomatic cases, however, radical resection which may potentially affect brainstem function is not currently recommended. Long-term follow-up with MRI/CT scan is necessary to monitor for recurrence of any residual lesion.6,8

Despite the low incidence of these lesions, it is crucial to be familiar with the presentation, radiographic and histopathologic characteristics and treatment options for NECs especially due to the remote but real possibility of malignant transformation.

**REFERENCES**