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Spinal intramedullar cord abscess which developed as a result of congenital spinal dermal sinus

Konjenital spinal dermal sinüs sonucu gelişen spinal intramedüller kord apsesi

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Abstract
Dermal sinus is a type of spina bifida occulta which develops as a result of incomplete separation of the cutaneous ectoderm from the underlying neural ectoderm. It generally has sacral location and may be accompanied by colour change, increased hairiness and sacral dimple. In half of cases the sinus may be associated with the spinal canal. Dermal sinus infection as a result of this may cause dramatic outcomes such as paraplegia and myelitis. The case is here presented of an 18-month old female infant who had started a fever 40 days previously, was taken to a doctor and treatment of antibiotics and antipyretics was started for a diagnosis of pharyngitis but the high fever continued despite the treatment. After 20 days, redness and swelling on the back was noticed together with the fever and as there was a yellow-coloured discharge from the area of the swelling, the doctor was again consulted and the antibiotics were changed. The patient was then brought to our clinic, as in the previous 3 days, there had been increasingly reduced movement of the feet. As a result of tests applied to the patient, intramedullary abscess was determined.
In neonates and infants, the spinal area must be evaluated well. It must be kept in mind that in patients with colour change, increased hair and especially sacral dimple, there could be midline closure defects.

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Key words: Dermal sinus, Sacral dimple, Abscess, Early diagnosis.

Özet
Dermal sinus, spina bifida occultanın bir tipi olup kütanöz ektoderm ile altında bulunan nöral ektodermin ayrılmasının tamamlanamaması sonucu gelişir.Genellikle sakral yerleşimlidir ve ciltte renk değişikliği, kıllanma artışı,sakral gamze eşlik edebilir. Olguların yarısında sinüs spinal kanal ile ilişkili olabilir. Bunun sonucu olarak dermal sinüs enfeksiyonu, parepleji, myelit gibi dramatik sonuçlara neden olabilir. Bir buçuk yaşında kız hasta 40 gün öncesinde ateşi başlamış ve başvurduğu doktor tarafından farenjit tanısıyla antibiyotik ve ateş düşürücü tedavisi başlanmış ancak tedaviye rağmen ateş yüksekliği devam etmiş. Yirmi gün önce ateş yüksekliği ile birlikte sırtında kızarıklık ve şişlik fark edilmiş ve şişlik yerinden sarı renkli akıntı gelmesi üzerine tekrar başvurduğu doktor tarafından antibiyotiği değiştirilmiş. Son üç gündür giderek artan ayak hareketlerinde azalma nedeniyle kliniğimize başvuran ve yapılan tetkikler sonucu intramedüller apse tespit edilen hasta sunuldu. Yenidoğan dönemindeki hastalarda spinal bölge iyi değerlendirilmeli, renk değişikliği, kıllanma artışı ve özellikle sakral gamzesi olan hastalarda orta hat kapanma defektleri olabileceği akılda tutulmalıdır.

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Anahtar sözcükler: Dermal sinüs, Sakral gamze, Apse, Erken teşhis.

Introduction

Congenital spinal dermal sinus (CSDS) is a type of spina bifida occulta which develops as a result of incomplete separation of the cutaneous ectoderm from the underlying neural ectoderm [1]. The incidence of CSDS has been reported as 1 per 2500 births at similar rates for both genders. It may be seen in any location from the occipital region to the coccyx, although it is observed most often in the lumbar and sacral regions [1,2]. There may be accompanying skin colour change, increased hairiness or sacral dimple and the most evident sign is a sinus orifice on the skin. This sinus orifice is often seen as an

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indentation or an atrophic area on the skin, the size of a pinhead, generally on the midline but it may very occasionally be paramedian. Hair may develop around the sinus orifice or a red mark may be seen. The hair is usually sparse and in the form of 'silky down' [3,4]. In half of cases the sinus may be associated with the spinal canal. Dermal sinus infection as a result of this may cause dramatic outcomes such as paraplegia and myelitis [5]. To determine the underlying spinal pathologies of these midline skin symptoms and to prevent neurological damage which may occur associated with these, there must be referral to imaging methods [6]. The case is here presented of an infant whose parents noticed a red mark in the lumbar region from birth and a sacral dimple. The infant was taken to a doctor because of fever, a discharge was noticed on the lower back and medical treatment was started. As a result of dermal sinus infection during follow-up, intramedullary abscess and paraplegia developed. This case of CSDS is here presented.

Case

An 18-month old female infant was brought to our polyclinic with the complaints of swelling and discharge on the lower back and lack of movement of the feet. Since birth, the parents had been aware of a red colour change and small orifice on the lower back over the hip of the child and although close relatives and the family doctor had said this was normal they had previously consulted the brain surgery clinic. Mental motor development was consistent with the age of the infant. The patient experienced no problems until 40 days prior to presentation at our clinic, when a fever developed and the patient was taken to a paediatrician. As a result of examination, treatment of antibiotics and antipyretics was started for a diagnosis of pharyngitis. As the fever did not subside, the antibiotics were changed. The fever continued intermittently and after 20 days, redness and swelling on the back was noticed together with the fever and as there was a yellow-coloured discharge from the area of the swelling, the doctor was again consulted and for the third time, the antibiotics were changed.

