CASE REPORT

Upper Cervical Spine Symptomatic Intradural Extramedullary Epithelial Cyst: A Case Report

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SUMMARY

We report a rare case of spinal intradural extramedullary epithelial cyst in 18 year old male. The patient presented with history of neck pain 1 year and weakness of all four limbs 2 months. A preoperative magnetic resonance image shows a well defined intradural extramedullary cystic lesion on the ventral side of cord at C2 to C4 level. The lesion was hyper intense in T2-weighted images and hypointense in T1-weighted images. The patient underwent C2-C4 laminectomy and excision of intradural and extramedullary cystic lesion. After having the cyst wall completely removed and adequate decompression of spinal cord, the patient improved neurologically. A histological study of the surgical specimens revealed that the cyst wall walls are lined by flattened to cuboidal cells with stratification of nuclei, cilia are also present focally. We suggest that the total excision of intradural extramedullary epithelial cysts creates adequate decompression of spinal cord and gives neurological improvement.

INTRODUCTION

The different varieties of developmental intradural spinal cord cysts include enterogenous cysts, teratomatous cysts, arachnoid cysts, neurenteric cysts, foregut cysts, bronchogenic cysts, colloid cysts, ependymal cysts, and epithelial cysts[4,5,10,11]. All of these cysts are infrequent, but spinal intramedullary epitheial cysts are particularly rare. Here, we report an additional case: a 18-year-old male with an intradural extramedullary cyst in the cervical spine C3-C4 level. Additionally, we discuss the diagnosis, operative management, and pathological features.

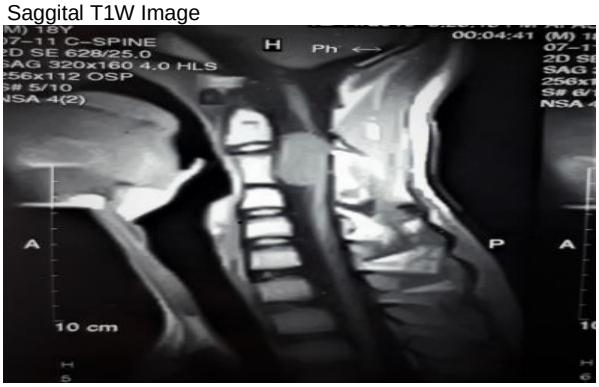
CASE REPORT

A 18 year old male who was shop keeper presented with a history of mild to moderate neck pain for 1 year, gradually progressive weakness of all four limbs for 2 months and complete weakness of all four limbs for 2 weeks however his sphincter control was normal. A neurological examination showed spastic quadriplegia with sensory level of C3. A magnetic resonance image (MRI) revealed a well-defined cystic lesion on the ventral side of the spinal cord at the C3 – C4 levels. The lesion was oval in shape and 36×15 mm in size. The spinal cord was compressed and displaced posteriorly and leftward. The lesion was hyperintense in T2-weighted images and hypointense in T1-weighted images (Fig. 1).

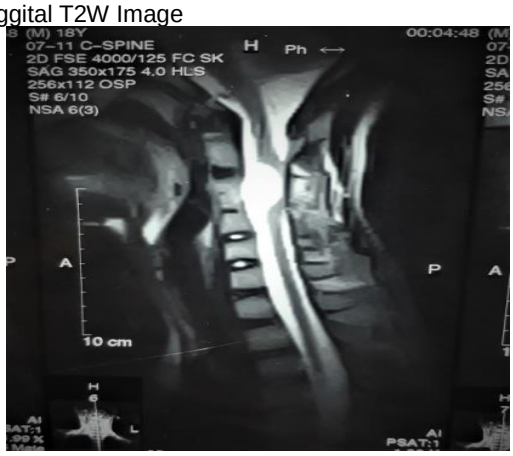
Fig.1: The magnetic resonance image revealing a well-defined cystic lesion on the ventral side of the spinal cord at the C2 to C4 levels. The lesion is a 36×15 mm oval intradural extramedullary cystic mass. The lesion is hyperintense in T2-weighted images (B and D) and hypointense in T1-weighted images (A and C). In the axial imaging, the cyst is observed to compress the spinal cord on the left dorsal side (C and D).

