

Intraspinal Neurenteric Cyst — a rare entity

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Abstract

Neurenteric cysts are uncommonly reported congenital abnormalities that are thought to be an abnormal connection between the endoderm and ectoderm during the 3rd week of life. We report a case of a 17 years old male who presented with one year history of pain in the right arm for which he had been taking pain killer medication from the local general practitioner. The pain progressed over several months and developed into numbness and weakness of the right arm. Magnetic resonance imaging revealed a cystic mass at the C7 to D2 location. The mass was removed surgically and the specimen sent to Armed Forces Institute of Pathology for histopathological examination. On the basis of morphological features, an endodermal mass was diagnosed as Intraspinal Neurenteric cyst.

Keywords: Enterogenous cyst, Neurenteric cyst, Spinal cord.

Introduction

Neurenteric cysts (NC) also called endodermal cysts,

enterogenous cyst, enteric cyst, gastrocytoma, dorsal enteric fistula, split notochord syndrome and teratoid cyst.¹ They are rare congenital cysts of the spine lined by endoderm derived epithelium. They may result from failure of separation of ectoderm from the endoderm during the third week of embryonic life with persistent canal of Kovalevsky.² NC are not only seen in the spinal cord but they may also be found in the abdomen, mediastinum, pelvis, brain and rarely subcutaneously. Persistence of the entire primitive Neurenteric canal results in vertebral anomalies, whereas persistence of only part of the canal results in isolated intraspinal cyst. Isolated intraspinal NC mainly occurs in the cervical region. The presentation may vary from a simple isolated intraspinal cyst with no other abnormalities to that of multiple visceral and vertebral anomalies. We report such a case of isolated Intraspinal NC with no visceral abnormality.

Case Report

A 17 years old male presented with one year history of pain in the right arm along with numbness and severe



Figure-1: MRI of the spinal cord showing ventral intraspinal cyst.

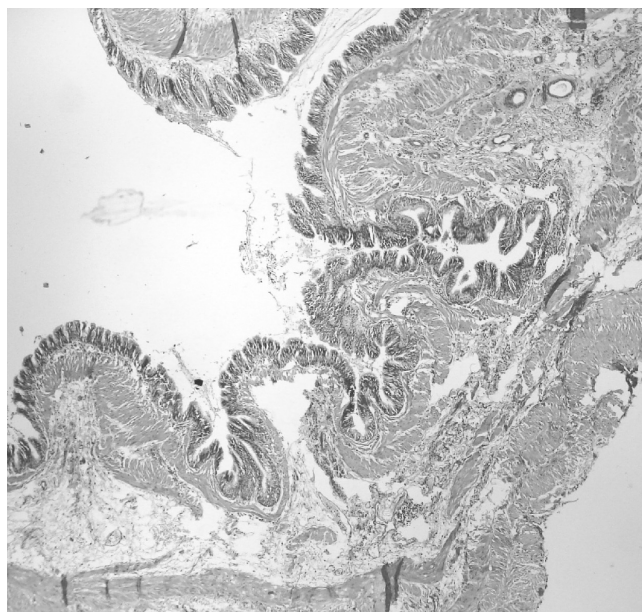


Figure-2: Medium power photomicrograph (original magnification, 200; H-E stain) of the cyst wall showing columnar lining epithelium thrown into villous structures and containing Seromucinous glands.

tingling sensation in the right hand. Patients Magnetic Resonance Imaging (MRI) of the spine revealed a cystic mass measuring at the ventral spine from C7 to D2 (Figure-1). The mass was intradural and extraparenchymal having a thick walled cyst. It was also associated with congenitally abnormal vertebral column at the same level. A radiological suspicion of a dermoid cyst was made. The cystic mass was surgically removed in its entirety and the specimen was sent to Armed Forces Institute of Pathology (AFIP) for histopathological examination.

On gross examination, specimen consisted of a cystic piece of tissue measuring 3 x 2 x 2 cm. On cut section, it was a unilocular cyst. The specimen was processed routinely and stained with haematoxylin and eosin (H&E). Microscopic examination revealed a cyst lined by columnar epithelium of intestinal type (Figure-2). The wall of the cyst contained fibro muscular tissue and thrown into villous structures. Seromucinous glands were also seen. Thus, a diagnosis of Intraspinal NC was rendered.

