Intramedullary inclusion cysts of the cervicothoracic junction

Report of two cases in adults and review of the literature

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Intramedullary inclusion cysts are extremely rare within the rostral spinal cord. In this case report the authors outline the clinical features and surgical treatment of one dermoid cyst and one epidermoid cyst of the cervicothoracic junction. The authors also include a relevant literature discussion regarding the treatment and the embryological origin of these lesions. (DOI: 10.3171/SPI-07/08/236)

KEY WORDS • dermoid cyst • epidermoid cyst • spinal cord tumor

Dermoid and epidermoid cysts are benign ectopic growths thought to be a consequence of embryological errors during neural tube closure. Their reported incidence within the spinal cord is extremely low, accounting for less than 1% of intramedullary spinal cord tumors.1,2,6,9,12–14,16,17,19–21 They more often involve the lumbosacral region and are extramedullary, and patients harboring these cysts usually present during the first decade of life. Intramedullary lesions are extremely rare, especially within the cervical and upper thoracic spinal cord, with only nine previous cases of cervical intramedullary dermoids6,11,12,16,17,19,21 and a single case of a cervical epidermoid cyst reported in the literature.6,18,23 The low incidence of dermoids in the cervical region is likely related to the embryological process of neural tube closure, which begins in the area of the neural tube destined to become the lower cervical cord and proceeds rostrally and caudally. We report on two cases of intramedullary tumors, one dermoid and one epidermoid, occurring within the cervical cord in adults. Although extremely rare, these lesions should be considered in the differential diagnosis when imaging characteristics are suggestive of an inclusion tumor. Preoperative MR imaging using fat-suppressed sequences can be helpful in distinguishing these lesions from other intramedullary tumors.

Case 1

History and Examination. This 23-year-old man was diagnosed with an intramedullary tumor at the cervicothoracic junction during a workup for episodic bowel incontinence 4 years prior to presentation at our institution. Ultimately, irritable bowel syndrome was diagnosed, and the lesion was considered asymptomatic and thus followed up with serial imaging studies. Three months prior to our initial evaluation, he noted the onset of symptoms consistent with myelopathy, including hand weakness and paresthesias and an unsteady gait. On physical examination, no cutaneous abnormalities were identified. A neurological examination revealed mild triceps muscle weakness bilaterally and mild left extensor hallucis longus muscle weakness. Reflexes were brisk throughout, with bilateral Hoffmann and Babinski signs.

Magnetic resonance imaging revealed a spherical mass within the spinal cord from C7–T1 with two satellite lesions at C-6 and C1–3 (Fig. 1). Signal characteristics were mixed on both T1- and T2-weighted sequences, and there

Abbreviation used in this paper: MR = magnetic resonance.
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Fig. 1. Case 1. a–c: Serial midsagittal MR images of the cervical spine demonstrating a well-circumscribed, intramedullary spinal cord mass lesion at the C7–T1 level, with multiple associated areas within the center of the cord up to C-2. The lesion is heterogeneous in signal intensity but predominantly bright on T1-weighted sequences (a) and relatively bright on T2-weighted sequences (b). The fat-suppressed T2-weighted sequence (c) confirms the lipid content in this lesion. d and e: Axial T2-weighted MR images confirming the intramedullary location of this complex mass and showing incompletely fused spinous processes at C-7 and T-1. Given the signal characteristics and the lack of any significant enhancement (images not shown), the most likely diagnosis is an intramedullary dermoid. If a diffusion weighted image of the cord were obtained, one would expect the lesion to be bright and demonstrate areas of restricted diffusion, a characteristic appearance of these true ectoderm-lined inclusion cysts. f: Postoperative image revealing a cystic cavity after removal of the dominant mass along with residual elements.

was no contrast enhancement. Axial imaging showed incompletely fused spinous processes and a holocord lesion at C-7 and T-1. Fat-suppressed images demonstrated signal characteristics consistent with lipid content in all the lesions identified.

Operation. The patient underwent a standard posterior cervicothoracic procedure with laminectomies from C6–T2 (Fig. 2). Once the dura mater was opened, an expanded spinal cord was immediately identified. A midline myelotomy was performed and tissue was resected for the purposes of decompression and biopsy procedures. The resected tissue had a yellow discoloration and a friable consistency with intermingled well-formed hair. Internal debulking of the lesion was continued until the capsule was reached. This capsule proved to be adherent to the surrounding neural tissue; therefore, we decided to perform a subtotal resection. A dural patch graft was sewn into place to enlarge the intrathecal volume to prevent recurrent growth. Because of the spinal abnormalities identified and the atrophy observed in the posterior cervical musculature, we decided to incorporate a posterior cervicothoracic stabilization and fusion.

