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Dermoid Cysts of the Conus Medullaris Associated with Sacral Meningocele. A Case Report

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Abstract

Spinal dermoid cysts are gradually progressive benign tumors that may be congenital or acquired. They account for 0.8% to 1.1% of all primary spinal tumors. MRI is the gold standard for the radiological assessment. Management is difficult for tumors involving highly eloquent areas such as the conus medullaris. In this report, we present a rare case of an adult woman with dermoid cysts in the conus medullaris associated with a sacral meningocele which was treated with a midline myelotomy that drained yellowish keratinous fluid and decompressed the cyst. No aggressive attempt at complete resection of the cyst wall was undertaken. The patient fully recovered, and her condition considerably improved after surgery and remains good at a 6-month follow-up.

Keywords

Dermoid Cysts, Intramedullary, Meningocele

1. Introduction

Spinal dermoid cysts are rare, benign, gradually progressive lesions that may be congenital or acquired [1]. They usually appear in childhood or adulthood with a history of trauma [1] [2] [3]. Spinal dermoid cysts are covered by keratinized squamous epithelium and can be distinguished from epidermoid cysts by the presence of dermis and dermal glands [4]. Spinal dermoid cysts ought to be considered as a distinct entity due to their anatomic localization, their surgical management, and their clinical course.

Spinal dermoid cysts are most commonly located in the lumbosacral region (cauda equina and conus medullaris), followed by the upper thoracic and cervical regions [5] [6]. They can be located intramedullary, intradural-extramedullary and extradurally [7] [8]. In some instances, spinal dermoid cysts are associated with bony malformations, myelomeningoceles, syringomyelia, hypertrichosis and/or dermal sinus tract [6] [9].

The diagnosis is established following a long-time lapse if the initial symptoms are considered, leading to tumors of large or even giant size [6] [7] [8] [9].

In this report, we describe a rare case of dermoid cysts in the conus medullaris associated with a sacral meningocele. Most publications addressing this topic are rare and only few case reports have been described in the literature [6] [12] [13].

This case confirms the importance of the pretherapeutic neurological status in functional prognosis.

2. Case Report

A 41-year-old woman presented with a 6-year history of low back pain progressing to poorly systematized bilateral sciatica. She complained of intermittent pollakiuria and paresthesia. She had no history of trauma, infection, lumbosacral surgery, congenital anomaly, or developmental disorders. On neurological examination, the muscle strength and deep tendon reflexes were normal. Urinary and bowel functions were intact.

MRI evidenced a large intradural mass located at the conus medullaris and attached to a fatty filum. According to the signal intensities on the MRI, the mass could be divided into two components (**Figure 1**). The upper component of the mass, which accounted for approximately two-thirds of the lesion, is located intramedullary and was iso- to hypointense on T1-weighted images and hyperintense on T2-weighted images relative to the spinal cord. The signals were heterogeneous on both T1- and T2-weighted images. In contrast, the lower component exhibited homogeneous hyperintensity on both T1- and T2-weighted images and was connected to the tight *filum terminale*. The mass was not enhanced after administration of gadolinium. Posterior rachischisis in the sacrum with meningocele extended from L5 to S4. Axial MRI sections confirmed the intramedullary location of the tumor (**Figure 2**).

An incision was made in the skin between the levels of the spinous processes of L1 and L2, and laminectomies were performed at L1 and L2. The dura mater was intact and incised at the midline (**Figure 3**). The spinal cord was bulging at the L2 level and the intradural mass was located in the conus medullaris. Tumor removal started by reducing the volume of the lesion with an ultrasonic aspirator before looking for a cleavage plane. Intratumoral resection was performed from inside to outside under a surgical microscope. After strict control of hemostasis, dissection was started laterally, on the side on which resection proved easiest. The rostral margin was adjacent to the shiny yellow intramedullary mass via arachnoid cysts. Using the microscope, evacuation of the intramedullary mass was performed.

