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Case Report

Epidermoid cysts associated with thoracic meningocele

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Summary

Intraspinal tumours of cutaneous origin associated with various spinal dysraphisms have been well documented in the literature. However, the metachronous development of intra- and extra-medullary tumours in conjunction with dorsal meningocele is rare. The authors report a patient with a thoracic dorsal meningocele and congenital intradural extramedullary epidermoid tumour. The patient developed an intramedullary epidermoid growth 12 years later. Subtotal resection of the tumour predisposed to a later recurrence.

Meningocele is not always an isolated clinical entity but the concurrent occult lesions are usually veiled by the more conspicuous surface anomaly. Thorough magnetic resonance imaging of the whole neural axis helps to identify associated pathologies. Delicate intradural exploration by a microsurgical approach is necessary to achieve appropriate treatment.

Keywords: Epidermoid; intramedullary tumour; meningocele; myelomeningocele; spinal dysraphism.

Introduction

A posterior spinal meningocele used to be considered as a simple and benign lesion. With the popularity of magnetic resonance imaging (MRI) and the advance of microsurgical techniques, more and more concurrent dysraphic pathologies such as epidermoids, dermoids, lipomas or dermal sinus tracts have been found to complicate this simple midline closure defect [1, 2, 5, 7, 10]. The wide spectrum of combined dysraphic anomalies continues to challenge not only our understanding of the normal and aberrant neural embryogenesis but also the strategies of management. The authors report a 16-year-old girl who developed an intramedullary epidermoid after removal of congenital intradural extramedullary epidermoids and repair of a thoracic meningocele as a neonate. The report illustrates an uncommon combination of dysraphic lesions associated with a simple spinal dysraphism and to our knowledge this metachronous development of intra- and extra-medullary epidermoids associated with a meningocele has not been reported previously.

Clinical report

The patient was initially admitted to hospital with a four-week history of upper back pain, incontinence and progressive lower limbs weakness. Tracing back her past history, she was a product of a full-term pregnancy and normal vaginal delivery but had undergone repair of a posterior thoracic meningocele five days after her birth. Pre-operative computed tomography (CT) indicated a cerebrospinal fluid containing cyst bulging through a spina bifida at T3–T5 vertebrae without other visible abnormalities. During intradural exploration, multiple small pearly nodules within the meningocele with some

