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Case Report

Surgical Findings of a Lumbar Mature Teratoma Accompanying the Preoperative Intracranial Dissemination of Fatty Droplets

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There are several mechanisms for the dissemination of lipid material from a mature teratoma into the subarachnoid space or ventricles, including iatrogenic or traumatic rupture, but spontaneous rupture of a mature teratoma is rare. We report the spontaneous rupture of a spinal mature teratoma into the subarachnoid space and ventricles. However, at surgery, there was no definite evidence of rupture into the perimedullary cerebrospinal fluid. We postulate that the central canal could be a migration pathway for ruptured material into the brain.

KEY WORDS : Mature teratoma.

INTRODUCTION

A spinal teratoma is a rare, benign, slow-growing tumor that arises from ectopic embryonic remains of the ectoderm and mesoderm in the spinal canal. There are several reported mechanisms for the dissemination of lipid material from a mature teratoma into the subarachnoid space or ventricles[3,15]. Nevertheless, spontaneous rupture of a mature teratoma is rare. We report a case of spontaneous rupture of a spinal mature teratoma into the subarachnoid space and ventricles. We postulate on how the cystic material from the teratoma could migrate into the brain based on a review of the literature.

CASE REPORT

A 44-year-old man presented with a history of voiding difficulty that started 10 years earlier and exacerbated 3 months before admission. He had no history of lumbar puncture or major operation. No gross abnormality was noted on his back. On neurological examination, he had voiding hesitancy, dysuria, and radiating pain and hyperesthesia in the left second sacral dermatome. The laboratory findings were non-specific. Magnetic resonance imaging (MRI) of the lumbar spine revealed a hyperintense intramedullary mass at the level of L3-L5 and the downward portion of the conus medullaris (Fig. 1). Computed tomography (CT) showed a non-enhancing low-density mass lesion at the same vertebral level, without spinal dysraphism

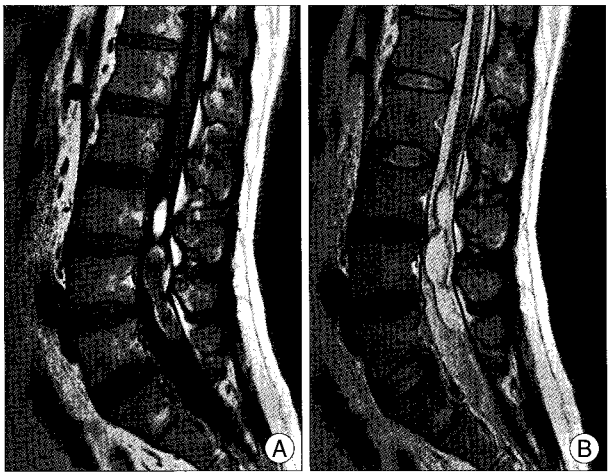

Fig. 1. Magnetic resonance imaging on admission. T1-weighted (A) and T2-weighted (B) sagittal images reveal a sausage-like abnormal signal lesion in the conus medullary area from the L2 to L4 level.

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(Fig. 2). Subsequently, CT and MRI were used to check for other central nervous system anomalies preoperatively. Multiple small, low-density lesions were scattered in the intraventricular and basal cisternal space (Fig. 3). Because these measured as 14 Hounsfield units, they were assumed to be fatty droplets.

Surgery was performed with the patient in the prone position under general anesthesia. After making a 13-cm midline skin incision, we exposed the lamina by dissecting the fascia and paraspinal muscle. After performing a bilateral total laminectomy from L2 to L5, we could find that there were no abnormal findings of the dura except slight bulging. On incising the dura, we found that the conus medullaris had a sausage-like appearance and its dorsal surface was lined with intact pia matter. There was no evidence of cyst rupture (Fig. 4A). Performing a careful midline myelotomy, we realized that the posterior wall of the cyst was encased by a -2-mm-thick normal conus medullaris (Fig. 4B). The cyst contained soft deep-ivory material with a creamy consistency, mixed with hairy tissue and subcutaneous appendages. The contents were removed without significant spillage into the cerebrospinal fluid (CSF).

Microscopic examination of the lumbar cyst revealed a squamous epithelium with keratin material, sebaceous glands, and mature fat (Fig. 5A). Eosinophilic fibrillary tissue consistent with a neural origin was scattered throughout the sample. Foci of mature enteric epithelium were found. These findings are consistent with a diagnosis of mature teratoma (Fig. 5B).

