A Rare Cutaneous Stigma of Severe Neurological Pathologies: Thoracal Dermal Sinus: Case Report

Ciddi Nörolojik Patolojilerin Nadir Görülen Bir Kutanöz Bulgusu: Torakal Dermal Sinüs

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ABSTRACT A 39 year old adult female patient was reported with a rare combination of thoracal dermal sinus tract, diastematomyelia, idiopathic syringomyelia and multipl vertebral anomalies in this patient. Spinal dermal sinus tracts were located most frequently in the lumbar and lumbosacral regions. Thoracal localization of congenital dermal sinus tracts is considerably infrequent. Because of this unusual localization, the lesion was confused with the chronic epidermal cyst which we mostly come across in clinical practice and further radiological examination was not thought. Our case has the property of being a rare case, showing the association of thoracal dermal sinus tract with diastematomyelia, idiopathic syringomyelia and multipl vertebral anomalies without any neurolojic deficit clinically. This case showed us that severe neurological pathologies could be found under a lesion considered as a simple cyst.

Key Words: Syringomyelia; spina bifida occulta

ÖZET Otuz dokuz yaşındaki erişkin kadın olgu, nadir görülen torakal yerleşimli dermal sinüs traktı, diastematomiyeli, idiyopatik siringomiyeli ve multipl vertebra anomalileri kombinasyonu bu yazıda sunulmuştur. Spinal dermal sinüs traktları çoğunlukla lumbal ve lumbosacral bölgelere yerleşmiştir. Doğumsal dermal sinüs traktlarının torakal yerleşimli olması nadirdir. Bu nadir yerleşiminden dolayı lezyon, pratikte sık karşılaştığımız kronik epidermal kist ile karıştırılmıştır ve ileri radyolojik tetkik düşünülmemiştir. Hastamız, klinik olarak hiçbir nörolojik defisit olmadan, torakal dermal sinüs ile idiopatik siringomiyeli, diastematomiyeli ve multipl vertebra anomalilerinin birlikte görüldüğü nadir bir olgu özelliği taşımaktadır. Bu olgu bize basit bir kist olarak düşünülen lezyonun altında ciddi nörolojik patolojilerin olabileceğini göstermiştir.

Anahtar Kelimeler: Siringomiyeli; spina bifida okülta

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Diastematomyelia is a rare congenital spinal cord malformation. Spinal cord is divided by an osseous, cartilaginous, or fibrous septum and there is a double neural tube formation.[1]

A syrinx is defined as a fluid-filled cavity that anatomically lies within the spinal cord parenchyma or the central canal. In many cases syringomyelia is being found incidentially because of mild neurolojic symptoms such as pain, temperature insensitivity.[2]

Dermal sinus tracts are remnants of incomplete neural tube closure and result from a failure of the surface ectoderm and dermal elements to sepa-

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rate from the neuroectoderm embryologically.[3] The association of spinal dermal sinus tracts with syringomyelia or diastematomyelia is very rare. However, dermal sinuses were located most frequently in the lumbar and lumbosacral regions. Here we report an adult patient with an unusual localization of dermal sinus tract associated with asympomatic diastematomyelia, syringomyelia and multipl vertebral anomalies. Because of this unusual localization and clinical findings we first thought, enfected epidermal cyst in diagnosis without need of further radiological examination.

CASE REPORT

39 years old woman admitted to our clinic with an inflamed lesion on her back which was red, sensitive and sometimes flowing. The patient was not suffering from any other medical history so it was considered as infected epidermal cyst. Anti-inflammatory, antibiotic therapy was initiated and the patient was operated after 1 week. Under local anesthesia an elliptical excision was made around the lesion but during exploration it was seen that the base of the lesion was extending up along the thoracic spine and spinal cord (Figure 1). At that time the patient was consulted with neurosurgeon and the handle of the lesion was ligated with no more dissection and excised, so after subcutaneous tissue and skin was closed primarly. Later, cervicothoracic magnetic resonance imaging (MRI) revealed partial and total segmentation anomalies of vertebral corpusus at the C4-5-6 and proximal thoracal level. Multipl syringomyelia cavities , largest craniocaudal length of 2 mm measured were seen at the servical and proximal thoracal medulla spinalis (Figure 2). Also a split appereance of diastematomyelia localized betwen C5-T2 level was observed (Figure 3). The patients has been treated conservatively and no further operation was considered for intradural tethering by neurosurgery as the patient had no neurolojic deficit.

