

An Intradural Extramedullary Bronchogenic Cyst in the Lumbar Spine

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Abstract

Background context: Bronchogenic cysts are rare congenital abnormalities of the central nervous system, identified as an epithelial endodermal cyst and can present as intradural extramedullary lesions

Purpose: Intradural extramedullary lumbar bronchogenic cysts present as a seldom cause of cauda equina syndrome. We discuss a case of a 51-year-old male presenting with sciatica and lower limb neurology, with the aforementioned cyst compressing both the conus medullaris and cauda equina.

Study Design/Setting (Patient Sample and Outcome Measures must be used in all Clinical Studies)

Results: Total surgical resection was curative and histology confirmed a bronchogenic cyst. The lesion is a rare congenital malformation thought to derive from remnants of primitive foregut.

Conclusion: Bronchogenic cyst should be considered as a differential in patients presenting with an intradural extramedullary lesion in the lumbar spine. Surgical excision offers cure with good results, and in our patient as exhibited in the post-operative MRI and clinical outcome

Keywords: Bronchogenic cyst; Cauda equina syndrome; Intramedullary cyst; Intraspinous cyst; Lumbar spine

Introduction

A 51-year old male presented initially with a three-month history of numbness in the buttocks radiating down to the right leg in the S1-dermatomal distribution. The numbness was associated with pain mostly at the buttock area. There was no perianal paraesthesia nor was there difficulty voiding his bladder, however the patient did complain of difficulty evacuating his bowels. The patient was otherwise medically fit. On clinical examination the patient had weakness of the right ankle in both dorsi and plantar-flexion, with Medical Research Council (MRC) scale for muscle strength of four in both. Straight leg raise was positive on the right at around 30 degrees.

The ankle jerk reflex was bilaterally absent and right knee reflex was decreased but the left was normal. Sensation was decreased in S1-S4 dermatomes distribution bilaterally and his anal tone was normal, however, voluntary constriction was decreased. The peripheral pulses were found to be normal. The

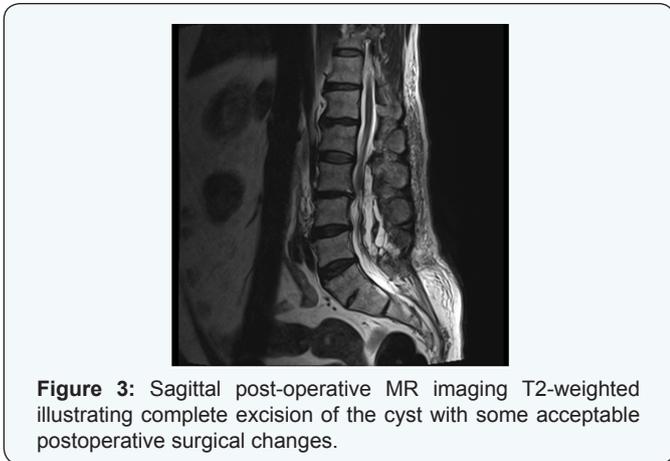
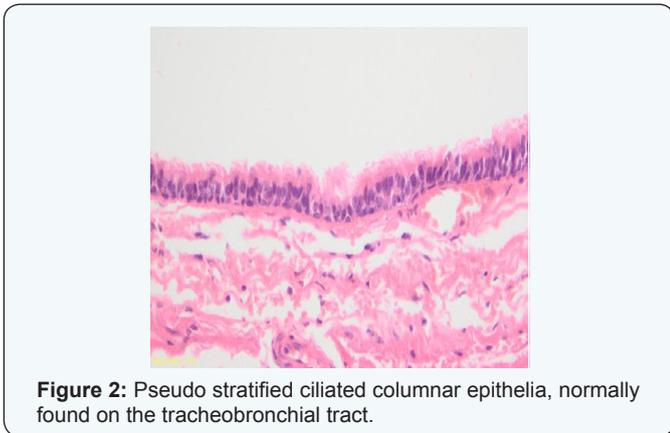


Figure 1: Sagittal pre-operative MR imaging T2-weighted with the lesion at the L3/L4 level.

Magnetic Resonance (MR) image of the patient's spine showed there to be an intradural extramedullary mass lesion centered at the level of L3, below the conus medullaris, and

expanding almost the entire length of the L4 vertebra. It was causing significant compression of the cauda equina nerves and was shown to be expanding the conus medullaris (Figure 1).

The lesion was initially thought to represent a dermoid or epidermoid cyst of the conus, causing tethering of the spinal cord. The patient underwent a midline osteotomy and laminectomy with subsequent durotomy performed, therefore gaining access at the levels of L2-L4 for the extensive surgical excision of this lesion guided by neurophysiology. Cystic fluid and tissue samples were sent for histopathological analysis (Figure 2 & 3). The patient in the immediate post-operative period had residual inability to evacuate his bowels and at follow-up this has been slowly improving.



Discussion

Bronchogenic cysts are rare congenital abnormalities of the central nervous system, identified as an epithelial endodermal cyst. Neuroenteric cysts are a more common type of endodermal cyst than the bronchogenic variety accounting for 0.7-1.3% of all spinal cord tumours [1]. Endodermal cysts are described as a combination of gastrointestinal and/or respiratory type epithelium with the absence of other germinal layers including non-intestinal components, a vertebral anomaly, and a classical cervical or upper thoracic location [2,3]. A lesion is termed a bronchogenic cyst if the endodermal lining is predominated

with respiratory tract epithelium, illustrated in (Figure 2) with pseudo stratified ciliated columnar epithelia that is normally found in the tracheobronchial tract. These cysts are therefore remnants of primitive foregut from which the respiratory system originates and, more commonly located in the mediastinum [2] (Table 1).

