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Case Report

Spinal Intra-Dural Bronchogenic Cysts

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Abstract

Background: Bronchogenic cysts are congenital malformations derived from anomalous budding of the embryonic foregut. An intradural extramedullary bronchogenic cyst is a congenital malformation and an extremely rare type of endodermal cyst.

Case Presentation: We present 2 cases with bronchogenic cyst. A 43 years old female and 52 years old male patients who presented with radicular and myelopathy symptoms in the limbs and their images revealed intra dural cystic lesions in the cervical and thoracic regions respectively. Both patients underwent total surgical excisions and histological examination revealed bronchogenic cysts.

Conclusion: Total excision is the treatment of choice with good prognosis. Incomplete resection has a high risk of recurrence. Radiological follow up is recommended.

Keywords: Bronchogenic Cyst; Endodermal Cyst; Intradural Extramedullary; Intraspinal Cyst

Introduction

Spinal Bronchogenic cyst is a rare entity and account for only 0.7 - 1.3% of all spinal cord tumour [1]. This congenital malformation is a rare endodermal cyst lined with respiratory epithelium. Bronchogenic cyst usually originates in the mediastinum but can be found in the sternum, skin, pericardium and diaphragm and rarely spine [2]. Spinal bronchogenic cysts are benign and are a subtype of neurenteric cysts. They arise due to embryological abnormalities of the developing foregut. Therefore, these cysts are also called as foregut cysts or enterogenous cysts [3]. These are usually solitary, but multiple may be found in a patient and can be filled with fluid or proteinaceous material. These have been also reported in more remote locations like the neck, abdomen and retroperitoneal space. Intraspinal bronchogenic cysts are extremely rare and most of them are extramedullary [4]. Compression of the spinal nerve root, anterior spinal artery, and/or the spinal cord by the cyst can cause features such as radiculopathy, myelopathy, or radiculomyelopathy [5,6]. Surgical excision of the SBC is the treatment of choice. Good neurological recovery following surgery has been reported in literature. Histological examination of the cyst is necessary for the confirmation of the diagnosis [7]. A lining of pseudostratified ciliated columnar epithelium resembling respiratory epithelium is the diagnostic hallmark of SBC. Rarely, malignant transformation of these cysts has been described in the mediastinal bronchogenic cysts but not in the spine [6].

Case1

42 years old civil servant presented with paraesthesia in both upper and lower limbs and numbness of fingers on both sides. She also complained of electric shock like sensation in her neck. On examination she had clear signs of myelopathy with mild weakness of her limbs (4/5). Deep tendon reflexes were all exaggerated and Hoffman's sign was positive. Plantar responses were however down going. An MRI scan of cervical spine showed an intra-dural but extramedullary cyst in C1/C2 region causing spinal cord compression. It was hyper intense in T2 weighted image and iso to mild hyper intense in T1 weighted image. It was not enhancing with contrast. She underwent C2 laminectomy and total excision of the cyst.

It was attached to the root on the right side which needed gentle dissection to separate, to ensure total removal. She recovered well from the operation and symptoms improved to a large extent over time. Histopathology confirmed lobulated cyst, lined by pseudostratified columnar ciliated epithelium as seen in respiratory mucosa. No glial element in GFAP stain. Microscopic appearance confirmed bronchogenic cyst. Post-operative scan confirmed total excision. A two year follow up shows that her myelopathic symptoms and signs have resolved, with no recurrence on imaging to date.

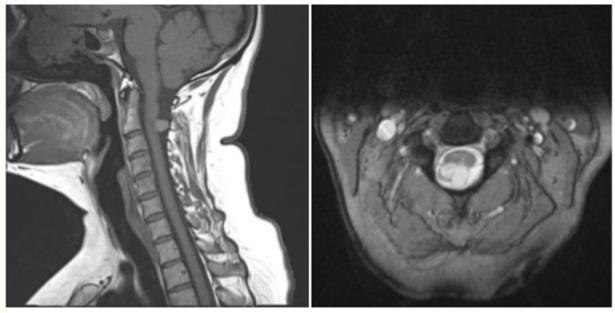

Figure 1: Sagittal and axial magnetic resonance imaging image showing cystic lesion, behind C2 posterior to the cord. Lesion is hypointense on T1-weighted images and hyperintense on T2-weighted images.