Postoperative Course. The postoperative period was uneventful and the patient was discharged on postoperative Day 5. At the 3-month follow-up he reported improvement in all of his preoperative deficits including hand strength, sensation, and ambulation. Follow-up MR images demonstrated residual tumor at the resection site with no change in the rostral lesions. Ample space had been created at the site of tumor resection as a result of the duroplasty (Fig. 1f).

Case 2

History and Examination. This 61-year-old woman presented with an 8-year history of left arm pain, new-onset right arm pain radiating to the fourth and fifth digits, and recent falls attributed to difficulty in controlling her left side. Her medical history was unremarkable. On examination we noted a small area of abnormally long, dark hair off-midline, adjacent to T-4. On physical examination she demonstrated mild weakness and atrophy of the intrinsic hand muscles, particularly in an ulnar distribution; otherwise, her muscle strength was normal. Her muscle tone was mildly increased throughout the left side, and the left side was diffusely hyperreflexic, although no pathological reflexes were elicited. Her sensory examination was grossly normal with the exception of slightly diminished distal left leg proprioception.
Imaging revealed a complex intradural lesion extending from C3–T1 with both intramedullary and extramedullary components and a focal mass at C-6 and C-7 with adjacent expansion of the osseous spinal canal (Fig. 3). Signal intensity on both T1- and T2-weighted images was heterogeneous, and the lesion did not enhance after contrast administration. An epidural defect was noted in association with the C-7 spinous process.

After midline incision and subperiosteal dissection, standard laminectomies were performed at C-6, C-7, and T-1 along with medial facetectomies. The osseous anatomy appeared completely normal, and no dermal sinus tract was identified. The dura was opened in the midline, and a durotomy was extended to expose the visible rostral and caudal extensions of the tumor (Fig. 4). The tumor was white and pearly with a large exophytic component, and during internal debulking it appeared to be composed mostly of flaky keratinized debris typical of an epidermoid cyst. The resection was continued from the inside out by using an ultrasonic aspirator until neural tissue could be visualized through the thinned-out tumor capsule all along the tumor margins. After hemostasis, the durotomy was repaired primarily, and the subarachnoid space was filled with warm saline before finishing a watertight closure. Somatosensory evoked potentials had revealed some baseline asymmetry and remained unchanged throughout the case.

Postoperative Course. The postoperative course was uncomplicated. After 2 days of bed rest, the patient was ambulatory and discharged home on postoperative Day 5. Currently, 18 months have passed since her surgery, and she still experiences bilateral arm pain and left leg spasticity but she has no functional impairment. Follow-up imaging reveals excellent cord decompression with near-complete resection of the dominant lesion.

Discussion

History and Embryological Characteristics

As noted in von Bostroem,24 the earliest description of a dermoid is attributed to Verratus (1745). The rarity of dermoids associated with the spine was recognized as early as 1889 by Sir John Bland-Sutton who, in a monograph describing occurrences of dermoids throughout the body, reported a single case of a 2-year-old boy with a lumbar dermoid associated with a “hairy patch” and “defective spines.” The earliest reference to an epidermoid cyst is attributed to Cruveilhier in 1830 who dubbed them “tumeurs perlées,” again in von Bostroem.24

As cases of dermoid and epidermoid cysts within the neuraxis along with cases of teratomas and lipomas were collected and reported, it was theorized that all these lesions might represent ectopic tissues arising from similar processes of embryological dysregulation. The first connection between inclusion cysts and developmental errors is attributed to von Remak, but this notion was greatly expanded on in 1897 by von Bostroem24 who set forth the classic, often-quoted theory: between the 3rd and 5th weeks of fetal development, cells fated for cutaneous ectoderm somehow become trapped within neural ectoderm during neural tube closure and eventually form tumors. According to this theory, the timing of the event (early or late)
determines cell potentiality, which in turn determines the type of tumor (teratoma or dermoid or epidermoid) that will form. The predilection for poles of the neuraxis, in particular the caudal end, is in some way related to the fact that the poles are the last segments of the neural tube to close.