The patient underwent surgery under general anesthesia, prone position, midline incision made after confirmation of level by fluoroscope, C2 – C4 laminectomy done, dura opened in linear pattern, cyst aspirated by fine needle and decompression attained. Cyst wall excised and removed in total.

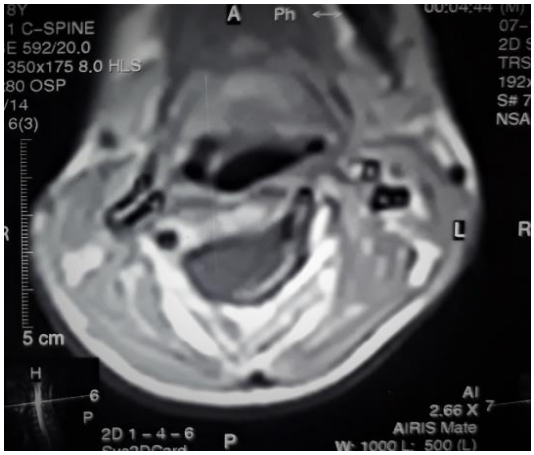
A: Saggital T1W Image



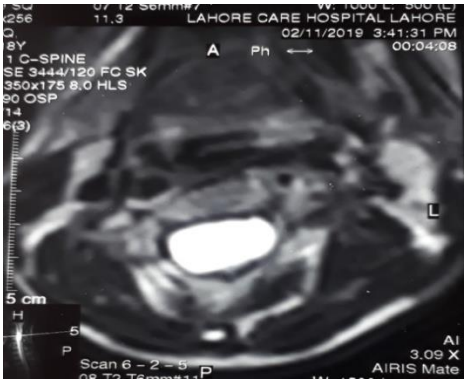
B: Saggital T2W Image



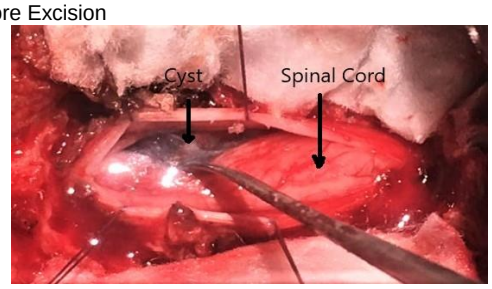
C: Axial T1W Image



D: Axial T2W Image



A) Before Excision



B) After Excision

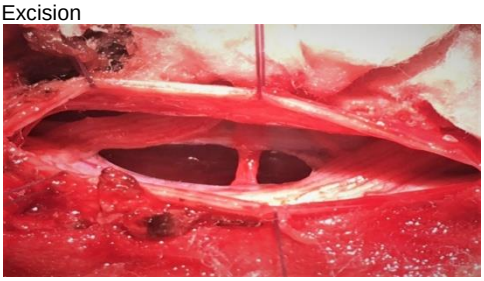


Fig. 2: A glistening white cyst at hemorrhage or parenchyma (A). The cyst wall is excised to obtain adequate decompression and plain for resection (B)

