Clinical Case Studies

Intradural intramedullary dermoid cyst in a 42-year-old man at the L1-L2 region

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ABSTRACT

Background: Intramedullary dermoid cysts within the spine are a rare benign tumor. We present this case, which has atypical presenting symptoms, in order to increase awareness of intradural dermoid cysts.

Clinical presentation: We present here a case of a 42 year old man with a 12-month history of lumbar spinal pain as well progressive left lower extremity loss of strength, as well as numbness and paresthesia radiating into the left foot. Magnetic resonance imaging scan revealed a 4 × 1 × 1.3cm intradural mass at the cauda equina L1-L2 region and was hyperintense in both T1 and T2 causing cord compression. L1-L2 laminectomy and intradural micro resection were performed with successful excision of the suspicious mass. Histopathological review revealed keratinaceous debris and adnexal structures consistent with a dermoid cyst.

Conclusions: Our case is unusual with the other reported cases of dermoid cysts due to superior involvement in the lumbar region compared to other case reports with predominantly lumbosacral involvement. This location of the cyst lead to radicular symptoms, rather than lumbosacral pain and sphincter incompetence that is more commonly represented in the literature.

Background

Dermoid cysts are considered a congenital anomaly, associated with the entrapment of ectodermal elements along the lines of embryonic closure, and are considered hamartomas [1]. They are considered benign tumors, with mature skin appendages, and commonly consist of keratinaceous debris and hair follicles [2]. While dermoid cysts are quite common on the head and scalp in the pediatric population, they consist of <1% of all adult spine tumors, with intramedullary involvement being extraordinarily rare [2,3]. In this patient, no congenital spinal abnormalities were reported, and symptoms were absent before the 4th decade of life. This indicates an atypical representation of the dermoid cyst.

We present this case, which has atypical presenting symptoms, in order to increase awareness of intradural dermoid cysts.

Consent

Written consent has been received from the subject.

Clinical presentation

We present the case of a 42-year-old male who presented to the clinic with a 12-month history of left-sided lower back pain, with radiation into the left foot. He stated that he had progressively worsening weakness in the left foot as well as intermittent numbness and paresthesia. Exam revealed 4/5 strength in the left hamstring, the straight-leg test was negative. Heel and toe walks were performed with difficulty. Lower back corticosteroid injections and physical therapy provided minimal relief. Radiographs of the lower back failed to reveal any pathology; therefore, Magnetic Resonance Imaging (MRI) was performed.

Imaging

There is a 2.6 × 1.2 × 1.0 cm heterogeneous mass in the posterior thecal sac at the L1-L2 level. This is intradural extramedullary with a conus terminating it at L1. This contains internal mixed signal including high T1 signal confirming fat within lesion. This is highly suspicious for an intrathecal dermoid cyst/teratoma (Fig. 1). MRI post contrast study of the lesion did not reveal any uptake of the contrast.

Management: Neurosurgical evaluation consisted of an MRI of the spinal to evaluate for drop metastases and computed tomography (CT) scans of the chest, abdomen, and pelvis. No other tumors or drop metastases were noted. Surgical recommendation was to proceed with laminectomy and intradural tumor resection at the cauda equina L1-L2 region. Surgery was performed and with initial laminectomy exposure of the dura at the L1-L2 region, there was an immediate notice of enlarge-
Fig. 1. Preoperative M.R.I of intradural intramedullary mass. (A) Sagittal T2 MRI revealing hyperintense 4 × 1 × 1.3 cm mass at the L1-L2 region. (B) Sagittal Pre-Contrast T1 MRI of intradural intramedullary mass. (C) Axial T2 MRI of intradural intramedullary mass. (D) Axial T1 MRI of intradural intramedullary mass.