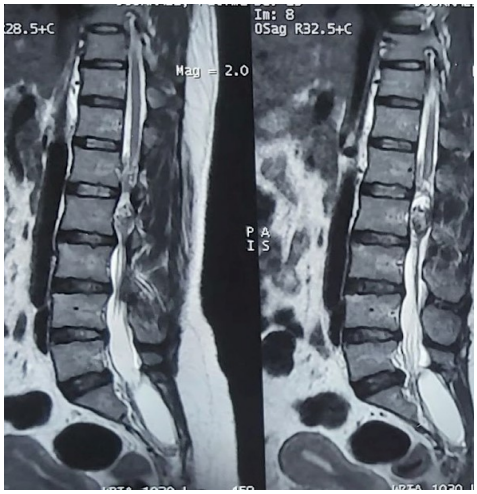

Figure 1. MRI sagittal sections. T2-weighted sequences showing the cyst in the conus medullaris and a sacral meningocele.

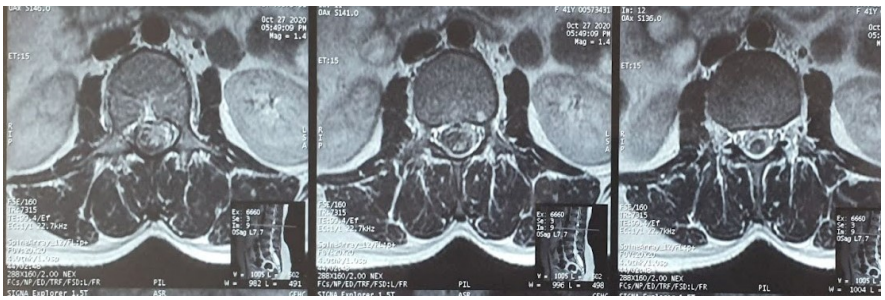

Figure 2. MRI axial sections. T2-weighted sequences showing the intramedullary tumor in the conus medullaris.

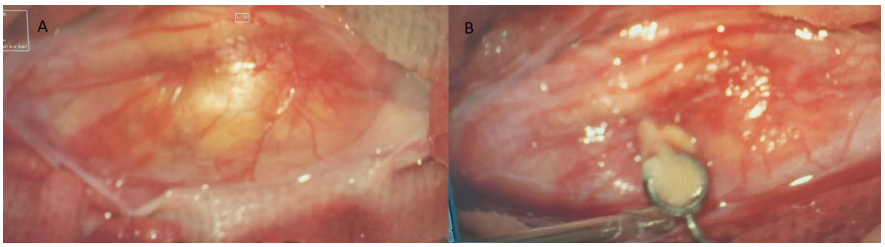

Figure 3. Intraoperative view of the yellowish cyst after incision of the dura mater (A) and the arachnoid membrane (B).

To explore the rostral margin of the intramedullary mass, another small window was made at approximately 3 cm rostral to the conus medullaris along the posterior median sulcus. The evacuation of the intramedullary mass was performed under microscope using a ring curette and suction through the small myelotomy. A yellowish, lipid rich material that contained tiny hairs and a small amount of calcification was collected. Subsequently, untethering of the cord with careful electrophysiological monitoring was done. Tumor removal begun by reducing the volume of the tumor with an ultrasonic aspirator. After strict control of hemostasis, dissection was started laterally. The pial edges were sutured with 5 - 0

nylon sutures, and the dura mater was closed using vicryl 4 - 0 sutures. Motor evoked potential (MEP) monitoring showed intact function throughout the surgery. Postoperatively, the patient did not show neurological deterioration.

Histopathological examination of the paraffin-embedded samples showed that the intradural cyst mainly consisted of mature adipocytes and contained Pacinian corpuscles. The intramedullary mass contained abundant yellowish lipid-rich debris in which hairs and a small amount of calcification were present. The wall of the mass was lined with stratified squamous epithelium and contained sebaceous glands and sweat glands, which confirmed the diagnosis of a dermoid cyst.

At 6-months follow-up visit, the patient showed a good evolution with a complete disappearance of symptoms described at admission to our department.

An informed consent has been obtained from the patient for the purpose of reporting her case.