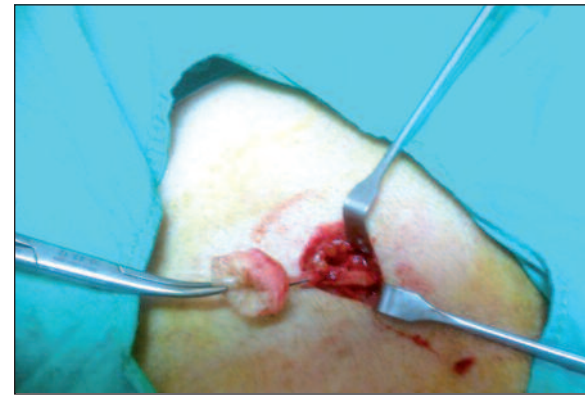

FIGURE 1: Dermal sinus tract with hair in case.

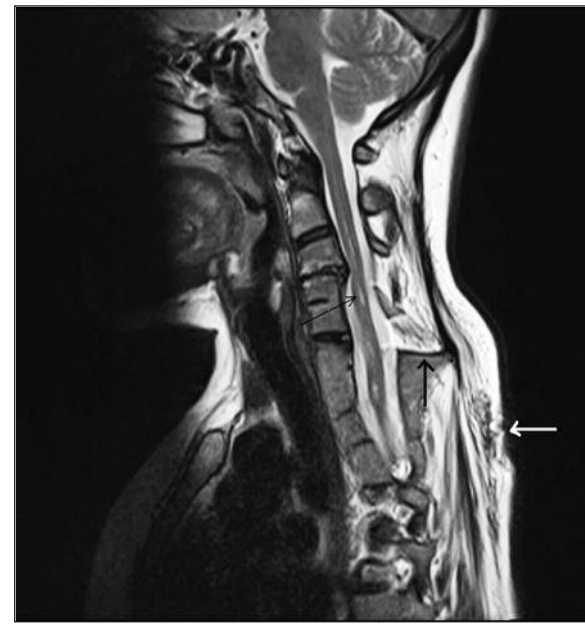

FIGURE 2: Sinus tract (bold arrow) and syringomyelia (thin arrow), clinically the localization of dermal sinus on thoracal skin (white arrow).

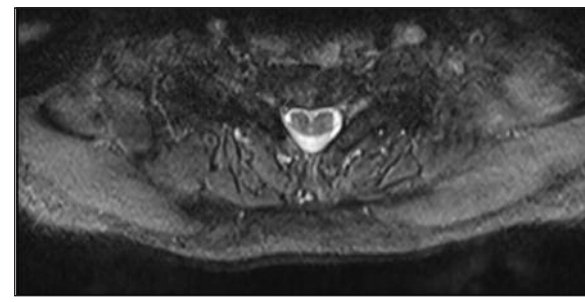

FIGURE 3: Diastematoyelia: Spinal cord divided by fibrous septum.

DISCUSSION

Congenital dermal sinuses represent cutaneous depressions or tracts that are lined by stratified squamous epithelium with surrounding dermal tissue. These sinuses are thought to result from abnormal separation of the cutaneous and neural ectoderm between the third and fifth week of intrauterine

life. They may occur anywhere along the craniospinal axis but the majority of these lesions ocur in the lumbar or lumbosacral region, followed by the occipital and thoracic regions, respectively.[4] They may extend rostrally a considerable distance to terminate several spinal segments above the cutaneous ostium.[5] They may terminate within the subcutaneous tissues or dura, or they may pass through the dura to terminate in the spinal cord, conus medullaris, a nevre root, or a fibrous nodule in the dorsal spinal cord.[6]

In our case we misdiagnosed this inflamed lesion as enfected epidermal cyst which are very common in our clinical practice especially on thoracic areas. However during surgical exploration the unexpected sinus tract progressing toward the spine was observed and further MRI revealed multipl syringomyelia cavities, diastematomyelia and partial and total segmentation anomalies of vertebral corpusus.

Cutaneous diorders are common in diastematomyelia. %20 of asymptomatic patients investigated and diagnosed as suspected cutaneous signs. Hypertrichosis is the most common while lipomas, hemangioma, sinus tract, sacral dimples may also be seen.[1] In our case the dermal sinus tract was the cutaneous sign of diastematomyelia and MRI showed the spinal cord divided by a fibrous septum.

Both idiopathic syringomyelia and diastematomyelia may be asymptomatic. Our case also reported no other complaints or systemic disease. Especially in syringomyelia cutaneous stigmatas are very rare thus syringomyelias are being found occasionally with widespread use of MRI.[7]

We found only one literature reporting the association of dermal sinüs tract, diastematomyelia and syringomyelia in a child with also other neurological pathologies.[8] Our patient is a rare case in the literature showing the association of this severe neurological pathologies with a dermal sinus tract as a skin stigma with no neurolojic deficit. So it should be kept in mind that a severe neurological pathologies could be found under a lesion which are considered as a simple cyst.

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