

Review Article

A challenging case of an intramedullary dermoid cyst of the cervical spine - A literature review



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ABSTRACT

A comprehensive differential diagnosis of intramedullary lesions of the sub-axial cervical spine must begin with the most common namely the diffuse fibrillary astrocytoma. Ependymoma's are not as common at this site and even rarer are epidermoids; teratomas; hemangioblastomas; lipomas, and drop metastases. Extremely rare lesions are intramedullary dermoid cysts and intramedullary schwannomas. We aimed to review the topic of intra-medullary dermoid cysts of the sub-axial cervical spine.

Dermoid tumors are histologically distinct from epidermoids in that they are lined with dermis containing skin appendages for example hair and sebaceous glands. Epidermoids on the other hand are lined with stratified squamous epithelium and contain only desquamated keratin. While congenital dermoid cysts account for only 1% of spinal cord tumors they are known to have a predilection for the lumbosacral spine. This predilection sees a 60% incidence at this site with only 10% occurring in the thoracic spine. A mere 5% of dermoid cysts occur in the cervical spine. Dermoid cysts are considered to be hamartomas that usually occur when mesodermal tissue becomes incorporated into the spinal canal during folding of the neural plate and the formation of the subsequent neural tube. While usually intradural and extramedullary in a dorsal location they do occur less commonly at an intramedullary site. An MRI is the diagnostic modality of choice and the features of fluid, soft tissue, calcium and fat are regarded as diagnostic of a dermoid tumor.

Albeit rare dermoid cysts do occur at a purely intramedullary location in the sub-axial cervical spine and a comprehensive differential diagnosis must include this tumor type. Whilst displaying linear growth and not the exponential growth of a true tumor the cyst wall of these lesions demand meticulous resection techniques.

1. Introduction

A comprehensive differential diagnosis of intramedullary lesions of the sub-axial cervical spine must begin with the most common namely the diffuse fibrillary astrocytoma although mixed neuroglial tumors such as gangliogliomas do occur in children. Ependymoma's are not as common at this site and even rarer are epidermoids; teratomas; hemangioblastomas; lipomas, and drop metastases. Extremely rare lesions are intramedullary dermoid cysts and intramedullary schwannomas [1]. We aimed to review the topic of intra-medullary dermoid cysts of the sub-axial cervical spine.

Dermoid tumors are histologically distinct from epidermoids in that they are lined with dermis containing skin appendages for example hair and sebaceous glands. Epidermoids on the other hand are lined with stratified squamous epithelium and contain only desquamated keratin. These two lesions must also be distinguished from mature cystic teratomas which contain derivatives from more than one embryological

cell layer [2]. While congenital dermoid cysts account for only 1% of spinal cord tumors, they are known to have a predilection for the lumbosacral spine. This predilection sees a 60% incidence at this site while 10% occur in the thoracic spine. A mere 5% of dermoid cysts occur in the cervical spine [3–6]. Epidemiologically spinal dermoid cysts have an increased prevalence in the pediatric population where they represent up to 8% of intra-spinal tumors [7]. Due to their slow growth in a linear fashion with the body they commonly present themselves during adolescence [8]. Their presentation in children is very rare and in a review of 1234 spinal cord tumors in children, although 197 were situated in the cervical region, none were dermoid tumors [9]. In our review we noted only three papers reporting isolated intramedullary dermoid cysts of the cervical spine without associated spinal dysraphism on MRI that were confirmed on intra-operative features and post-operative histology [10–12].

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Fig. 1. Pre-operative MRI: T1 weighted MRI sagittal image showing the hyper-intense fat content of an intramedullary spinal dermoid cyst. The anterior vertebral body scalloping eludes to the chronicity of the disease and posteriorly the markers of a previous resection attempt namely multilevel laminectomies are visible.

2. Materials and methods

We present an unfortunate case of a 14 year old female patient whom presented to our unit with a 2 day history of sudden onset quadriplegic and associated respiratory distress. The child had been diagnosed with an intramedullary dermoid tumor of her cervical spine as much as 10 years prior to this admission and had undergone a debulking procedure at that time. Since then she had, according to her mother, been doing well and had hence not attended her follow-up appointments nor her surveillance MRI imaging. Her MRI was repeated which showed an impressive tumor almost completely replacing the involved cervical spinal cord [Figs. 1–3]. Under neurophysiological

monitoring she was taken for emergency surgery and the tumor displayed the typical characteristics of her prior diagnosis [Fig. 4–5]. Unfortunately intra-operatively there was a gradual loss of SSEP and MEP action potentials indicating cord infarction and post-operatively she was ventilator dependent and quadriplegic. Several months later she succumbed to a ventilator acquired pneumonia.

