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Meningocele with Cervical Dermoid Sinus Tract Presenting with Congenital Mirror Movement and Recurrent Meningitis

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Dermoid sinuses and meningoceles are seldom encountered in the cervical region. Besides, to the best of our knowledge, the coexistence of these types of congenital abnormalities with recurrent meningitis, as well as with mirror movement, has never been reported before. A 14-year-old female with the diagnosis of recurrent meningitis was referred to our clinic from the Department of Infectious Diseases. She had a cervical meningocele mass that was leaking cerebro-spinal fluid (CSF) and an associated mirror movement symptom. Spina bifida, dermoid sinus and meningocele lesions were demonstrated at the C2 level on computed tomography (CT) and magnetic resonance imaging (MRI). She underwent an operation to remove the sinus tract together with the sac, and at the same time the tethered cord between the sac base and the distal end of the spinal cord was detached. The diagnosis of dermoid sinus and meningocele was confirmed histopathologically. These kinds of congenital pathologies in the cervical region may also predispose the patient to other diseases or symptoms. Herein, a case of meningocele associated with cervical dermoid sinus tract which presented with recurrent meningitis and a rare manifestation of mirror movement is discussed. Neurosurgeons should consider the possible coexistence of mirror movement and recurrent meningitis in the treatment of these types of congenital abnormalities.

Key Words: Dermal sinus, meningocele, mirror movement, recurrent meningitis, spinal dysraphism

INTRODUCTION

Dermoid sinuses are epithelial tissue-covered tracts extending from neural structures to the skin. They are congenital lesions that occur in embryonic life due to defective separation of the ectoderm into skin and neural components and are encountered in 1/1500th of normal births.[1] Dermoid sinuses are usually found in the lumbosacral region, whereas its occurrence at a cervical location represents only about 1.5% of all cases.[2-4] In our case, a meningocele sac found at the C2-3 level, which is a rare occurrence, was associated with a dermoid sinus and presented with mirror movement and recurrent meningitis. The combination of these signs and symptoms is very rare and is presented here.

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CASE REPORT

A 14-year-old girl was referred to our clinic with the prediagnosis of recurrent meningitis. On the day of admission, the presence of a 2 × 2 × 2 cm sac in the cervical region was noted, which protruded from the skin with the partial thinning of the overlying skin causing CSF leak (Fig. 1). It had previously been suggested to her that she needed to be operated on because of the cervical lesion, however her parents refused this intervention, which ultimately led her suffering a meningitis attack 5 years ago. Her deep tendon responses were found to be hyperactive on examination. She exhibited a marked involuntary mirror movement on the opposite side, which occurred whenever she was told to move her hand or one finger on one side. This mirror movement reportedly existed since childhood. Bone window CT scans indicated the presence of a fusion abnor-

mality of the spinal process of the C2 vertebral bone (Fig. 2). On T1-weighted axial and sagittal MRI, the spinal cord was found to be tethered to the backside by a tight fibrous band at the C2-3 level, and a hydromyelia cavitation located proximal to the neck of the sac was found to extend all the way up through the tract, culminating in a meningocele mass within the sac (Fig. 3 and 4). Cranial CT and MRI were otherwise unremarkable.

The patient underwent an operation in the prone position. An elliptical skin incision was made around the lesion and extended up along the vertical axis, in order to remove the lesion and for the sake of safe skin closure. The lesion was dissected from the skin and the subcutaneous tissue. The paravertebral muscles were decollated carefully from the laminae and spinal processes. The C2 spinal process abnormality at the midline was recognized, when the sinus tract was entirely exposed. The defect at the C2 level was widened by laminectomy, in order to expose the tract safely. The tract was followed along the sides down to the subarachnoid space. In this region, the walls were found to be thinned and the spinal cord was stretched backwards by a tethering fibrous band extending from the caudal part to the sac base. During the course of the operation, this band and the other arachnoidal adhesions were removed. The lateral parts of the tract were not amenable to being advanced to the midline, and the tract was therefore closed, mainly by

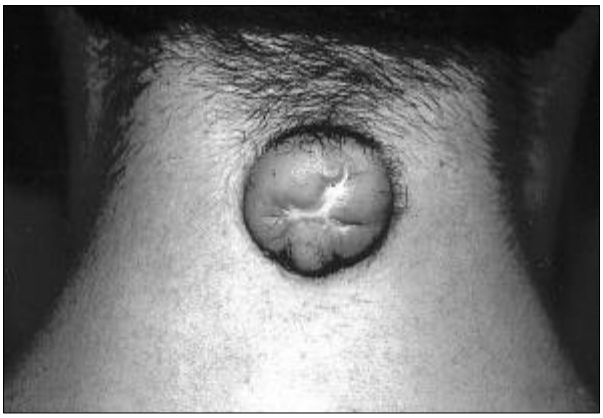

Fig. 1. Cervical meningocele is demonstrated.

