CASE REPORT

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Coexistence of Subcutaneous Dermoid Cyst and Lipomyelomeningocele

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Although lipomyelomeningocele and dermoid cyst are formed during a similar embryologic developmental stage of 'neural tube formation', they are caused by entirely different embryologic mechanisms. We encountered a case, which has not been previously reported, that had a lumbar subcutaneous dermoid cyst associated with lipomyelomeningocele. A 52-year-old man presented with a slowly growing lumbar mass of 3 years duration, which had been present since birth. Lumbosacral magnetic resonance imaging(MRI) showed lipomyelomeningocele at the L3, 4 and 5 levels and a subcutaneous cystic mass of high signal intensity on the T2 weighted image and iso or low signal intensity on the T1 weighted image. He underwent total resection of the lumbar subcutaneous mass. Intraoperative findings and histological examination were consistent with the preoperative diagnosis of a 'dermoid cyst.' We demonstrate that the formation of a dermoid cyst might coexist with lipomyelomeningocele during the embryologic developmental stage.

KEY WORDS : Coexistence · Dermoid cyst · Lipomyelomeningocele · Neural tube formation.

Introduction

The term spinal dysraphism refers to a group of congenital malformations of the back and spine[4,11]. Spinal dysraphisms may be categorized clinically into two subsets. In open spinal dysraphisms, the placode (non-neurulated neural tissue) is exposed to the environment. These disorders which include myelomeningocele, myeloschisis, hemimyelomeningocele, and hemimyelocele are always associated with a Chiari II malformation. Occult spinal dysraphisms are covered by intact skin, although cutaneous stigmata usually indicate their presence. Two subsets of occult spinal dysraphism may be identified based on whether a subcutaneous mass is present in the low back. Occult spinal dysraphisms with mass comprise lipomyeloschisis, lipomyelomeningocele, meningocele, and myelocystocele. Occult spinal dysraphisms without mass comprise complex dysraphic states (ranging from complete dorsal enteric fistula to neurenteric cysts, split cord malformations, dermal sinuses, caudal regression, and spinal segmental dysgenesis), bony spina bifida, tight filum terminale, filar and intradural lipomas, and persistent terminal ventricle[12].

The term lipomyelomeningocele(LMMC) is used by many neurosurgeons to address all occult spinal dysraphisms in which the spinal cord is tethered by a lipoma[1,4]. Lipomyelomeningocele is the most common form of occult spinal dysraphism and causes progressive impairment of, the lower extremities or bladder

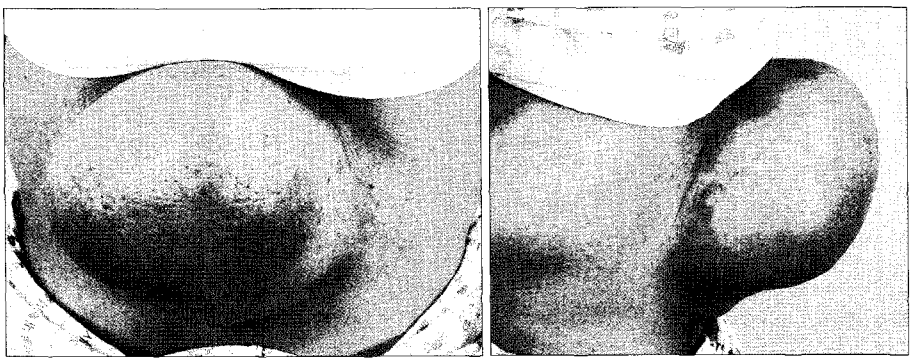

Fig. 1. A huge soft mass, measuring 20×15×10cm, is observed in the lumbar area. No evidence of the existence of telangiectasis, hypertrichosis, skin dimple or sinus tract.

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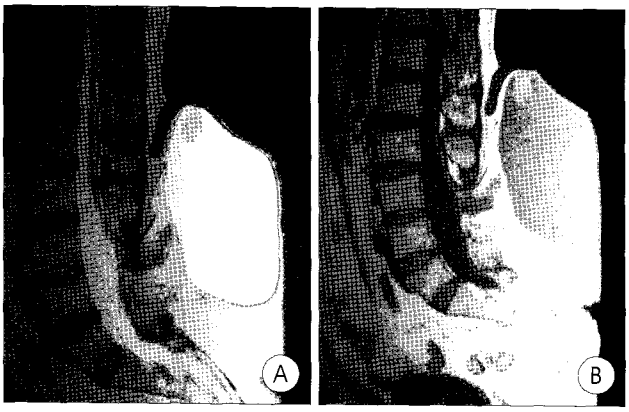

Fig. 2. Magnetic resonance image showing lipomyelomeningocele and high signal intensity on the T2 weighted image (A). The mass is iso or low signal intensity on the T1 weighted image. There is no evidence of a connection between the mass and any spinal canal component (B).

Fig. 3. The mass is composed of a densely fibrous connective tissue capsule and contained heterogenous materials including hair and a greasy gray colored debris.

Fig. 4. The wall of mass is composed of epithelial cells (H&E, ×200) (A). The mass includes a sebaceous gland, which is a kind of dermal appendages (H&E, ×100) (B).

or bowel function through spinal cord tethering and compression. Dermoid cyst is defined as a benign cystic lesion derived from ectopic inclusions of epithelial cells or dermal appendages during neural tube closure[5]. Spinal epidermoid or dermoid manifests as two forms : one is a part of the congenital dermal sinus tract, which belongs to occult spinal dysraphism, and the other is a subcutaneous dermoid cyst, which does not involve the dermal sinus tract.

We encountered a patient having a lumbar subcutaneous dermoid cyst associated with lipomyelomeningocele without dermal sinus tract involvement for the first time.