Discussion

The term intraspinal enterogenous cyst was used for the first time by Harriman³ to describe cysts previously known as endodermal or respiratory cysts. NC typically present with spinal cord or cranial nerve compression. Our patient presented with signs and symptoms of nerve compression with pain and numbness of the hands. The age of presentation varies from 5 weeks to 52 years. More than 160 cases of intraspinal NC have been reported in the international literature. In Pakistan, one case has been reported by Saleem et al in 1999.⁴

Various theories have been proposed to explain the embryological basis of these cysts. The main assumption postulates that the cyst forms as a result of primary adhesion of endoderm anterior to the notochord, incomplete fusion of the notochord, persistence of the neurenteric canal or formation of a split notochord and displacement of endodermal cells.⁵ Hemivertebrae, unsegmentation of vertebra, spina bifida, and clefts in the vertebral bodies are all vertebral abnormalities that can be associated with neurenteric cysts. In our case, there was associated structural anomaly at the level of the cyst.

Most NC are located in intradural and extramedullary region. They may be dorsal or ventral to the spinal canal.⁶ In our case, the cyst was located ventrally. According to the literature, the most common location is the cervicodorsal region as was in our case. Plain radiographs may reveal widening of the spinal canal at the site of the cyst along with associated skeletal anomalies and scoliosis. MRI appears to be the best modality for identifying the complex anatomy of spinal cysts at present. In our case, the radiological appearance was of an intradural, extra-parenchymal thick

walled cyst containing mucinous liquid contents and a suspicion of dermoid cyst was made on MRI.

Wilkins and Odum first described the histological variants of neurenteric cysts. According to their classification, there are three groups of NC.⁷ In group I, cysts lined by single pseudostratified, cuboidal or columnar epithelium with or without cilia are included. Group II includes more complex elements such as mucous glands, serous glands, smooth muscle, fat, cartilage, bone, elastic fibers, lymphoid tissue and nerve tissue (Figure-1). Group III have ependymal or glial elements in addition to the elements seen in group II. Most NC belongs to group II, as was in our case. In our case complete intestinal lining with muscular layer was seen. In other studies published rarely such complete lining epithelium has been observed, usually it is scanty lining epithelium. The wall of these cysts takes up immuno-histochemical stains such as cytokeratin, epithelial membrane antigen, and carcinoembryonic antigen while it is negative for S100 and glial fibrillary acidic protein which confirms their endodermal origin.⁸ The final diagnosis depends on the histological assessment of the cysts.

The choice of surgical technique varies according to the type of cyst and its exact anatomical location. However, surgical exploration aims at preventing spinal cord compression and cyst

refilling after surgery. Our patient recovered well after surgery and was followed up monthly up till six months.

Conclusion

Intraspinal NC are benign embryological defects, which are diagnosed histopathologically and require simple excision in most of the cases.

References

1. Paleologos TS, Thom M, Thomas DG: Spinal neurenteric cysts without associated malformations. Are they the same as those presenting in spinal dysraphism?. *Br J Neurosurg* 2000; 14: 185-94.
2. Prasad VS, Reddy DR, Murty JM. Cervico-thoracic neurenteric cyst: clinicoradiological correlation with embryogenesis. *Childs Nerv Syst* 1996; 12: 48-51.
3. Miyagi K, Mukawa J, Mekaru S, Ishikawa Y, Kinjo T, Nakasone S. Enterogenous cyst in the cervical spinal canal. Case report. *J Neurosurg* 1988; 68: 292-6.
4. Saleem M, Sadiq J, Rauf A, Shaukat M. Intra-Spinal Enteric Cyst. *Pak J Paed Surg* 1999; 1-2: 40-8.
5. Macdonald RL, Schwartz ML, Lewis AJ. Neurenteric cyst located dorsal to the cervical spine: case report. *Neurosurgery* 1991; 28: 583-7.
6. Sreedhar M, Menon S, Varma G, Ghosal N. Cervico-Thoracic Neurenteric Cyst - A Case Report. *Ind J Radiol Imag* 2006; 16: 99-102.
7. Wilkins RH, Odum GL. Spinal intradural cysts. In: Vinken PJ, Bruyn GW, eds. *Handbook of Clinical Neurology*. Amsterdam: North Holland, 1976; pp 55-102.
8. Kapoor V, Johnson DR, Fukui MB, Rothfus WE, Jho HD. Neuroradiologic-pathologic correlation in a neurenteric cyst of the clivus. *AJNR Am J Neuroradiol* 2002; 23: 476-9.