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RESEARCH ARTICLE

NEURENTERIC CYSTS WITHOUT VERTEBRAL ANOMALIES: AN UNUSUAL PRESENTATION

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ABSTRACT

Neurenteric cysts are congenital lesions of the spinal cord which usually communicate to other endodermally derived structures of the body, the embryological theory of which is yet not properly established. They are usually located in the cervico-dorsal region of the spinal cord with an associated vertebral anomaly like spina bifida, butterfly vertebra, lack of segmentation, partial fusion, scoliosis together with the cyst. However, only few cases have been reported in literature where the cysts have not been associated with any vertebral anomalies.

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INTRODUCTION

Neurenteric cysts are a relatively rare malformation that causes compression of the spinal cord. Several hypotheses concern the embryonic origin of neurenteric cysts, of which the "split notochord" theory is the most accepted one. These cysts have been recognized, classified and assigned various nomenclature since the early 20th century. These cysts were commonly associated with vertebral body defects, diplomyelia, spina bifida, and even imperforate anus and intestinal prolapse. Failure of complete obliteration of the neurenteric canal explains the spectrum of neurenteric cysts encountered clinically.

MATERIALS AND METHODS

All patients presenting to our institute with features suggestive of myelopathy, radiculopathy or focal pain were advised MRI. Patients with spinal SOL were screened and those with features suggestive of a cystic lesion were selected. Out of the 358 cases of spinal SOL that presented to our institute over the last 10 years, 4 cases of enterogenous cyst were diagnosed.

RESULTS

In the present series, 4 cases of neurenteric cysts were studied. Age of the patients ranged from 6.5 years to 19 years, 3 of them being in the second decade. There was equal number of male and female subjects in the study. The earliest duration of presentation was at 6 months in one of the subjects while rest presented at a later phase. In our study, three of the cysts were located in the cervical spine while one cyst was located in the dorsal spine. All the cysts were intradural-extramedullary in their location and all were ventrally located to the spinal cord. No associated vertebral abnormality was detected in any of the cases. All except one of the cysts were isointense on T1 sequence and hyperintense on T2 sequence. One of the cysts showed layering effect on MRI. The majority of subjects presented with neck pain with features of quadriparesis, while the subject with dorsal lesion had back pain and associated with paraparesis. Sphincter involvement was present in only one of the subjects. All the subjects were treated by a surgical decompression by a posterior approach. Complete surgical resection was not possible due to associated subarachnoid adhesions. Hence, partial resection of the cyst was done. Histopathological examination of the cysts revealed that three cysts belonged to the Group II of Wilkins and Odum's classification, while one belonged to Group I. All the patients recovered to their premorbid neurological status following surgery except one in which there was no deterioration. The patients were followed up for a period of 3 years. There was recurrence in one subject at the same site (D5-D6).

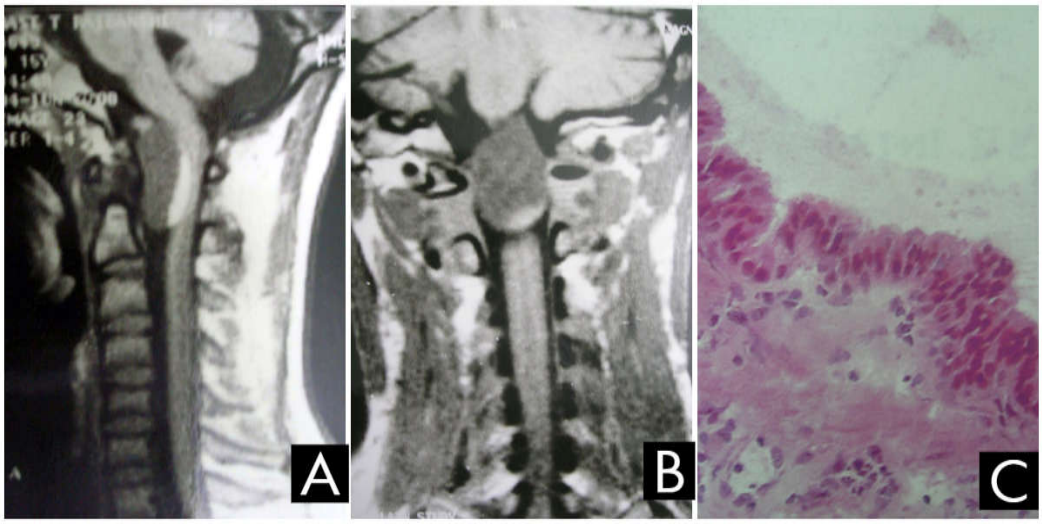

Fig. 1. [A] T1-contrast sagittal and [B] coronal images of neurenteric cyst in front of foramen magnum, C1, C2 vertebra. [C] Histopathology showing Group I pattern of cyst wall lining