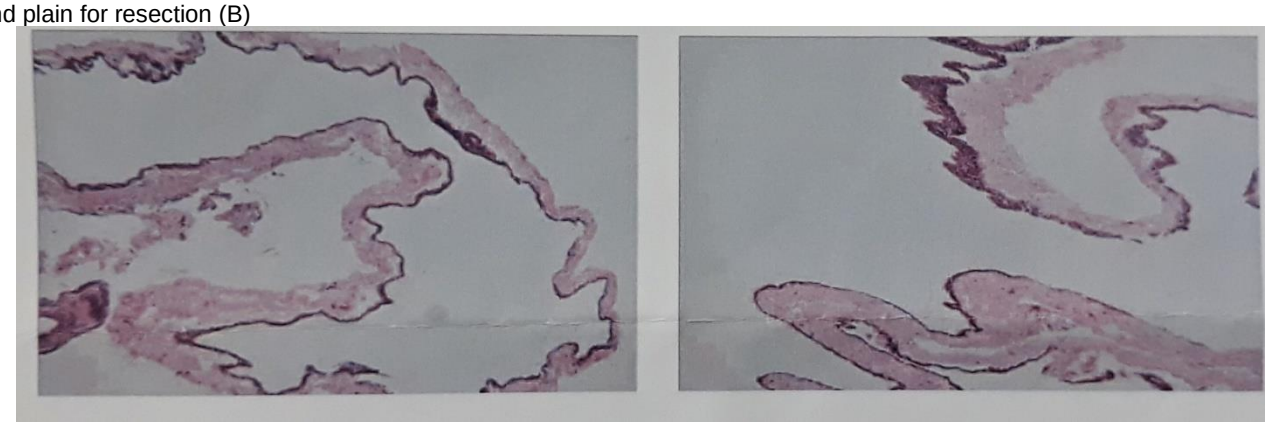
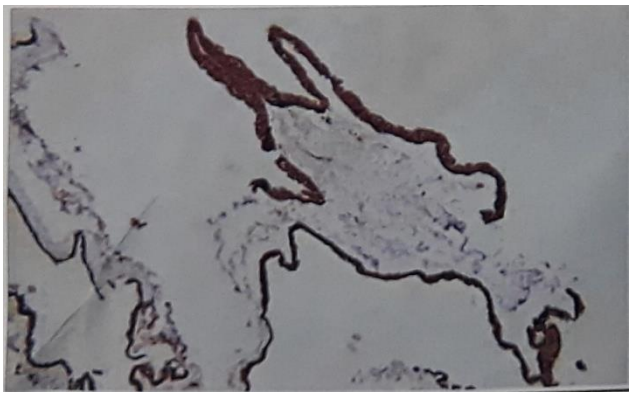


Fig.3: In the H & E staining Epithelial cysts are cystic lesion, walls are lined by flattened to cuboidal cells with stratification of nuclei, cilia are also present focally). These findings are consistent with an epithelial cyst diagnosis.



DISCUSSION

Epithelial cysts are cystic lesion, walls are lined by flattened to cuboidal cells with stratification of nuclei, cilia are also present focally. During spinal cord development, the walls of the recently closed neural tube consist of neuroepithelial cells. These cells extend over the entire thickness of the wall and form a thick pseudostratified epithelium. They are connected at their lumens by junctional complexes. They divide rapidly during the neural groove stage and immediately after tube closure. During this time period, these cells produce an increasing number of epithelial cells. Collectively, they constitute the neuroepithelial layer[9]. Once the neural tube closes, the neuroepithelial cells begin to give rise to another cell type that is characterized by a large round nucleus with a pale nucleoplasm and a dark-staining nucleolus. These cells are the primitive nerve cells or neuroblasts. Neuroepithelial cells following the other pathway differentiate into ependymal cells, which line the ventricular spaces and the central cana[9].

The most widely accepted hypothesis regarding the genesis of ependymal cysts holds that the floor plate of the neural tube is evaginated on the ventral side and becomes isolated to form an ependymal cyst[5,14,16]. The locations of the isolated ependymal cells determine whether the cyst presentation is intramedullary or extramedullary[5]. Additionally, the nature of the cyst presentation may cause the cyst to be present anywhere along the cranio-spinal axis[5]. There have been frequent reports of extramedullary spinal ependymal cysts. However, intradural extramedullary spinal epithelial cysts are rare lesions.

CONCLUSION

We report a rare spinal intradural extramedullary epithelial cyst case. These cysts are benign in nature, and there is usually good functional recovery and a low recurrence rate after complete excision. Hence, we consider total excision is safe and optimal treatment for intradural extramedullary epithelial cysts.

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