We conducted a PubMed search using keywords “intramedullary cervical dermoid cysts” or synonyms thereof. The articles reviewed were analyzed for epidemiological variables, tumor characteristics and the optimal surgical technique for resection. This review explores these variables and thereby provides a valuable insight into a rare site of occurrence of this histological tumor type.

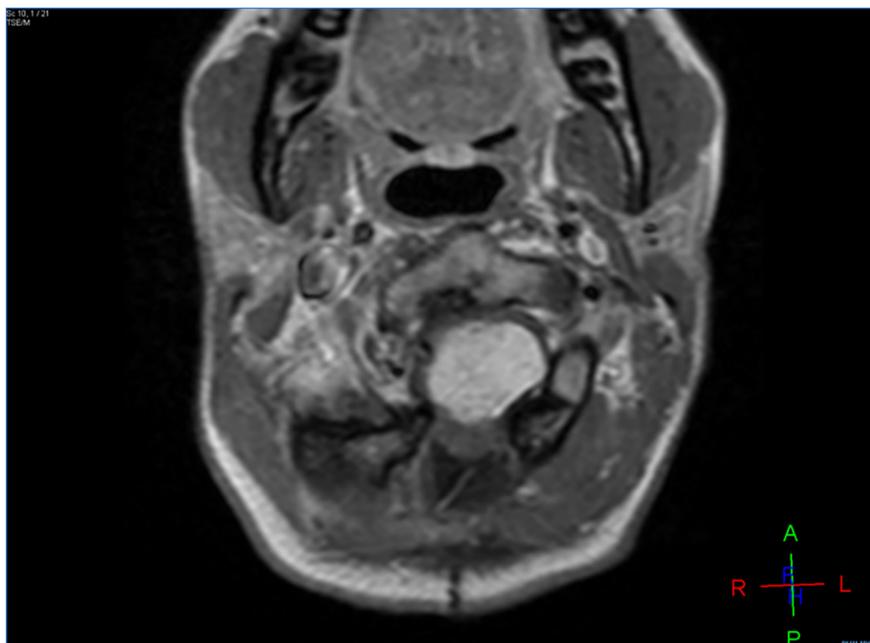


Fig. 2. Pre-operative T1 weighted MRI: axial view showing the purely intramedullary location of this spinal dermoid cyst which has almost completely replaced the cord.



Fig. 3. Pre-operative T2 weighted MRI: showed the heterogeneity of the cyst contents. Note again the wide opening of the foramen magnum eluding to the chronicity of the disease.

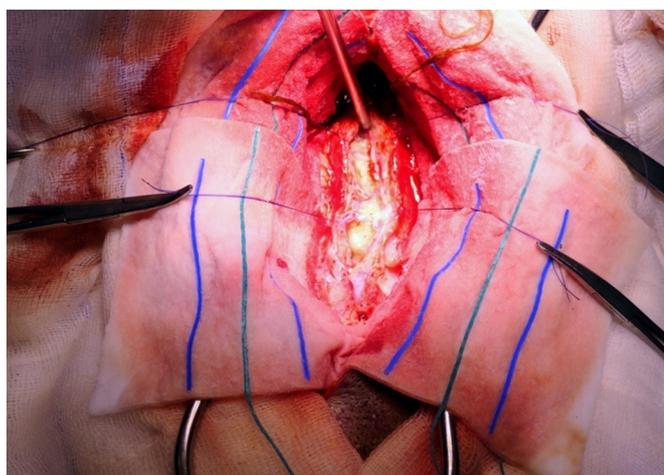


Fig. 4. Intra-operative photograph: upon opening of the dura. Note the sebaceous material within this intramedullary dermoid cyst.



Fig. 5. Intra-operative photograph: showing the accompanying benign dermal derivatives of sebaceous material and hair which spilled from the lesion.

3. Results

Dermoid cysts are considered to be hamartomas that occur when mesodermal tissue becomes incorporated into the spinal canal during folding of the neural plate and the formation of the subsequent neural tube. While usually intra-dural and extramedullary in a dorsal location they do occur less commonly at an intramedullary site [10].

Albeit rare endodermal involvement does occur also the mechanism is more complex and several theories are currently debated. The first of these is that during the early embryonic period in the anterior part of the trilaminar germ disc, in the region of the buccopharyngeal membrane adjacent to the neural plate, there is a time when the developing notochordal plate becomes, for a brief period, part of the roof of the developing foregut—a process called incalation. The notochordal plate would then, as part of normal development, separate from the endoderm to situate itself between the neuroectoderm and the primitive foregut—a process of excalation. Incomplete excalation is postulated as leading to diverticula of endodermal tissue becoming trapped within

the spinal canal and explains the presence of this tissue in spinal dermoid cysts [13]. The alternative “split notochord theory” proposes the causality for endodermal derivatives in dermoid cysts as occurring earlier in embryonic development at the stage when the germ disc is still bilaminar. Adhesions between the neuroectoderm and the endoderm prevent the normal complete separation of these layers. These adhesions obstruct the developing notochord as it grows in a cephalad direction and this in turn causes a cleft in the notochord containing endodermal germ cells in association with the derivatives of the ectodermal and mesodermal layers [14]. A third etiology is inoculation dermoid cysts which occur iatrogenically during for example the repeated lumbar punctures associated with tuberculous meningitis complicated by hydrocephalus [15]. This theory was confirmed in an animal model using rats [16].