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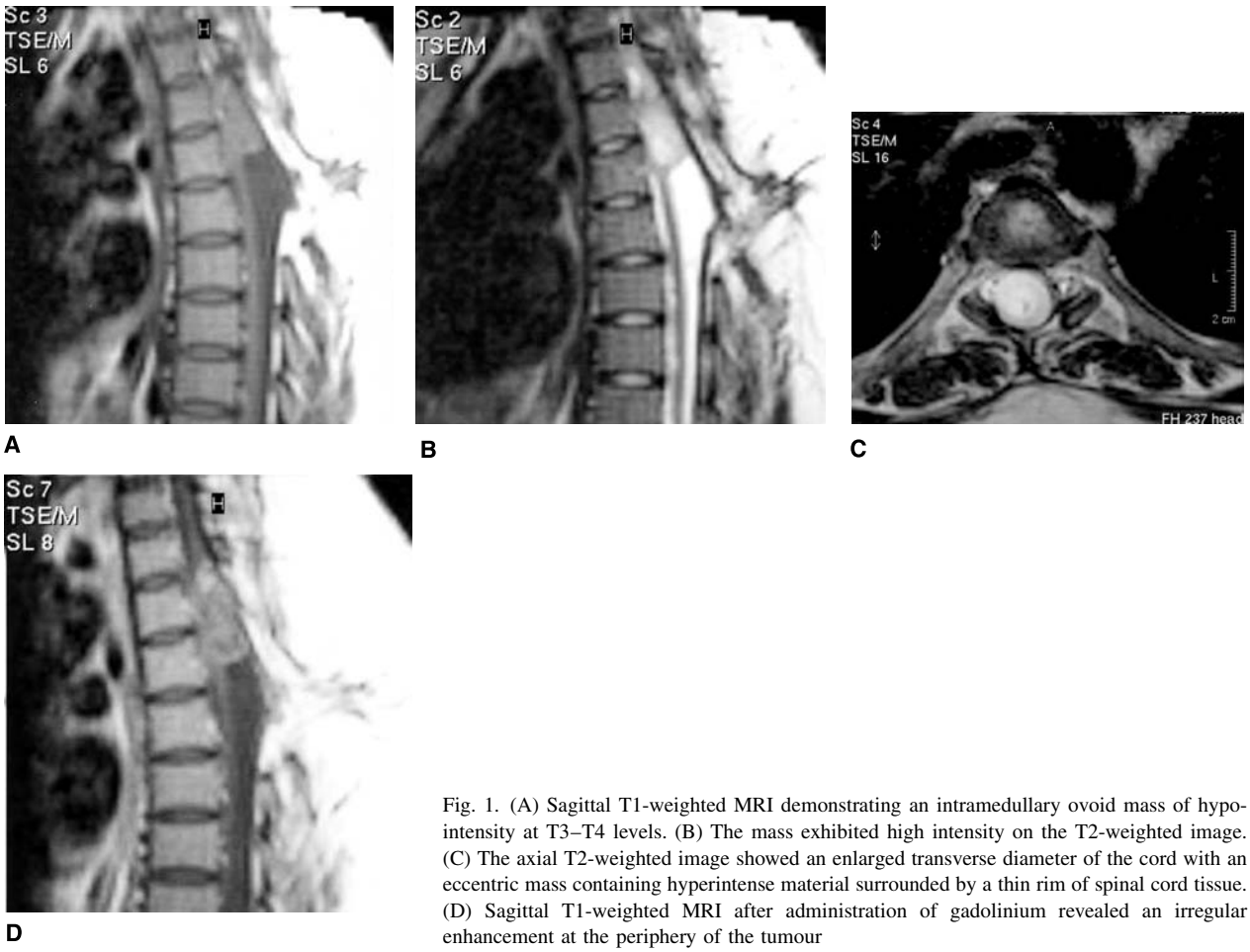


Fig. 1. (A) Sagittal T1-weighted MRI demonstrating an intramedullary ovoid mass of hypointensity at T3–T4 levels. (B) The mass exhibited high intensity on the T2-weighted image. (C) The axial T2-weighted image showed an enlarged transverse diameter of the cord with an eccentric mass containing hyperintense material surrounded by a thin rim of spinal cord tissue. (D) Sagittal T1-weighted MRI after administration of gadolinium revealed an irregular enhancement at the periphery of the tumour

of them attaching to the dorsal spinal cord surface were noted. These lesions were totally removed without difficulty followed by ligation of the meningocele sac. Microscopic examination of these nodules demonstrated the appearance of an epidermoid. The post-operative course was uneventful and the subsequent developmental milestones of the patient were normal. However, she developed an unsteady gait and occasional urinary incontinence at the age of 8 years. MRI of the spine at that time ruled out recurrence of the epidermoid or a tethered spinal cord. CT of the head revealed hydrocephalus which necessitated a ventriculo-peritoneal shunt. The patient recovered from the symptoms after shunting although a mild urinary disturbance remained. At age 12 years, she developed incontinence and progressive paraparesis. A CT of the head showed normal size ventricles, but the MRI demonstrated an intraspinal intramedullary mass at the site of previous meningocele repair. Near total removal of the tumour was done but some parts of the capsule were left in place for fear of injury to the spinal parenchyma. The weakness of the lower limbs recovered progressively and she also regained continence. Four years later, she was admitted to hospital again with a recurrence of similar symptoms.

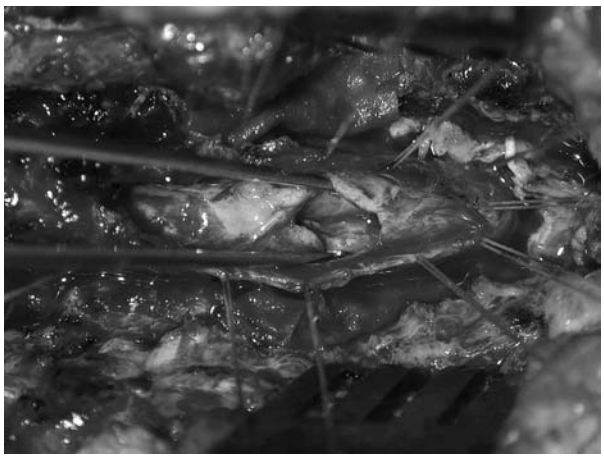
Neurological examination demonstrated both a paraparesis (grade 3/5) as well as paraesthesiae below the level of the nipples. Decreased anal tone and deep tendon reflexes of the lower limbs were also detected.

