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CASE STUDY

NEURENTERIC CYST OF CRANIOVERTEBRAL JUNCTION MANIFESTING DURING THIRD TRIMESTER OF PREGNANCY

***Dr. Venkidesh, K.**

Department of Neurosurgery, Government Medical College, Thrissur-Kerala, India

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ABSTRACT

Neurenteric cysts of the spine usually occur at the lower cervical and upper thoracic spine. Neurenteric cyst of the Craniovertebral junction is extremely rare. They are believed to arise from persistent abnormal communication between endodermal and neuroectodermal tissues in the embryonic phase. On T2-weighted MR images, appear as hyperintense cystic lesions without edema is generally observed. An immunohistochemical study is the key to diagnosis and demonstrates the endodermic origin of the lesion. The neurenteric cyst shows epithelial membrane antigen, cytokeratin, and carcinoembryonic antigen. Here we present a rare case of Neurenteric cyst of the Craniovertebral junction diagnosed during third trimester of pregnancy, its management, and outcome with review of literature.

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INTRODUCTION

Twenty five year old lady presented to the obstetrics and Gynecology department at 36weeks of gestation with spastic quadriparesis worsening over last two months. There was wasting of the upper limbs with severe grip weakness. MRI revealed a T2 hyperintense and T1 hypointense intradural extramedullary lesion without contrast enhancement extending from pontomedullary junction to lower boarder of C7 ventral to spinal cord (Fig. 1a and Fig. 1b). There were no associated vertebral anomalies. She underwent elective caesarian section at 37weeks of gestation. The power of the upper and lower limbs improved marginally after the delivery. One week after the delivery she was taken up for definitive surgery for the cervical lesion. She underwent Sub-occipital approach combined with laminectomy. On opening the dura, a thick walled cyst was seen ventral to the cord, severely compressing and displacing the cord posteriorly (Fig 2a, 2b and 2c). On opening the cyst fluid having cholesterol crystals were seen. After evacuating the contents, cyst wall excised completely. Post-operately patient's power significantly improved and she is on constant follow-up. Histopatholgical findings were consistent with neurenteric cyst.

****Corresponding author: Dr. Venkidesh, K.***
Department of Neurosurgery, Government Medical College, Thrissur-Kerala, India.

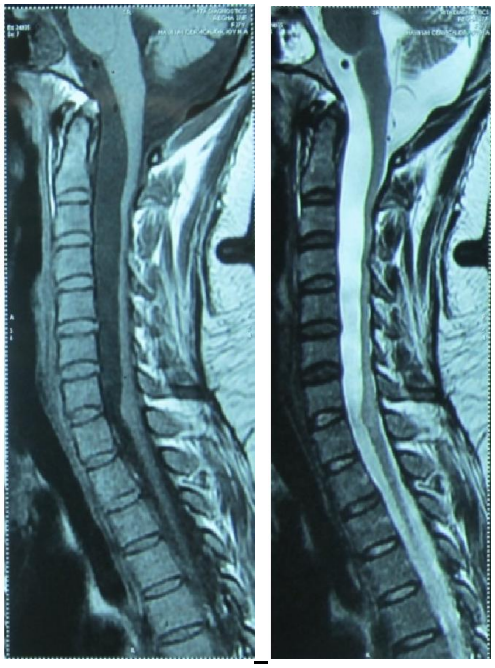

Fig. 1a. MRI T1 and T2W sagittal images showing the extent of lesion from ponto-medullary junction to C7

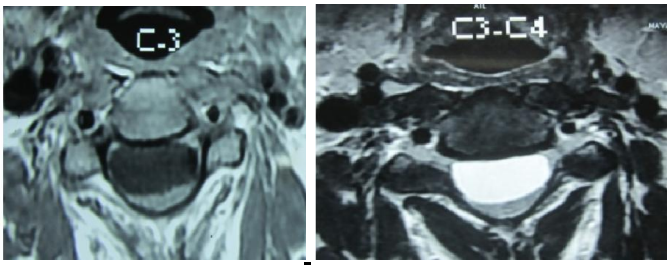

Fig. 1b. MRI T1 and T2W axial images revealing the cyst ventral to the cord and severely compressing and displacing the cord posterior

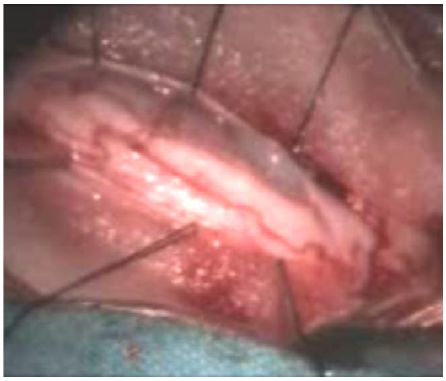

Fig. 2a. Intra-operative image on opening the dura, the cyst seen ventral to the cord displacing it posteriorly

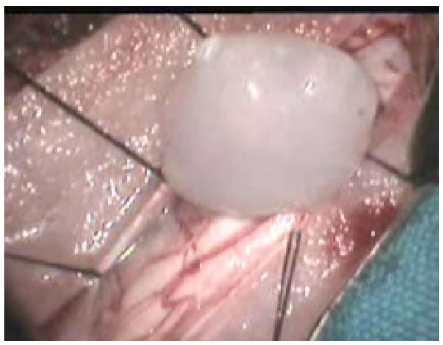

Fig. 2b. Intra-operative image revealing the un-ruptured cyst delivered posteriorly by gentle traction.