The innocuous growth of a spinal dermoid cyst in the childhood years may go unnoticed as the child remains asymptomatic. The presentation of these tumors during adolescent years can either be by gradually progressive neurological deterioration or by a catastrophic

neurological event [17,18]. This acute presentation can be due to the development of hydrocephalus [19] or cyst rupture into the subarachnoid space where the patient will present with symptoms that include headache, nausea and vomiting, visual disturbances, vertigo, cognitive disturbances and coma. Chemical meningitis from irritation of the meninges by the fat content of the cyst in the subarachnoid space will present with pyrexia, vomiting and meningism, making it difficult to distinguish clinically from other causes of meningitis [18,20].

The computed tomographic features of a spinal dermoid cyst are that of a well-defined lesion with distinct borders; the presence of calcification or teeth may be seen. An MRI is however the diagnostic modality of choice. The MRI features of fluid, soft tissue, calcium and fat are regarded as diagnostic of a dermoid tumor [21,22]. The lesion is often heterogeneous, however the fat content in the tumor is clearly visible on T1W images and is especially specific for showing the fatty sebaceous secretions, liquid lipid metabolites and cholesterol in the subarachnoid space that occurs with cyst rupture [18,21,22].

Differentiating mature cystic teratomas from dermoid cysts without histological evidence is a diagnostic dilemma as the teratoma is similarly comprised of derivatives from more than one germ cell layer [23]. This becomes even more difficult as the absence of endodermal derivatives, similarly to a dermoid cyst, does not exclude the diagnosis. A teratoma is however a true neoplasm, displaying a growth rate that is accelerated as compared to the normal body tissues [24]. On MRI this may be reflected by the tumor cells of a teratoma showing contrast-medium enhancement as compared to the non-enhancing benign dermoid cyst. Only on histological grounds, by displaying the cellular changes of tumoral cells, can this differentiation be conclusively made [23].

Intra-operatively while complete resection of a dermoid cyst should be the goal of therapy with emphasis on preventing spillage of cyst contents there is a real risk of complications if the cyst ruptures or if the wall is resected as it is often adhered to the surrounding spinal cord tissue [5,25]. While the dermal cyst contents are relatively easy to be removed the intramedullary dermal cyst wall fuses with the surrounding neural tissue at the embryological stage making removal difficult without causing damage [26]. Another paper reported a high rate of total removal of intramedullary dermoid cysts and emphasizes the importance of intra-operative neurophysiological monitoring as an adjunct to ensuring a good functional outcome [27]. Recurrent surgery is however not uncommon despite gross total resection having been achieved and the mechanisms behind this are poorly understood [26]. Several studies report that failure to remove the cyst wall completely incurs a high recurrence rate and this should always be the ultimate goal of surgery if it can be done safely [26,28].

4. Conclusion

Albeit rare dermoid cysts do occur at a purely intramedullary location in the sub-axial cervical spine and a comprehensive differential diagnosis must include this tumor type. Whilst displaying linear growth and not the exponential growth of a true tumor the cyst wall of these lesions demand meticulous resection techniques.

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Declaration of Competing Interest