The MRI of the thoracic spine revealed a bony defect at T3–T5 levels with sequestrated cerebrospinal fluid. At the level of T3–T4, an ovoid intramedullary tumour was identified. The mass was hypointense on T1-weighted images but hyperintense on T2-weighted images (Fig. 1). The periphery of the tumour enhanced after administration of gadolinium on the T1-weighted sequence.

Operative findings

At operation, an ovoid expansion of the spinal cord was identified with some pearly white material rupturing to the subarachnoid space (Fig. 2). After a midline myelotomy, a mass containing avascular, soft and caseous ma-

A



B

Fig. 2. Intraoperative photographs demonstrating the thoracic spinal cord before (A) and (B) after removal of the intramedullary epidermoid. Note that some epidermoid tissue has ruptured through the spinal cord surface

terial was found immediately underneath the cord surface and was removed gently. The surrounding capsule was carefully stripped from the interior surface of the thoracic cord. Microscopic examination of the specimen demonstrated keratin and degenerating squamous epithelium compatible with an epidermoid.

Postoperative course

The patient developed mild fever and meningeal signs three days following surgery. Aseptic meningitis was suspected but soon came under control. She received a short course of steroids which was then tapered progressively. The patient began to recover lower limb strength soon and was able to walk without support two months later. She also regained control over the bladder. During follow-up at outpatient clinic, no evidence of tumour recurrence has been evident 4 years after surgery.

Discussion

Intraspinal epidermoid cysts could be of acquired or congenital origin. Incidental introduction of epidermal elements into spinal canal by lumbar puncture is the most common aetiology for the acquired tumours. Another rare iatrogenic cause is inadequate excision of epithelium around the neural placode at the time of repair of a myelomeningocele [8]. The pathogenesis of congenital intraspinal epidermoids is considered to arise from entrapment of epithelial rests within the neural axis at the stage of closure of the neural tube and separation of neural and cutaneous ectoderm during the third or fourth weeks of embryonic life.

Epidermoids, as well as other inclusions associated with midline closure defects have been well recognised [3]. Intraspinal inclusion cysts associated with myelomeningocele are more common. Nelson *et al.* found dermoid inclusion cysts in 16% of the patients in whom surgery was performed to release a tethered placode [4]. In Reigel's series of 102 patients with tethered spinal cord, most were subsequent to repair of a myelomeningocele or meningocele, 13 had epidermoid tumours and 2 had dermoid tumours [6]. Inadvertent inclusion of epithelial tissue at the repair of the defect has been considered to be the possible cause of post-operative dermoid or epidermoid lesions but sometimes the inclusion cysts might have existed before repair of the myelomeningocele and unnoticed during surgery. Storrs examined fresh myelomeningoceles and specimens from tethered cord release and concluded that hamartomatous lesions are a common feature of the myelodysplastic sequences [9].

Although one dysraphic state is occasionally associated with another, meningocele, an infrequent form of dysraphism is often clinically benign and thought to have less concurrent spinal or cranial anomalies. However, more and more patients with dorsal meningoceles have been reported to be accompanied with other dysraphic lesions [1, 2, 5, 7, 10]. In a series of 22 newly diagnosed meningocele patients reported by Ersahin, 90% were associated with other occult lesions including epidermoids, dermoids, fibrous bands, tight filum and split cord malformation [2].