Microdissection techniques under microscopic guidance were used to carefully open the dura. The tumor was noted to be an encapsulated yellowish mass. We carefully dissected the capsule from the surrounding nerves until we reached the area where the capsule was connected to part of the cauda equina. At this point we clamped the proximal tip of the cyst and cut it off from the cauda equina. We were able to not spill any of the cyst content. The capsule opened after placement on a sterile tray. We observed a thick gelatinous consistency with calcified 3-4 mm particles and six 5mm long hairs present. The portion of the tumor that was mostly calcified and clumped within the nerves was carefully dissected and removed. We were able to remove all visible tumor under the microscope, and specimens were sent to pathology. Acid-fast stains, as well as cultures, were sent as well. Neuromonitoring was utilized throughout the case, which remained stable.

Post-operatively the patient noted reduction in his lower back radicular leg pain symptoms and was able to ambulate without any difficulty. His left Hamstring strength was improved to 4+/5. He was discharged to home 2 days after surgery. Treatment of these cysts is primarily with surgical resection and clinical monitoring with regular follow-up for recurrence.

Pathology: Macroscopically, the mass was found to have a smooth wall that encased keratinaceous debris consistent with a dermoid cyst. A single hair was confirmed microscopically, and keratotic debris were recovered along with a keratin pearl. No lining epithelium was identified. These findings are consistent with the adnexal structures typically found within dermoid cysts that distinguish them from epidermoid cysts [1] (Fig. 2).

Discussion

The majority of dermoid cysts (60%) are diagnosed within the first 5 years of life and occur most commonly on the head/scalp [4]. Spinal involvement of dermoid cysts, especially intramedullary spinal tumors are an exceptional minority in the adult population, contributing to less
than 1% of all spinal tumors [3]. When they are discovered, they are more likely to involve the lumbosacral spine (60%) [3].

Dermoid cysts are thought to result from failed separation of neuroectoderm and surface ectoderm and the etiology of these cysts is thought to be from an injury to the ectodermal cells [4]. Commonly, dermoid cysts become symptomatic in childhood and are associated with other congenital spine abnormalities as well as trauma to the area [5]. These cysts most commonly present with compressive symptoms and resultant neurological deficits that help to aid in localization. Most commonly, presenting symptoms can be paralysis and sphincter complications [6]. This case is rather atypical since the patient remained asymptomatic before the 4th decade of life and did not declare a history of other spinal abnormalities or trauma to the area of involvement. This case presents with radicular lower back pain and weakness, rather than the typical aforementioned symptoms. This is due to the upper lumbar involvement of the lesion that is more commonly associated with epidermoid cysts distinguishes it from the published literature [6,7].

When first discovered on MRI, dermoid cysts are viewed as an intradural intramedullary tumor, and the differential diagnosis is extensive (dermoid, epidermoid, meningioma, neurofibroma, glial neoplasm, hemangioblastoma, intramedullary metastasis, lipoma, infectious abscess, etc.) with epidermoid being the most common intramedullary spinal cord tumor in adults [8]. As a result of this expansive differential, the only way to diagnose these lesions is via surgical excision and histopathological examination.

Histopathological examination of dermoid cyst reveals stratified squamous epithelial tissue, lined by an outer collagenous layer [2]. Dermoid cysts are differentiated from epidermoid cysts by the presence of hair follicles, sebaceous glands, and other adnexal structures. This is relevant since epidermoid cysts are more common than dermoid cysts in the lumbar region [3].

Recurrent cysts have been treated successfully with radiotherapy [9]. Intramedullary involvement of dermoid cysts leads to a more difficult surgical resection as the capsule is likely to adhere to the spinal cord and this can make complete resection difficult [10]. This case did involve capsule adherence to part of the cauda equina, but successful micro resection of the entire cyst was accomplished.

Conclusion

Our case is unusual with the other reported cases of dermoid cysts due to superior involvement in the lumbar region compared to other case reports with predominantly lumbosacral involvement. This location of the cyst lead to radicular symptoms, rather than lumbosacral pain and sphincter incompetence that is more commonly represented in the literature. It is important to keep a wide differential for all intramedullary spinal cord masses and to be cautious during surgical resection in order to preserve the maximum amount of neural tissue possible. This case also illustrates the importance of paying careful attention to minimize any cyst rupture or dissemination of cyst contents as failure to adequately control cyst contents can lead to meningeal irritation.

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Declaration of Competing Interest

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.jxnsj.2022.100124.

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