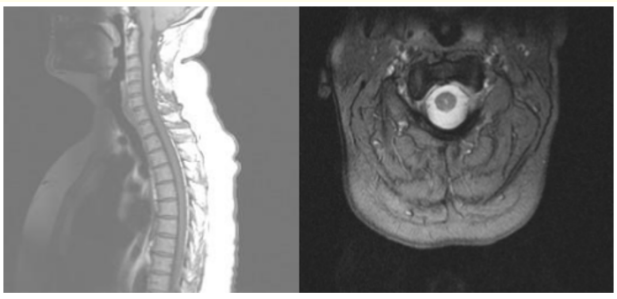

Figure 2: Postoperative images in sagittal and axial sections of T1 and T2-weighted magnetic resonance imaging showing complete excision of cystic lesion behind C2 posterior to the cord with no evidence of residual.

Case 2

50 year old gentleman presented with longstanding LBP and right sided sciatica. On neurological examination he did not have any deficit. His SLR was restricted on the right side. He had an MRI scan of spine which identified a right sided L5/S1 disc prolapse and T10/11 intradural lesion. It was isointense on T1 and hyper intense on T2 weighted images. There was no contrast enhancement. Radiologist suggested a possibility of a proteinaceous cyst in the absence of contrast enhancement. The lesion was approximately 1 cm in size. He underwent T10/11 laminectomy and excision of the cyst, along with a right sided L5/S1 micro discectomy. The cyst was attached to the dorsal aspect of spinal cord. He recovered well from the operation and his right sided sciatica improved and back pain got better. Histopathological study has shown a columnar ciliated epithelium lining of the cyst. Immune staining confirmed it to be an endodermal cyst with strongly positive cytokeratin CK5 stain. A one year follow up shows good clinical outcome with confirmation of total excision on imaging.

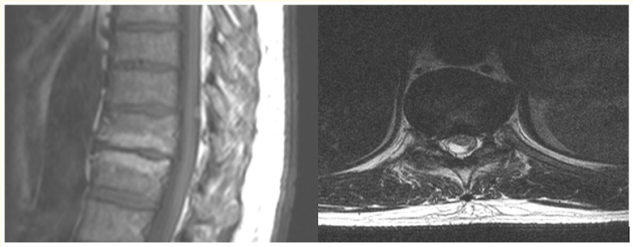

Figure 3: Sagittal and axial magnetic resonance imaging image showing cystic lesion at T10/11 , extending posterior to the cord. Lesion is hypointense on T1-weighted images and hyperintense on T2-weighted images.

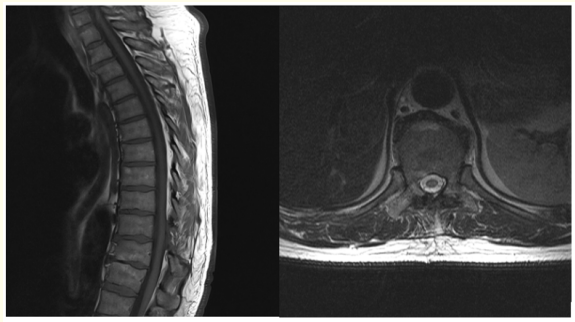

Figure 4: Sagittal and axial magnetic resonance imaging image showing complete excision of the lesion at the previously noted cystic lesion at T10/11 with no evidence of residual clearly in both sagittal T1 and axial T2 sequences

Literature Review and Discussion

With the appearance of magnetic resonance imaging, it became easy to diagnose the spinal cysts. Base on anatomical presentation, these cysts include intramedullary, intradural, extradural cysts, perineural cysts, as well as synovial, and discal cysts. Spinal meningeal cysts are classified as intradural and extradural ones. Intradural cysts include arachnoid cysts, enterogen (endodermal, neuroenteric) cysts, and ependymal cysts. Among the cysts in the spinal canal, the arachnoid derived lesions are most common.

Bronchogenic cysts are also known as neurogenetic cyst or enterogenous cyst or endodermal cyst or foregut cyst. They originate from congenital remnants of primitive foregut. Age of presentation is variable and generally has no gender preference. It can be associated with Spina bifida.