Table 1: William and Odom formulated a system to classify intrathecal cysts based upon three histopathological categories and a bronchogenic lesion is a type A cyst [4].

Characteristics	Type A	Type B	Type C
Single layer of pseudostratified columnar or cuboidal cells mimicking respiratory or gastrointestinal epithelium	+	+	+
Complex invaginations with glandular organization; mucinous or serous production; nerve ganglion, lymphoid, skeletal muscle, smooth muscle, fat, cartilage, and/or bone elements	-	+	-
Ependymal or glial tissue	-	-	+

Mesoderm separates the first two embryonic layers endoderm and ectoderm, and grows forward from both sides of the primitive streak. Originating from the cephalic end of the primitive streak, the mesodermal notochord grows forward in the central axis. The growing notochord becomes transiently incorporated into the dorsal endoderm wall that later becomes the foregut. The endoderm extrudes it, moving dorsally between the endodermal tube and ectoderm. Somites form both sides of the notochord rod are segments of the paraxial mesoderm, with the vertebral bodies and inter vertebral discs forming from the sclerotomes fusing around the notochord. The notochord induces the surface ectoderm to form the neuro-ectoderm which forms the neural tube and subsequently the brain and spinal cord. The neural tube closes cranially too caudally, with the former opening (neuropore) closing at day 26 and latter by day 27, with failure in closure resulting in anencephaly and spina bifida, respectively. Vertebral structures, meninges and skin from the remaining ectoderm develop around the closed neural tube [3,5].

The pathogenesis and mechanism for development of Bronchogenic cysts is unknown, but three hypothesis have been proposed, including [1]; ecto-endodermal adhesion which results in incomplete germ cell layer separation [2]. The cysts are of ectodermal origin, and being part of the primitive streak they are capable of differentiation to both endoderm and mesoderm [3]. The notochord syndrome theory more commonly seen in the lumbrosacral level, where partial duplication and separation of the notochord leads to a ventral herniation of the yolk sac or remnant gut endoderm through the notochord and subsequent fistula with the amniotic cavity. A cystic mass forms from closure of the fistula as the embryo grows from differentiated cells from the remnant of the foregut/endodermal cell origin [3,5].

The most common MR imaging findings associated with intradural extramedullary epithelial endodermal cysts are non-contrast enhancing lesions that are isointense on T1-weighted sequences and hyperintense on T2-weighted imaging. The appearance of epithelial endodermal cyst, neuroenteric or bronchogenic, on diagnostic imaging is that of a lobulated homogenous mass without an associated mural nodule [2,6]. Surgical resection appears to be the most effective treatment of symptomatic spinal bronchogenic cysts and recurrence is attributed to partial resection, with a rate of 11.6% reported in the literature [2]. It should also not be ignored that the presence of spinal development anomalies such as spina bifida may indicate

further developmental spinal or vertebral malformations including spinal cysts [3,5]. Nine cases of spinal bronchogenic cysts reported in the English literature, all the lesions were intradural extramedullary, and 5 of these arose in the cervical or upper thoracic region, 1 in the lower thoracic spine, 1 in the thoracolumbar region, 1 in the lumbar region, and 1 in the sacral region [6]. In our case report the patient is fifth decade of life and the clinical presentation is an indolent progressive deficit in lower limb neurology and with constipation, a reflection of the lesion compressing his cauda equine. The bronchogenic cyst is a slowly growing lesion owing to the cell micro-architecture including the tight junction between cells [3,5] (Table 2).

Table 2: Summary of the literature review of reported cases of spinal bronchogenic cysts.

Authors, Year	Symptoms	Lesion Location	Type of Lesion	Extent of Resection	Follow-up period
Yamashita et al. [7]	Intermittent neck & left arm pain for 4 years	C6-7	Intradural extramedullary	Total	11 months
Ho & Tiel [8]	Numbness on the right arm & leg for 6 weeks	C5-T2	Intradural extramedullary	Total	-
Wilkinson et al. [9]	Pain, paraesthesia of the right arm for 2 weeks	C3-4	Intradural		
	extramedullary	Partial			1 year
Baba et al.[10]	Suboccipital pain for 1 year	C1	Intradural extramedullary	Total	1 year
Rao et al. [11]	Pain, progressive weakness of rt arm for 6 wks	C2-3	Intradural extramedullary	Total	3 months
Baumann et al. [12]	Acute leg pain	T12-L1	Intradural extramedullary	Partial	3 months
Chongyi et al. [13]	Chronic lumbago for 1 year, progressive weakness & numbness of left leg for 2 weeks	L1	Intradural extramedullary	Partial	-
Ko et al. [14]	Skin dimple on the sacrum	S2	Intradural extramedullary	Total	9 days
Arnold et al. [2]	Chronic lumbago for 6 months, lower limb weakness and paraesthesia, urinary incontinence	T4	Intradural extramedullary	Total	1 year
Yilmaz et al. [15]	Chronic lumbago for 2 months and paraesthesia in both legs	T12	Intradural extramedullary	Partial	6 months

We conclude that bronchogenic cyst should be considered as a differential albeit a rare differential with only 10 reported cases in the literature, in patients presenting with an intradural extramedullary lesion in the lumbar spine [2,7-9,10-15]. Surgical excision offers cure with good results, and in our patient as exhibited in the post-operative MRI and clinical outcome.

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