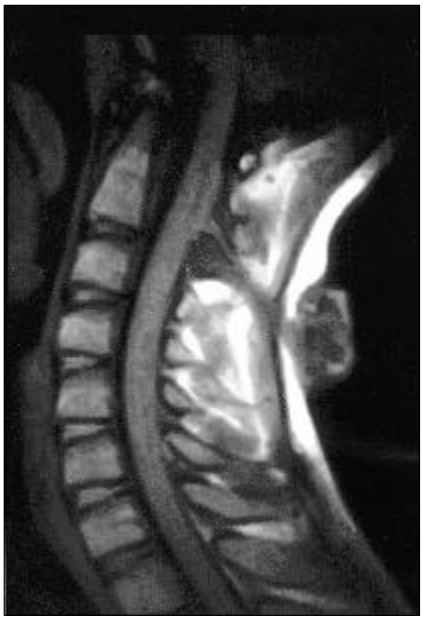

Fig. 3. Dermoid sinus, tethering cord, and meningocele sac are shown on T1-weighted MR image.

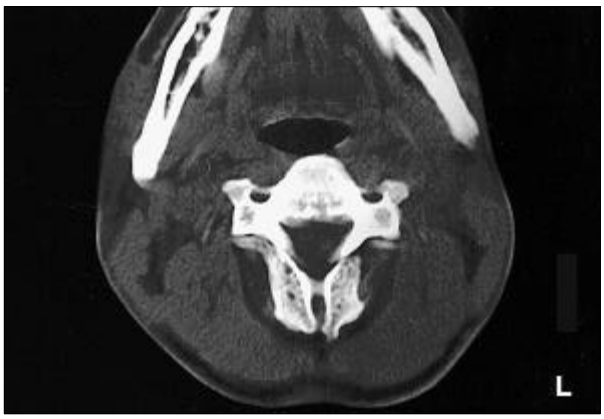

Fig. 2. Fusion abnormality of the posterior elements of C2 vertebra is seen on bone window-CT scan.

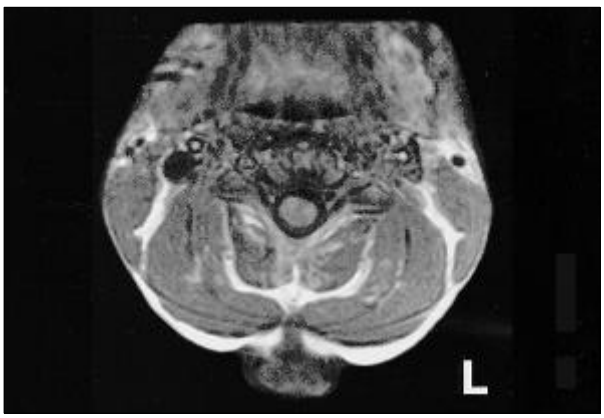

Fig. 4. Dermoid sinus is delineated on T1-weighted axial MR image.

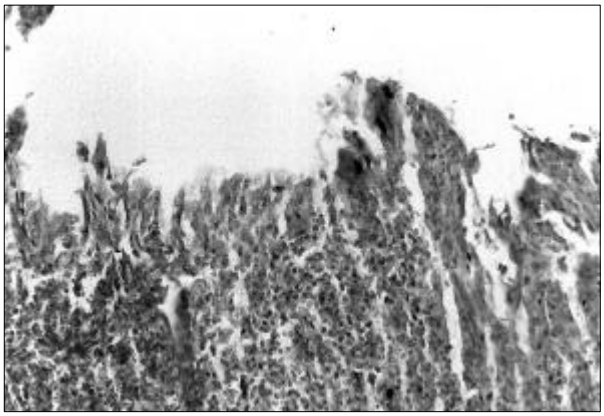

Fig. 5. Dermoid sinus tract lined with squamous epithelial cells and the surrounding profound lymphocyte infiltration. (HE ×40).

means of a fascial graft, in order to protect the intradural compartment. Arachnoidal elements and fibroadipous-like tissue were identified upon histopathological examination of the sac. A typical dermoid sinus lined on the inside with squamous epithelial tissue was found upon microscopic investigation of the tract (Fig. 5). There were no postoperative complications. However, the mirror movement persisted during the postoperative period.

DISCUSSION

The embryogenesis of cervical dysraphic lesions can be explained by incomplete fusion of the neural tube at the lesion level.[4-7] The designation, "spinal dermal sinus", was first introduced by Walker and Bucy[8] and this disorder is considered to be in the dysraphism category.