DISCUSSION

Neurenteric cyst is a relatively rare cause of spinal cord compression accounting for 0.7 – 1.3% of spinal tumours (Fortuna *et al.,* 1983). These lesions were first described by Kubie and Fulton in 1928 as teratomatous cysts and later by Puusepp (Puusepp, 1934) as intestinomas in 1934. Holcomb and Matson (Holcomb *et al.,* 1954) coined the term neurenteric cyst in 1954. Several theories have been postulated to explain the embryological basis of enterogenous cyst. According to Bently and Smith (1960), these cysts form a part of the spectrum of diseases that occur following herniation of endodermal components through a partially duplicated notochord, labeled as "split notochord syndrome". Cohen and Sledge (1968) further expressed that this single embryological abnormality accounts for the association of cutaneous defects, vertebral anomalies, intestinal duplication, mediastinal and spinal enterogenous cysts, and diastematomyelia. Bently *et al.* (1960), stated that these cysts are invariably associated with vertebral cleft, however multiple cases of intraspinal enterogenous cysts not associated with spinal abnormalities have been reported in literature. In the present series, none of the subjects had any associated spinal abnormalities. Dorsally situated intraspinal cysts which occur less frequently have been postulated to be of ectodermal origin, as the primitive streak ectoderm is probably capable of differentiating into endoderm as well as paraxial mesoderm (Reddy *et al.,* 1968). Lippman *et al.* (2001), in their review stated that 90% of neurenteric cysts of spine are located in intradural/ extramedullary compartment, while the remaining 10% are distributed in intramedullary and extradural locations. They may be located dorsal or ventral to the spinal cord, but ventrally located cysts are more common. According to literature, cervico-dorsal location is most common for neurenteric cysts (Khadim *et al.,* 2011). In our series all the cysts were located in the intradural/ extramedullary compartment, ventral to the spinal cord in the cervico-dorsal region. Individuals with neurenteric cysts most frequently present in the second or third decade of life. Majority of adult patients present with features of focal pain, fluctuating myelopathy or radiculopathy. Neurenteric cysts are allied with bony abnormalities of the spine in approximately 50% of cases and are associated with a variety of conditions including spinal dysraphism, scoliosis, spina bifida, split cord malformation, and Klippel-Feil syndrome (Brooks *et al.,* 1993). Majority of our subjects were from second decade and presented with focal pain and features of myelopathy. There were no associated bony abnormalities in any of the subjects. Wilkins and Odum (1976) first described the histological variants of neurenteric cysts. According to their classification, there are three groups of neurenteric cysts. In group I, cysts lined by single pseudostratifed, 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. Group III have ependymal or glial elements in addition to the elements seen in group II. Most neurenteric cysts belong to group II. All except one patient in our study had Group II pattern on histopathology. MRI is superior to any other investigation in delineating the cyst and its relation with adjacent structures (Brooks *et al.,* 1993). Most commonly the lesions were non-contrast enhancing that were isointense on T1-weighted sequences and hyperintense on T2-weighted sequences (Brooks *et al.,* 1993). However, Mazumdar *et al,* have described neurenteric cysts with abscess or granuloma like presentation, with peripheral ring enhancement of the lesion (Muzumdar *et al.,* 2008). Kimura et al, in their series of 18 patients, found that in 16 patients, the cysts were hyperintense on T1 sequence. All cysts were also hyperintense to CSF in FLAIR sequence. Paolini and colleagues have reported presence of a false mural nodule within the cystic lesion which was later determined to be a collection of mucinous debris along the inferior aspect of the cystic chamber (Paolini *et al.,* 2003). All these studies suggest that variation in MRI findings is frequent and atypical findings should not exclude the potential diagnosis of neurenteric cysts. In the present study all except one of the cysts were non-contrast enhancing that were isointense on T1 - weighted sequences and hyperintense on T2 -weighted sequences. Surgical resection is the first-line treatment for neurenteric cysts with the goal of total excision, given the possibility of cyst recurrence following partial resection. Three approaches to the spine have been described for cyst removal – namely anterior, lateral and posterior. However, the potential advantage of one approach over the other has not been described in literarture (Savage *et al.,* 2010). As per de Oliveira et al, the posterior approach has been most frequently involved in surgical resection of the cyst, despite the ventral preponderance of the cysts, as this approach was associated

with the least complications (De Oliveira *et al.,* 2005). Posterior resection may occasionally require aspiration of contents which carry a greater risk of membrane rupture and consequent meningitis. However, cyst aspiration, partial resection with marsupialization and cysto-subarachnoid shunting has been reported as alternative treatment modalities when complete resection of the cyst was not possible due to dense subarachnoid adhesions or intra-medullary location of the cysts (Savage *et al.,* 2010). In our study all the lesions were approached through a posterior approach. Complete resection was not possible for the ventral cysts due to dense subarachnoid adhesions. Partial resection and decompression was done in all cases. Kim et al, and Cai et al, have observed no recurrence in their series (Cai *et al.,* 2008). Holmes et al reported a recurrence of 4% in their patients (Holmes *et al.,* 1978). One of the patients in the present study had a recurrence of lesion at the same site at 3 years follow-up.

Conclusion

Neurenteric cyst is a rare lesion, incidence in our series being 1.12%. These lesions may or may not be associated with a vertebral abnormality. Operative outcome is satisfactory even after subtotal removal of the lesion. However, larger studies are suggested for confirmation of our findings.

Conflict of interest: There were no conflicts of interest in this study.

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