With our knowledge of human embryology evolving, the underlying mechanisms of spinal dysraphism are better understood. Several distinct theories have been proposed to explain the developmental error accounting for a specific phenotype of dysraphism and its related variants. However, the more complex combinations of various dysraphic entities continue to challenge the existing hypothesis. Coincidental independent developmental

errors might be possible; nevertheless, it is always tempting to hypothesise a common embryological origin for the concurrent events. This unusual example we present can be explained by the theory of limited dorsal myeloschisis in which the final fusion of the neural plate is almost complete but for a small limited area in the midline where the original attachment between neuroectoderm and cutaneous ectoderm persists [5]. As the neural tube is displaced ventrally by the proliferating mesoderm, the ectodermal attachment which contributes to the adhesion band in some cases regresses but the remnants might predispose to the later development of extra- and intra-medullary epidermoids. In our patient, no evidence exists that any previous operative procedures was the cause of the delayed development of the intramedullary tumour. Simple removal of an epidermoid on the cord surface seems unlikely to introduce epidermoid tissue into the spinal cord parenchyma. The recurrence of the epidermoid four years later should be related to the residual capsule left within the thoracic cord from previous incomplete excision.

MRI is an effective tool for the diagnosis of intraspinal epidermoids. The lesion is usually isointense or hypointense on T1-weighted images and hyperintense on T2-weighted images. After infusion of gadolinium, an irregular enhanced rim can be identified surrounding the heterogeneous central cavity. The enhanced rim is considered as the wall composed of tumour cells, and the central cavity is a mixture of keratin and cholesterin.

The management of intraspinal epidermoids is surgical, and complete removal is the goal of treatment. Because an epidermoid is well encapsulated, it facilitates dissection and removal of the whole tumour without causing appreciable neural damage. Although the intimate adherence between the capsule and spinal cord sometimes makes the aim impossible and carries risk of cord injury, with the help of microsurgical techniques and neurophysiological recording, complete removal of the capsule will decrease the chance of recurrence. Careful protection to avoid dissemination of the cystic contents into the subarachnoid space is also important to prevent post-operative chemical meningitis.

Conclusion

A meningocele is not always an isolated lesion and may be associated with other occult abnormalities at the same or different spinal levels. Apart from that, patients with spinal dysraphism also carry a higher risk of developing hydrocephalus and tethered cord syndrome which may present with similar clinical manifestations as intraspinal tumours. Early radiological evaluation of the whole spine is necessary. Intraspinal tumours should always be kept in mind in the differential diagnosis once patients with spinal dysraphism develop later neurological deterioration and treatment should be initiated as soon as possible before irreversible spinal cord damage is caused by the space occupying lesion.

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Comments

Intramedullary spinal epidermoids are seen under two circumstances: they may be found in isolation (without evidence of spinal dysraphism) or they may be in continuity with a dermal sinus. There are several reports dealing with these two presentations of intramedullary epidermoids.

Epidermoid tumours have also been described in association with open spinal dysraphism, namely with meningoceles or myelomeningoceles. In this case, they may occur as the result of the inadvertent entrapment of skin elements at the time of myelomeningocele closure. In a recent review from our Hospital we found 3 instances of intradural epidermoids in 11 cases of postmyelomeningocele-repair tethered cord accounting for an incidence of 27% for this occurrence [1]. Interestingly, Mazzola *et al.* have documented inclusion dermoid cysts in cases of intrauterine myelomeningocele repair, a fact that they attribute to the accidental implant of skin or to the use of duraplasty with acelular human dermis [2]. More rarely myelomeningoceles and inclusion cysts of epidermal origin are coexistent lesions, that is, the epidermoid cyst is unrelated to myelomeningocele repair and probably originates at the same time as myelomeningocele develops.

The occurrence of an intramedullary epidermoid in the thoracic cord associated with open spinal dysraphism, as in the case reported here, really represents an exceptional event. As the authors state, this infrequent association can be part of the spectrum of combined anomalies that can be found in congenital malformations, as occurs in spinal

dysraphism. Also, there exists the possibility of one of the epidermoid cyst's rests of the previous operation growing into the substance of the spinal cord and attaining a large size whose origin would be difficult to ascertain. In other words, the remnants of the previous epidermoid cyst capsule could have grown within the spinal cord excavating and eroding the spinal cord parenchyma. An important issue for the management of this late complication of meningocele repair is the awareness of that the appearance of new symptoms and signs added to a pre-existent neurological deficit in patients with open spinal dysraphism does not always represent the natural evolution of the disease.

In our view, this case is worthy of report as it contributes to the knowledge of the possibility of an epidermoid growing within the spinal cord as a late complication in the repair of spinal dysraphism.

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