Dermoid sinuses and tumors can involve the cranium, from the nasion to the occipital region, and from the spinal canal down to the sacrum. In the case where the spinal canal is affected, these sinuses and tumors have a predilection for the lumbosacral region, whereas they are seldom encountered in the cervical region. Schiffer[4] reported on a series of 245 cases, of which 4 cases (1.6%) presented with cervical involvement. Dystrophic lesions such as diastematomyelia, diplomyelia, epidermoid cyst and hamartoma are rarely located in the cervical region.[4,6]

The presence of a small sinus ostium in cases of dermoid sinus may be recognized on physical examination, but usually cannot be diagnosed until the time the child contracts meningitis. In most of the cases in which there is a small sinus ostium, the neurological examination is unremarkable. MRI is superior to contrast CT, since it is more sensitive and noninvasive in the identification and demonstration of the dermal sinus and the associated abnormalities.[7]

Abnormalities which coexist with dermal sinus tracts, such as meningocele, myelocystocele and myelomeningocele, may also be associated with various kinds of skin lesions. Also, in some situations such as in spina bifida and Klippel-Feil Syndrome, the lesion may be associated with bone abnormalities, and can propagate into the spinal canal and may cause some pathologies, such as tethered cord, filum terminale at the dura/cord level.[4,6,9,10] In our case, the tract was found to end with a meningocele sac.

In meningocele lesions, there is a band of tissue extending from the posterior part of the cervical cord to the duramater and lamina. There is usually only a single band, extending in the midline to the base of the sac.[5,11] However, in some cases, it may extend to the dome of the sac.[12] In our case, the band of tissue was found to extend to the base of the sac. The lesion was a meningocele in continuity with only the subarachnoid space, which was demonstrated by cervical MRI. It was found to be devoid of neural tissue during the operation, which was also confirmed by histopathological investigation. Some studies showed a direct association between cervical cord tethering and congenital mirror movements.[13] However, the tethering of the spinal cord is not directly responsible for mirror movements. The abnormal organization of the spinal pathways at the site of tethering may underlie the mirror movements or at least may contribute to them.

In contrast to the frequent abnormalities in the skin and spinal interspace, which are observed in cases of spinal dysraphism, neurological problems are seldom encountered in pathologies only involving the subarachnoid space, such as dermoid sinus, meningocele and myelocystocele.[6,7] Similarly, no pathological sign was observed in our patient, except for the mirror movement. Spina bifida cystica, as in our case, is usually found in

the thoracic and lumbar regions, but is seldom located in the cervical region, for which the incidence rate is only 3.7%,[14] and bone abnormalities at this location are usually associated with neural abnormalities.[10]

The neurophysiological mechanism involved in the type of movement disorder which is referred to as "mirror movement" is thought to be due to medullary pyramidal decussation decompression (please remove "de") or incompetence and incomplete maturation of the motor inhibition center responsible for non-synergic movements.[15] Because of these discrete origins, several cases have been reported in which mirror movement was reported together with various neurological disorders, such as frontal lobe or corpus callosum lesions, cerebral tumors, subarachnoid hemorrhage, extrapyramidal disorders and cerebrovascular diseases.[10,15] Furthermore, signs of mirror movement are also observed in cases of cord lesions that are associated with abnormal pathways within the pyramidal tract or at a cervical location.[10,15,16] In the present case, the tethering band found during the operation, extending from the backside of the spinal cord to the base of the sac, was thought to exacerbate the abnormal movement.

The series referred to above, concerning cases of cervical cord tethering, consisted entirely of pediatric patients. However, it is not easy to detect mirror movements in this age group and, therefore, it is likely that mirror movements were present in some of these patients, without this being recognized by the clinician. Also, many such patients may be subclinically affected, and therefore may simply be more clumsy at two-handed activities.[13,17]

A risk of contracting meningitis exists in all cases in which the skin and subarachnoid space are in communication with each other, since the dermoid sinus tract is lined with epithelial tissue, which is in continuity with the skin surface and the deep neural structures. Many authors have documented cases of meningitis (including the recurrent type)[7,18,19] and even intramedullary abscess[20,21] complications in similar types of reports, and they particularly emphasized the high incidence of surgical intervention, which was required even after proper antibiotics therapy for the recurrent meningitis.

Dysraphisms are seldom encountered in the cervical region. They are usually associated with various underlying pathologies. These kinds of lesions should be considered to be a likely cause of recurrent meningitis. Although many causes have been suggested for the development of mirror movement, we assumed the tethering band of tissue, which extended from the back of the spinal cord to the base of the sac, to be the likely cause in our case. However, further clinical studies, as well as electrophysiological investigations, are required to confirm this conclusion.

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