Multiple Intramedullary and Intradural Epidermoid Cysts in the Conus Medullaris and the Lumbar Spine
- Case Report -

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Intramedullary epidermoid tumors are rare. To our knowledge, this is the first description of a multiple intraspinal epidermoid tumor, including the intramedullary conus medullaris and an intradural lesion in the lumbar spine. The authors report the case of a 43-year-old woman presenting with progressive paraparesis. A thoraco-lumbar lesion was confirmed as an epidermoid cyst from histopathology. The clinical, radiological and surgical features, as well as a brief critical review of the literature are included.

KEY WORDS: Multiple epidermoid cysts · Intramedullary tumor.

Introduction

Intraspinal epidermoid cysts account for less than 1% of all intraspinal tumors in adults. Intramedullary localization of an epidermoid cyst is extremely rare. To date, approximately fifty cases with an intramedullary epidermoid have been reported. However, multiple intraspinal epidermoid tumors have not previously been reported. We review the literature regarding intramedullary epidermoid tumors, and present this case.

Case Report

A 43-year-old woman visited our hospital with a 20-year history of back pain and progressive paraparesis, which began with left foot drop and then extended to both legs. These symptoms were aggravated one month prior to presentation. The patient had no history of lumbar puncture or meningocele repairs. A neurological examination revealed paraparetic gait (muscle strength grade 4 in lower limbs). There was hypesthesia in the L1-5 and anesthesia bilaterally in the sacral dermatomes bilaterally. Her bilateral ankle and knee reflexes were diminished. She had been performed left nephrectomy due to hydronephrosis, two years earlier. Since then, she is doing self-catheterization for voiding.

Plain X-rays of the spine showed a vertebral body and pedicular erosion at T12, L1, L2, L3 and L4. Magnetic resonance (MR) imaging revealed a nonenhancing, sharply circumcised, intramedullary lesion at the conus medullaris and an intradural lesion at the lumbar region, which was hypointense on T1-weighted images, and hyperintense on T2-weighted images (Fig. 1). Computed tomography (CT) scan revealed a small calcified lesion at the conus medullaris (Fig. 2).

A T12-L4 laminectomy was performed. A fusiform enlargement of the dural sac was noted. At opening the dural mater at the lumbar spine, an avascular mass, lying within the intradural space, was seen. It contained a white, waxy material, which was easily removed by suction. The thin capsule was intimately adhered to the rootlets.
A 1-cm myelotome, centered over the lesion, was performed at the conus medullaris, and then another separating mass was seen, completely within the substance of the cord at the conus medullaris. It contained same material as was noted in the lumbar spine. We tried to remove the calcified capsule that had adhered intimately to the parenchyma, however, the adhesiolysis was unsuccessful. Histopathological examination of both the conus and cauda equinae lesions showed cholesterol clefts and strands of laminated keratin material. We found matured squamous cells and amorphous contents of a cyst has often been regarded as the method of choice.4,8,33. In fact, the potential regrowth of a partially excised congenital tumor must be recognized according to Collins rule.1 An intramedullary epidermoid cyst usually tightly adheres to the surrounding cord tissue. In this case, the spinal cord was damaged during the removal of the calcified capsule and the patient had a monoplegic state of the right leg.

Discussion

An epidermoid tumor is a benign congenital tumor, which develops during the third to fourth week of fetal life by the inclusion of cutaneous ectoderm into the neural ectoderm.10,24. Multiple spinal punctures and meningeal repairs have contributed to the reduced incidence of spinal epidermoid tumors.2,6. An intramedullary localization of an epidermoid tumor is extremely rare, and there has been no report of multiple epidermoid tumors in the spinal canal.

MR image may not be specific in the preoperative diagnosis, because there are other intramedullary spinal tumors that appear hypointense on T1-weighted images, and hyperintense on T2-weighted images, such as, ependymomas, astrocytomas and hemangiomas [40% of which are cystic].10. A calcified lesion on a CT scan could be helpful in differential diagnosis. The logical treatment of an intramedullary epidermoid is total surgical excision, if possible, to prevent a recurrence.4,5,7.

Tekkok, et al claimed that a microsurgical technique allowed adequate dissection and removal of the adherent capsular fragments without appreciable damage to the spinal cord.13. However, part of the tumor capsule adhering to the nervous tissue should be left in situ, because its removal might lead to greater neurologic deficits.4,5,7. Thus, the evacuation of the amorphous contents of a cyst has often been regarded as the method of choice.3,4,8. In fact, the potential regrowth of a partially excised congenital tumor must be recognized according to Collins rule.1 An intramedullary epidermoid cyst usually tightly adheres to the surrounding cord tissue. In this case, the spinal cord was damaged during the removal of the calcified capsule and the patient had a monoplegic state of the right leg.

Conclusion

In conclusion, before surgical intervention is performed, it is important to establish a surgical plan. Radical removal should be the method of choice. However, when the capsule is adherent to the neural structure, radical removal should be avoided, so as not to lead to neurologic deficit.

References


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