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References

- [1] B. Steyn, Intramedullary spinal cord tumors, *Clin. Neurosurg.* 308 (1983) 717–741.
- [2] B. Karaaslan, G. Ulku, M. Ucar, T. Demirdag, A. Inan, A. Borcek, Intramedullary dermoid cyst infection mimicking holocord tumor: should radical resection be mandatory?—a case report, *Childs Nerv. Syst.* 32 (2016) 2249–2253.
- [3] A. Shuba, S. Mohanty, K. Das, I. Garg, Congenital inclusion tumours in spinal dysraphism, *Indian J. Pediatr.* 77 (2010) 167–170.
- [4] S. Kumar, S. Gupta, V. Puri, R. Gupta, R. Malik, Intramedullary dermoids in children, *Indian Pediatr.* 27 (1990) 626–629.
- [5] M. Najjar, J. Kusske, A. Hasso, Dorsal intramedullary dermoids, *Neurosurg. Rev.* 28 (28) (2005) 320–325.
- [6] C. List, Intraspinal epidermoids, dermoids, and dermal sinuses, *Surg. Gynecol. Obstet.* 73 (1941) 525–538.
- [7] Yamamoto Y, Raffel C. Spinal extradural neoplasms and intradural extramedullary neoplasms. In Albright A, Pollack I, Adelson P. Principles and Practice of Pediatric Neurosurgery. Thieme, New York, 685–696.
- [8] M. Dias, M. Walker, The embryogenesis of complex dysraphic malformations: a disorder of gastrulation? *Pediatr. Neurosurg.* 18 (1992) 229–253.
- [9] N. Di Lorenzo, R. Giuffre, A. Fortuna, Primary spinal neoplasms in childhood: analysis of 1234 published cases (including 56 personal cases) by pathology, sex, age and site. Differences from the situation in adults, *Neurochirurgia* 25 (1982) 153–164.
- [10] A. Olgen, A. Khandjii, P. McCormick, M. Kaiser, Intramedullary inclusion cysts of the cervicothoracic junction, *J. Neurosurg. Spine* 7 (2007) 236–242.
- [11] A. Patankar, J. Sheth, Dermoid cyst: a rare intramedullary inclusion cyst, *Asian J. Neurosurg.* 7 (2) (2012) 81–83.
- [12] F. Signorelli, N. Montano, A. Scerrati, B. Cioni, F. Papacci, Isolated intramedullary cervical dermoid cyst, *Acta Neurol. Belg.* 114 (2014) 223–224.
- [13] M. Fallon, A. Gordon, A.C. Lendum, Mediastinal cysts of fore-gut origin associated with vertebral abnormalities, *Br. J. Surg.* 41 (1994) 154–159.
- [14] J.F. Bentley, J.R. Smith, Developmental posterior enteric remnants and spinal malformations: the split notochord syndrome, *Arch. Dis. Child.* 35 (1960) 76.
- [15] R. Wilkins, G. Odom, Spinal intradural cysts: epidermoid and dermoid cysts, in: P. Vinken, G. Bruyn (Eds.), *Handbook of Clinical Neurology*, Vol 20, Chp 4, North-Holland, Amsterdam, 1976, pp. 73–82.
- [16] S. Kumar, R. Gulati, S. Mann, Intraspinous dermoids, *Neurochirurgia* 20 (1977) 105–108.
- [17] M. Yasargil, C. Abernathy, A. Sarioglu, Microsurgical treatment of intracranial dermoid and epidermoid tumors, *Neurosurg* 24 (1989) 561–567.
- [18] P. Barsi, J. Kenez, G. Varallyay, L. Gergely, Unusual origin of free subarachnoid fat drops: a ruptured spinal dermoid tumour, *Neuroradiology* 34 (4) (1992) 343–344.
- [19] R. Martin, A. Krone, B. Schuknecht, W. Kuhn, Rapid development of occlusion hydrocephalus by intraventricular fat possibly derived from a ruptured dermoid cyst, *J. Neurol. Neurosurg. Psychiatry* 52 (1989) 134–135.
- [20] T.A. Scearce, C.M. Shaw, A.D. Bronstein, P.D. Swanson, Intraventricular fat from a ruptured sacral dermoid cyst: clinical, radiographic, and pathological correlation. Case report, *J. Neurosurg.* 78 (4) (1993) 666–668.
- [21] J.G. Cha, S.H. Paik, J.S. Park, S.J. Park, D.H. Kim, H.K. Lee, Ruptured spinal dermoid cyst with disseminated intracranial fat droplets, *Br. J. Radiol.* 79 (938) (2006) 167–169.
- [22] A. Garg, V. Gupta, S. Gaikwad, et al., Isolated central canal rupture of spinal dermoid: report of two cases, *Australas. Radiol.* 47 (2) (2003) 194–197.
- [23] H. Altay, O. Kitis, C. Calli, N. Yunten, A spinal dermoid tumor that ruptured into the subarachnoid space and syrinx cavity, *Diagn. Interv. Radiol.* 12 (4) (2006) 171–173.
- [24] T.J. Rosenbaum, E.H. Soule, B.M. Onofrio, Teratomatous cyst of the spinal canal. Case report, *J. Neurosurg.* 49 (2) (1978) 292–297.
- [25] Slooff J. Primary Intramedullary Tumors of the Spinal Cord and Filum Terminale. Saunders, Philadelphia.
- [26] S. Guo, Y. Xing, A review of five cases of intramedullary dermoid cysts, *Childs Nerv. Syst.* 30 (2014) 659–664.
- [27] I. Sandalcioğlu, T. Gasser, S. Asgari, A. Lanzorisak, T. Engelhorn, T. Egelhof, D. Stolke, H. Wiedelmayer, Functional outcome after surgical treatment of intramedullary spinal cord tumors: experience with 78 patients, *Spinal Cord* 43 (1) (2005) 34–41.
- [28] S. Maurice, F. Burstein, Disappearing dermoid: fact or fiction? *J. Craniofac. Surg.* 23 (1) (2012) e31–e33.