Postoperative MRI showed that nearly all of the mass had been removed (Fig. 6). Postoperatively, the patient's voiding difficulty improved gradually over several weeks.

DISCUSSION

Intraspinal mature teratomas constitute approximately 0.1 to 0.2% of all spinal tumors. Intraspinal teratomas are

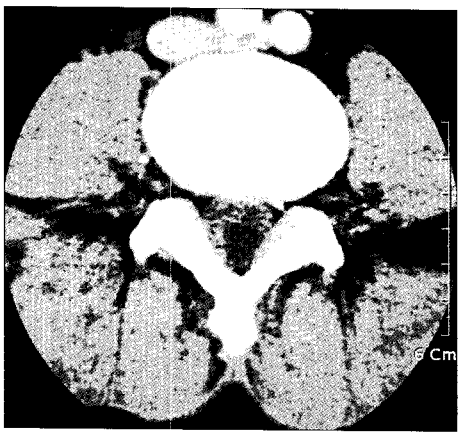

Fig. 2. Preoperative axial computed tomography showing the abnormal low-density lesion in the intramedullary area from L2 to L5.

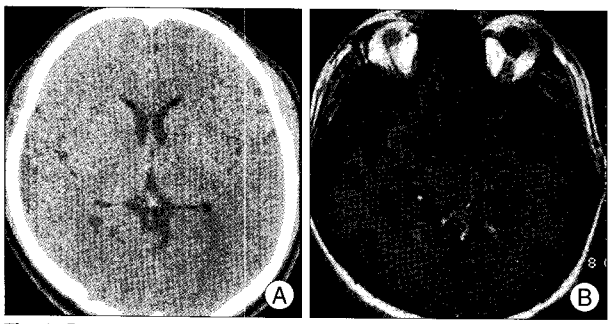

Fig. 3. Preoperative computed tomography and m magnetic resonance imaging showing multiple fat droplets in the basal cistern and lateral ventricle.

Fig. 4. A : On operative finding, there is no evidence of rupture of mature teratoma. B : During midline myelotomy, the teratoma is shown to be encased with intact pia and normal neural tissue.

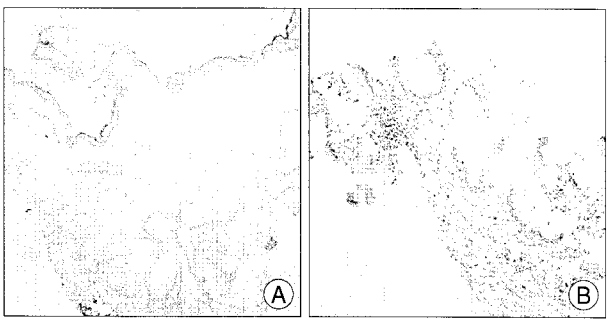

Fig. 5. A : Microscopic examination reveals a stratified squamous epithelium with keratin material, sebaceous glands, and mature fat. B : An enteric type epithelium and chronic inflammation are also noted. (H&E, ×40 and ×100).

most often located posteriorly in the cervical or lower thoracic and upper lumbar regions[11]. The pluripotent embryonic caudal mesenchyme gives rise to teratomas and other congenital tumors via the dysfunction of several factors, which probably involve gene function and cellular inductive interactions[9]. Typically, there is no communication between the cyst and subarachnoid space.

Mature teratomas are slight more predominant in males. They may be related to bony malformations, myelomeningoceles[13], hypertrichosis, or a dermal sinus tract[12]. The lumbosacral region is the most common site (60%), involving the cauda equina and conus medullaris, followed by the upper thoracic (10%) and cervical (5%) regions[7]. Our patient had no gross abnormality of the skin or bone in his lumbar area.

Although a mature teratoma may start to develop during the embryonic period, symptoms may not occur until adulthood due to their slow growth[10]. The symptoms and signs secondary to a space-occupying lesion are location-dependent and are due to the irritative effect on or compression of the adjacent structures. When accompanied by a tethered cord, particularly with small lesions, neurological symptoms can be elicited without a mass effect. In our case, the patient's neurological deficit was due to the mass effect of the mature teratoma and a tethered cord in the lumbar intramedullary region.