Case Report

A 52-year-old man presented with a slowly growing lumbar mass of 3 years duration, though the mass had been present since birth. The inherent mass hindered his driving a taxi, and lying supine. His symptoms included gait disturbance, urinary difficulty, a tingling sensation, and a numbness of both lower extremities over a period of several decades. About 40 years previously a nail penetrated his foot and resulted in a right below knee amputation operation because of a complicated infection. In addition 14 years previously he had undergone an internal fixation operation of left ankle due to deformity. Both ankle & foot motion had since been impossible. A neurologic examination showed the presence of saddle anesthesia and both below knee paresthesia and hypesthesia without any definite motor weakness. A physical examination revealed a huge soft mass, measuring 20×15×10cm, not associated with telangiectasia or hypertrichosis (Fig. 1). Lumbosacral MRI showed L3, 4 and 5 level posterior element defects, connection between the spinal cord and the subcutaneous soft tissue through these defects and spinal cord tethering at this level. In addition, the MRI showed a high signal intensity on T2 weighted imaging and iso or low signal intensity on T1 weighted imaging, corresponding to the subcutaneous cystic mass (Fig. 2). There was no evidence of connection between the mass and any other components in the spinal canal. The preoperative diagnosis was 'lipomyelomeningocele and

an associated dermoid cyst or myelocystocele'.

The patient underwent total resection of the lumbar mass without untethering. Intraoperative findings showed that the mass was composed of a densely fibrous connective tissue capsule and heterogenous cysic materials including hair and greasy gray colored debris (Fig. 3). Moreover, it was connected without any other components in the spinal cord. A histologic examination confirmed this mass as a dermoid cyst, as had been expected preoperatively (Fig. 4).

Discussion

Spinal cord development occurs through three consecutive periods of gastrulation (weeks 2~3), primary neurulation (weeks 3~4), and secondary neurulation (weeks 5~6)[10]. Gastrulation (weeks 2~3) is characterized by the conversion of the embryonic disk from a bilaminar to a trilaminar arrangement and the establishment of a notochord. Primary neurulation (weeks 3~4) produces the uppermost nine tenths of the spinal cord, whereas secondary neurulation and retrogressive differentiation (weeks 5~6) result in the formation of the conus tip and filum terminale. Defects in these early embryonic stages produce spinal dysraphisms, which are characterized by anom-alous differentiation and fusion of dorsal midline structures[12].

Dermoid cysts are defined as benign cystic lesions derived from ectopic inclusions of epithelial cells or dermal appendages during neural tube closure[5]. They reflect a problem at the gastrulation stage of development, with primary disruption of tissues derived from surface ectoderm. Of these lumbosacral dermoid cysts can be classified into two forms : one includes those connected by a dermal sinus with the skin, and the other lack a dermal sinus[6]. The frequency ratio of the former to the latter is 1 to 4[6]. The former may arise from expansion of the dermal sinus, which develops from incomplete separation of the neural ectoderm from the cutaneous ectoderm[11]. The latter develop from "cell rest", which may be the result of cutaneous tissue sequestration during neural tube formation[2,11].

Congenital lumbosacral lipoma, used by more inclusive meaning than lipomyelomeningocele regardless of tethering spinal cord, forms because of the separation of primitive ectoderm into neuroectoderm, neural crest, and cutaneous ectoderm and the improper simultaneous fusion posteriorly of the resulting neural plate edges and of the overlying edges of cutaneous ectoderm[14]. The paraxial mesenchyme can migrate through the gap of between the cutaneous ectoderm and the neural tube. This mesenchyme becomes primarily fat under the inductive influence of the inner ependymal surface of the placode[8].

In the present case the patient had both a subcutaneous dermoid cyst and lipoma tethering the spinal cord simultaneously between intact skin and spinal structures. Similar embryological developmental abnormalities can cause and generate both dermoid cyst and lipomyelomeningocele. However, no case report has been issued concerning the coexistence of a subcutaneous dermoid cyst without an associated dermal sinus tract to a lipomyelomeningocele.

Three possible mechanisms of the coexistence should be considered. First, the simultaneous formation of both a solitary subcutaneous dermoid cyst without any dermal sinus tract and lipomyelomeningocele could occur during primary neurulation. Second, the dermoid cyst could persist within the lipoma of a lipomyelomeningocele. During migration of paraxial mesenchymal tissue, a primarily formed lipoma can include tissue of mixed embryonic origin, such as bone, cartilage, smooth and striated muscle, collagen, renal epithelium, and gastric mucosa[1,3,13]. The possibility that the lipoma capsule wrapping the dermoid cyst thinned over several decades should be considered. And finally, as we could not discover the dermal sinus tract associated with the dermoid cyst it is likely that it had degenerated and become atrophic over several decades. The last hypothesis may be less possible, since no report has mentioned a disappearing or disappeared dermal sinus tract, and this patient had no history of recurrent meningitis. All the three hypotheses are possible, unfortunately we are unable to prove which is true.

There is some doubt as to whether untethering should be undertaken in such a patient or not. His neurologic deteriorations, i.e., lower extremity hypesthesia and paresthesia, and urinary difficulty had been stationary for decades. Motor power improvement of his lower extremities was meaningless because his right leg had been amputated below the knee and his left ankle joint had no motion. Accordingly an intra-spinal procedure including untethering would have provided little benefit and substantially more risk to this patient.

Conclusion

Lipomyelomeningocele and dermoid cyst are caused by different mechanisms from an embryologic point of view. The former is a 'separation failure and mesodermal defect', whereas the latter is an 'ectopic inclusion'.

However, we supposed the possibility of the coexistence of these embryologic developmental abnormalities, and here for the first time, encountered a case with a lumbar subcutaneous dermoid cyst associated with lipomyelomeningocele without any dermal sinus tract.

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