Bronchogenic cysts are found along the tracheobronchial tree in the mediastinum or with the lung parenchyma. Very Rarely, these cysts occur in other locations, as skin, subcutaneous tissues, the pericardium, the diaphragm, the abdomen and the spinal cord [8].

The main explanation for the existence of these malformations might be due to faulty separation between the ectodermal and endodermal layers which results into inclusion of endodermal tissue within the ectodermal layers of the spinal cord. Later, the endodermal layer may differentiate into digestive or respiratory tissues and the bronchogenic cysts may present with both types of histological features [8].

Clinical presentation usually depends on location of the cyst and it's pressure effect on neural structure. Our first case was causing myelo-radiculopathy and the second case caused local pain. MRI scan is the mainstay for diagnosis. Most cases of neurogenetic cyst have shown iso or hypo intensity on T1 and hyper intensity on T2 weighted MR scan. Our first case was iso to hyper intense on T1 and hyper intense on T2 weighted image. They do not enhance with contrast. Our first case was adhered to nerve root of one side and the second case was adhered to the spinal cord. This can be challenging during operation. Surgical excision is the treatment of choice and generally has good outcome. Total excision is the goal. Garg., *et al* (2008) reported no recurrence with total excision and 63% recurrence with partial resection. Tight adhesion may prevent total excision [9].

There has been few cases mentioned in the literature, all were surgically excised. There was no clear pattern of distribution through the spine. Its adherence to the surrounding neural structure makes its total excision a great challenge. Also there was a clear understanding that total excision has a great impact on the neurological recovery avoiding the risk of later recurrence. In these cases, radiological follow up is recommended. On the other hand, most of the cases were intradural and extramedullary despite they may still be intramedullary. The pathological process and behavioural history still not clearly understood.

Conclusion

Total excision is the treatment of choice with good prognosis. Incomplete resection has a high risk of recurrence. Radiological follow up is recommended as the pathological process and behavioural history still not clearly understood.

Consent for publication

Not applicable.

Declarations

Ethics approval and consent to participate

This is a case report so ethical approval is not required.

Informed written consent

Patient gave consent to participate.

Availability of data and material

(Hospital records) on the scanned documents on hospital online records.

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Authors' contributions

Record and document the manuscript.

Acknowledgements

Not applicable.

Trial Registration

Not applicable as it is not a prospective trial it is a case report The authors certify that they have obtained all appropriate forms.

Conflicts of Interest

There are no conflicts of interest.

Abbreviations List

Not applicable.

Bibliography

1. Chen J., *et al*. "Case Report Series and Review of Rare Intradural Extramedullary Neoplasms-Bronchiogenic Cysts". *Medicine (Baltimore)* 94.49 (2015): e2039.

2. Weng JC., *et al*. "Intradural Extramedullary Bronchogenic Cyst: Clinical and Radiologic Characteristics, Surgical Outcomes, and Literature Review". *World Neurosurgery* 109 (2018): e571-e80.

3. Sundaram C., *et al*. "Cysts of the central nervous system: a clinicopathologic study of 145 cases". *Neurology India* 49.3 (2001): 237-242.

4. Baumann CR., *et al*. "Thoracolumbar intradural extramedullary bronchiogenic cyst". *Acta Neurochirurgica (Wien)* 147.3 (2005): 317-319.

5. Zhao H., *et al*. "Effects of critical ultrasonic management of Peking Union Medical College Hospital on the etiological diagnosis of patients with acute respiratory failure]". *Zhonghua Yi Xue Za Zhi* 95.47 (2015): 3843-3847.

6. Fiorelli A., *et al*. "Malignant transformation of bronchogenic cyst revealed by 99mTc-MIBI-SPECT". *Asian Cardiovascular and Thoracic Annals* 20.3 (2012): 347-349.

7. Jha R., *et al*. "Upper cervical bronchogenic cyst: A rare condition at a rare location". *Journal of Neurosciences in Rural Practice* 9.1 (2018): 149-151.

8. Ko KS., *et al*. "Sacral intraspinal bronchogenic cyst: a case report". *Journal of Korean Medical Science* 23.5 (2008): 895-897.

9. Garg N., *et al*. "Is total excision of spinal neurenteric cysts possible?" *British Journal of Neurosurgery* 22.2 (2008): 241-251.