The relatively high signal from fat during MRIs, especially on T1-weighted images, makes the identification of lipid droplets easy, particularly within the cerebral sulci, fissures, perimedullary subarachnoid space, and central canal of the spinal cord[16].

A mature teratoma may have two distinct regions: a lipid and a more solid or fluid one[7], as in our case, with fat tissue in the peripheral portion of the tumor. In addition, especially with the leakage of lipid material, the use of intravenous contrast medium facilitates the diagnosis of meningeal inflammation.

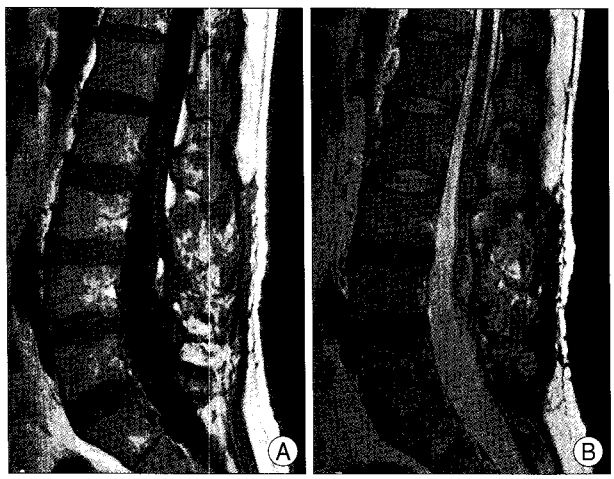

Fig. 6. Postoperative magnetic resonance imaging. T1-weighted (A) and T2-weighted (B) sagittal images reveals no abnormal signals in the conus medullary area from the L2 to L4 level.

Once the cyst ruptures, acute symptoms related to chemical or aseptic meningitis[13], headache, or seizures may develop due to dissemination of the lipid droplets in the CSF. Lumbar arachnoiditis may develop as a result of the leakage of fat and proteinaceous material into the subarachnoid space. The highly irritative lipid content of a mature teratoma may cause a severe inflammatory response, although the spread of fat into the CSF may also be silent clinically[4]. Our patient had no complications relating rupture.

A mature teratoma is composed of fully differentiated, "adult-type" ectodermal, mesodermal, and endodermal elements. The more common ectodermal components include skin, brain, and choroid plexus. Mesodermal representatives include cartilage, bone, fat, and muscle. Cysts lined by a respiratory or enteric type epithelium are the usual endodermal components. Epidermoid and dermoid tumors are made of epithelium, connective tissue, and skin appendages; neural, mesenchymal, or glandular tissues are not identified[1]. The pathological findings in our case included enteric type epithelium, mature fat, skin, sebaceous glands, and neural tissue. As it contained elements derived from all three germ cell layers without immature elements, a diagnosis of mature teratoma was made.

After a mature teratoma ruptures, lipid droplets float in the CSF and are conveyed passively by CSF movement. They can spread throughout the subarachnoid space and ventricular system. Scearce et al.[14] reported that fat droplets reach the ventricles from the perimedullary subarachnoid space via retrograde flow through the foramina of Luschka and Magendie. To the best of our knowledge, few cases of ruptureed mature teratoma have been reported[2,5,6,8,14]. In our case, the mature teratoma appeared to have ruptured spontaneously with the intracranial dissemination of fatty droplets. At surgery, however, no site of rupture was identified. We wondered how it was possible for small fatty droplets to exist in the brain without evidence of rupture into the perimedullary CSF. We thought that the fatty droplets either migrated via the central canal or were incidental findings of a congenital lesion. However, the small fatty droplets were found diffusely in the subarachnoid space and ventricular system, and were unremarkable in terms of size or the typical locations of such droplets (the third ventricle, intraventricular area, or suprasellar region). Therefore, it is possible that the central canal can serve as a migration pathway for ruptured material into the brain.

Moreover, we know that a ruptured teratoma usually causes severe symptoms. However, our patient was clinically asymptomatic in terms of the migration of ruptured material into the brain, and it was not necessary to treat this.

CONCLUSION

We report a patient who had a spinal mature teratoma with coexisting intradural fatty droplets in the intracranial subarachnoid space and ventricles preoperatively. The patient showed only insidious clinical course. At surgery, there was no definite evidence of rupture into the perimedullary CSF. We suggest that the central canal might be another slow migration pathway for the content of tumor material into the brain.

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