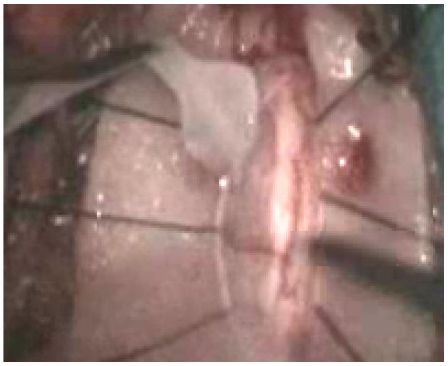

Fig. 2c. Collapsed cyst wall with spilled out cholesterol crystals seen adjacent

DISCUSSION

Neurenteric cysts of spine are considered rare, constituting 07.-1.3% of spinal axial tumors with much lower incidences of location in the intracranial site or craniocervical junction [1,17-28]. On the basis of the literature review, only 23 cases with the craniocervical neurenteric cysts treated by surgery have been reported in the literature [2, 44, 45, 46, 47]. The Lesion was first described by Kubie and Fulton in 1928 as teratomatous cysts and later by Puusepp in 1934 as instestinomas [6, 7]. The term Neurenteric cyst was coined by Holcomb and Matson in 1954[8]. The term enterogenous cyst was first used by Harriman to describe cysts previously known as neurenteric, endodermal or respiratory cyts [14]. It is believed to arise from persistent abnormal communication between endodermal and neuroectodermal tissues in the embryonic phase associated with vertebral anomalies [19, 25, 26]. They can arise anywhere from brainstem to conus with predilection for lower cervical, upper thoracic, cervicothoracic junction and thoracolumbar regions. In rare cases it can be found anterior to CCJ [41, 42, 43].

Most CVJ cysts are located ventrally in the midline anterior to brainstem [9]. About half of the cases are associated with congenital anomalies such as spina bifida or fused vertebrae, hemivertebrae or an anterior spina bifida [15, 16]. They are usually intradural extra-axial lesions. There was only one case of intramedullary location described [53]. Though Wilkins and Odom has sub classified into three types based on histopathology, there appears to be no association between Wilkins and Odom subtypes, and the site, extent, or outcome after the resection of the neurenteric cyst[10]. These cysts are seen in all ages, with the average age around the third and fourth decade, with a male predominance [33]. The most frequent initial symptom is neck pain or occipital headache sometimes accompanied by neck stiffness. Motor weakness and ataxia are the most frequent neurological signs. Rarely there can be lower cranial nerve paresis. Aseptic meningitis resulting from cyst rupture in the subarachnoid space is reported in two cases [9].

In 50% of cases there are associated bony abnormalities like spinal dysraphism, scoliosis, spina bifida, split cord malformation and Kippel-Fiel syndrome [29]. In addition there can be associated malformations of gastrointestinal tract, renal defects, cardiac abnormalities and overlying cutaneous changes [19]. The most common MRI findings associated with neurenteric cysts are non-contrast-enhancing lesions that are isointense on T1weighted sequences and hyperintense on T2weighed imaging. These lesions display characteristic histopathology including well differentiated columnar or cuboidal epithelium with or without cilia and mucous globules. Immunohistochemical studies show positivity for Epithelial Membrane Antigen, Cytokeratin, Carcino Embryonic Antigen, and CA19-9, while they are uniformly negative for Glial Fibrillary Acid Protein, Vimentin and S100 markers. These help in differentiating them from arachnoid cysts, ependymal cysts and cysts of neuroectodermal origin.

Management

Surgical resection is the first line treatment with the goal of total resection. However, strong adhesion of the cyst wall to the surrounding neurovascular structures (brainstem) may sometimes necessitate only subtotal removal of the cyst capsule. In these special cases, a more conservative approach should be taken. After sub-total excision or cyst fenestration,

the residual cyst wall is said to have proliferative potential and can rarely undergo malignant transformation [48, 49]. Signs of aseptic meningitis or increasing levels of CA19-9 in the CSF are possible indicators of recurrence [12, 13] necessitating extended serial follow-up imaging [12] for at least 10 years following surgery [50] and reoperation should be considered in cases of recurrence. An unusual case of holospinal dissemination of multiple neurenteric cysts occurring after fenestration of a fourth ventricle's neurenteric is also described [51]. The sub-occipital approach, sometimes associated with laminectomy, is the classical option to approach this craniocervical junction lesion [52]. A similar approach was used for our patient and carried out total excision of the cyst.

A transoral approach has also been described for cysts located in the midline and attached to the anterior surface of the brainstem. A limited operative field and the risk of postoperative infection limit its use [32].

Conclusion

Neurenteric cyst of the craniocervical junction is rare lesions. MRI is the gold standard for characterizing neurenteric cysts; however CT plays an important role in defining bony abnormalities that are often co-existent with neurenteric cysts. A complete surgical resection is the goal of the treatment. However a more conservative approach should be taken where there is severe adhesion to the surrounding vital structures. Long term follow-ups had revealed excellent outcome following surgical resection with nominal morbidity and mortality. With partial resection the incidence of recurrence is very high and has to be kept on long term follow-up with serial imaging.

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