3. Discussion

Spinal dermoid cysts are rare slow-growing benign lesions, accounting for 0.8% to 1.1% of all spinal tumors [6]. They may be congenital, believed to arise from ectopic embryonic remnants of ectoderm within the spinal canal at the time of neural tube closure or acquired following trauma or surgery [2] [3].

Despite occurring at any age group, these lesions are rare and only account for 2% to 4% of all central nervous system tumors and 15% of all primary intradural tumors in adults [10]. Symptoms may vary depending on the location, extent of the lesions and are consequence of compression of adjacent structures by the tumors. Motor disturbances, pain, sensory disturbance, and urological dysfunction often occur [11] [12].

Spinal dermoid cysts are often associated with some form of spinal dysraphism including bony or split cord malformation, hypertrichosis, dermal sinus tracts, meningocele as it was the case of our patient, or a combination of these elements [6] [9].

Even if a diagnosis cannot be made without histopathological examination, certain neuroimaging findings are characteristic and useful in helping to distinguish inclusion cysts from other more common spinal lesions. Presently, MRI is the best and, in most cases, the only examination to perform in investigating these cases [13]. On MRI, the tumor appears hypointense on T1-weighted sequences and hyperintense on T2-weighted ones as compared with the conus medullaris, which behaves like CSF. CT scan helps in confirming the fatty composition of the tumor [14] [15]. The differential diagnosis of dermoid cysts on imaging includes lesions with high lipid content such as teratomas and lipomas. Other differential diagnostic possibilities include cysticercosis cyst, epidermoid cyst and myxopapillary ependymoma [15].

The management of patients with intramedullary spinal cysts remains controversial. Given the slow progression and the benign nature of these tumors, some authors are in favor of conservative management for asymptomatic lesions [9] [16]. On the other hand, even if complete removal of the cyst wall may re-

duce the recurrence rate, it can also involve a risk of spinal cord damage that may cause severe postoperative neurological deterioration [17] [18]. In fact, most cases of spinal dermoids cannot be totally excised, as demonstrated in many surgical series [6] [9] [14] and the risk of cyst recurrence after incomplete removal has not been reported to justify the risk involved in attempting complete cyst wall resection [12] [19]. In those patients, where the resection of the lesion was incomplete, some authors have recommended clinical observation until tumor re-growth gives rise to novel symptoms, which will only happen in a small number of patients with long-term follow-up [9] [20].

At surgery, these tumors are usually filled with a fatty, soft, whitish yellow waxy substance that contain hair and sometimes nails [9] [21]. Dissemination to the central nervous system might exist, but most of the time this dissemination consists in fatty droplets, with no evolution [14].

Histologically, spinal dermoid cysts are usually surrounded by a multi-layered, cornified epithelium, with a collagen stroma, lying in the dermis and hypodermis with cutaneous appendages, such as hair follicles, hair, sebaceous and sweat glands, as well as sebum. Calcifications can also be observed in the cyst content [14] [20]. Blood vessels usually are detected only in the connective tissue surrounding the tumor and they do not penetrate the epithelial wall of the dermoid cyst [20].

The use of postoperative radiation therapy in the tumor recurrence has been described by some literature reports [18]. The authors suggest that radiation therapy might have an effect in preventing future cystic re-accumulation in partial resections and may be an alternative to surgery in tumor recurrences [21]. In any case, the effectiveness of radiotherapy is still unclear and has not been demonstrated, especially for those patients with low-grade tumors [18]. In our patient no postoperative radiotherapy was performed and at 6 months follow-up, the patient has had a good evolution with disappearance of symptoms at admission.

4. Conclusion

Dermoid cysts usually carry a favorable prognosis. The management of the surgical resection remains a challenge for all neurosurgeons. Whenever possible, total tumor removal should be performed for preventing cystic recurrence. However, subtotal extirpation is a reasonable treatment when the lesions are tightly adhered to the nervous tissue and carry a risk of neurological deterioration with gross tumor removal. At present, magnetic resonance imaging (MRI) is the best and, in most cases, the only examination to perform in investigating these cases.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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