In 1933 Holmdahl attempted to connect the high incidence of lumbosacral lesions to his discovery that the caudal spine formed not from ectoderm but from a distinct, undifferentiated cell mass belonging to no particular germinal layer. Because cells from these lumbosacral precursors were less differentiated than ectoderm, he postulated that they were more likely to form tumors with pluripotentiality. Over the next 20 years, numerous authors challenged this idea, providing evidence that ectodermal cells could revert to form cells of other germinal layers. A rival theory was proposed, stating that teratomas, dermoids, and epidermoid cysts of the neuraxis arise from cells whose position is correct but that somehow fail to receive the correct differentiation signals. Interestingly, very little has been done to clarify these issues in the last 50 years. Indeed, recognition of the ability of many differentiated cells to maintain or revert to a pluripotential state under the correct experimental conditions challenges many classic assumptions regarding cell fate.

**Epidemiological Features**

In the absence of conclusive experimental data, we have...
accrued a good deal of clinical information that is useful to a modern clinical practice and may ultimately help to shed some light on the genesis of dermoid tumors. Some authors have used a collection of cases reported prior to 1978 to quote a 5% frequency rate of dermoids in the cervical spine. Taking into account redundant reporting, we found a total of 180 cases of spinal dermoid tumors in the literature, 2% of which involved any level above T-2. Of these, only five were intramedullary and two were associated with a dermal sinus tract, which had been removed 20 years prior to presentation of the mass (Table 1). A similar review for intramedullary epidermoid cysts was performed by Roux et al., who found 47 cases in the literature; in none of these cases did the tumor extend into the cervical cord. The only reported case of an epidermoid involving the cervical cord is from Higazi, who described an extramedullary epidermoid lesion adjacent to C-4 in a 5-year-old patient with a dermal sinus tract.

More instructive and likely more accurate numbers regarding the frequency and location of dermoid and epidermoid lesions in the spinal cord are to be gleaned from large case series. Of the 680 primary spinal tumors mostly in adult patients who underwent surgical treatment at La Sapienza in Rome between 1952 and 1986, 10 of the 479 extramedullary lesions and six of the 201 intramedullary lesions were dermoid or epidermoid cysts. None involved the cervical or upper thoracic cord. Two large series of intramedullary tumors in 51 and 78 adults each included a single case of a dermoid or epidermoid lesion; neither of these tumors were found in the cervical spine. Combining results in these series produces an incidence of 1.8% of intramedullary dermoid or epidermoid lesions in adults and no cervical cases. In the pediatric population, the prevalence of dermoid and epidermoid tumors is much greater, with dermoids accounting for 5 to 17% of all intradural lesions in reported series. Interestingly, of the

Fig. 4. Case 2. Intraoperative photographs obtained in a 61-year-old woman with an intradural lesion. a: A patch of hypertrichosis (arrow) was noted off-midline at around the T-4 spinous process level. b: After laminectomy and midline durotomy, a shiny, glistening intramedullary mass with a large exophytic component was readily apparent. c: The extramedullary portion was excised, and internal debulking was performed on the intramedullary portion of the lesion whose capsule proved to be densely adherent to neural tissue and could not be safely dissected free.
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Clinical Management and Prognosis

Dermoid and epidermoid tumors are benign, indolent, congenital lesions. In the absence of an associated dural sinus tract, resection is indicated for symptomatic progression and compression of neural elements on neuroimaging. In the presence of a dural sinus tract, surgery is mandated either to treat or to avoid bacterial meningitis. In the literature, bacterial meningitis during presentation appears to be a greater problem in older patients, presumably because of the prevalence of diagnostic delays in that population. Preoperative chemical meningitis in association with spinal inclusion cysts has rarely been reported, but postoperative chemical meningitis appears to be more prevalent and may be associated with residual tumor after resection. Patients who have aseptic meningitis postoperatively can also have chemical meningitis. Preoperative imaging has questionable value. In our experience, revision surgery for recurrent tumor should be reserved for patients demonstrating a clinical deterioration. This strategy has led to local control and stable symptoms in patients with more than one decade of follow-up, although symptomatic recurrences will arise in a small percentage of patients and thus mandate long-term clinical follow-up.

<table>
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<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex</th>
<th>Tumor Type</th>
<th>Lesion Location</th>
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</table>

*NP = information not provided.

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