The patient was then brought to our polyclinic, as together with the continuing fever, inability to stand and lack of movement of the feet had started 3 days previously. The physical examination determined subcutaneous oedema between T5-L5 and redness and swelling of 3 x3 cm at L5-S3. The patient was paraplegic in the lower extremities and there was no anal sphincter tonus. The patient also exhibited profound hypesthesia below the C7 dermatome. Bilateral biceps jerks were weak, and bilateral knee and ankle jerks were absent. Meningeal irritation sign was observed. The laboratory test results showed CRP 54.3 mg/L, white cell 31620 K/UI and haemoglobin 8.9 g/dl. Contrast lumbar magnetic resonance imaging (MRI) was applied to the patient who was paraplegic with swelling, redness and discharge in the lumbar region. On MRI, oedema was determined in the central cord between T5-T9 and a meningeal type contrast enhanced appearance with a central cystic nature from the periphery at the dorsolumbar cord level between T9-S2. On lumbar CT, dorsal dermal sinus + posterior bone at the lower sacral level was determined as a fusion defect in the spinal canal (Figures 1 and 2). The patient was admitted for surgery, the abscess was drained and antibiotic therapy was started. Multiple pathogens were suspected and the patient received vancomycin and cefotaxime for empirical therapy. Later, *Peptoniphilus asaccharolyticus* and an unidentified anaerobic rod were isolated from the pus and we added metronidazole to the drug regimen. The patient received the triple antibiotics for 8 weeks following the operation. On review 6 months later, the motor deficit in the left foot recovered and in the right foot a deficit of 3+/5 remained.

Discussion

In CSDS, although skin findings are seen from birth, neurological findings may not be seen in the early stages. Therefore, diagnosis may be delayed. In the current case, although the parents had been aware of the skin redness and sacral dimple since birth, the paediatrician they had consulted had said it was normal and as a midline closure defect was not considered, the diagnosis was delayed.

For a diagnosis of CSDS in infants determined with sacral dimple in the physical examination, spinal surface ultrasonography can provide information about the relationship of the spinal canal with the sinus, as the posterior spinal elements are cartilage structures in the first 3-6 months. However, the gold standard in imaging is MRI (6). Early stage excision of

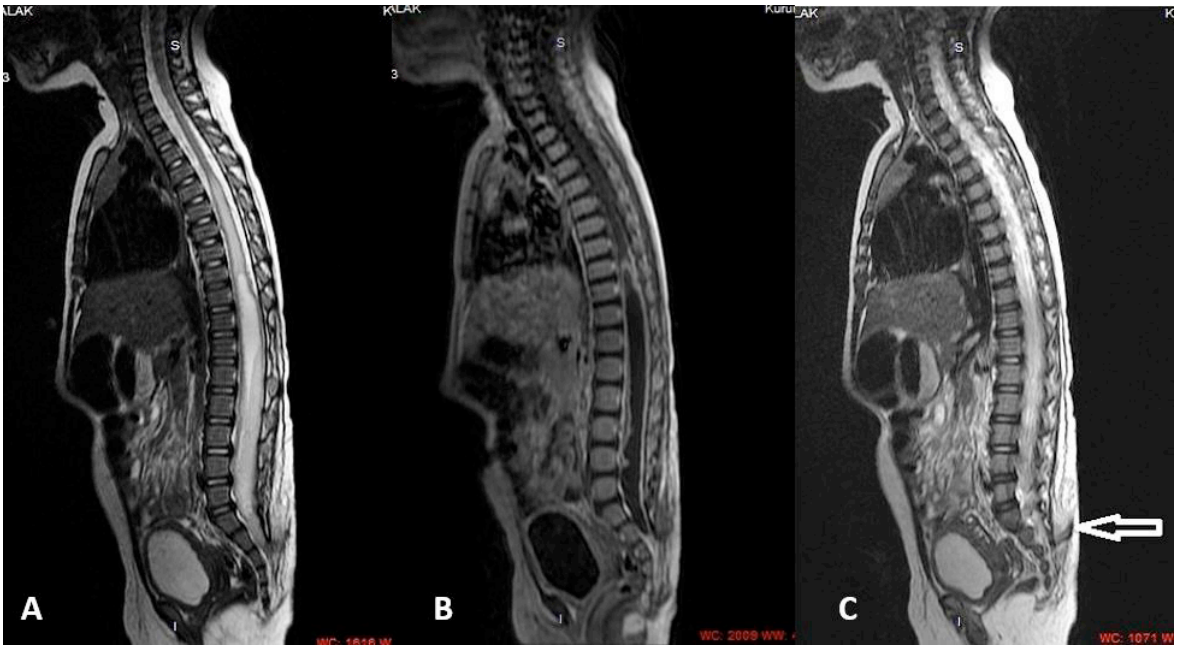

Figure 1. On the sagittal MRI, oedema was determined in the central cord between T5-T9 (A) and a meningeal type contrast enhanced appearance with a central cystic nature from the periphery at the dorsolumbar cord level between T9-S2 (B). A dermal sinus tract (arrow) is seen from the skin dimple to a bony defect at the S2 level (C).

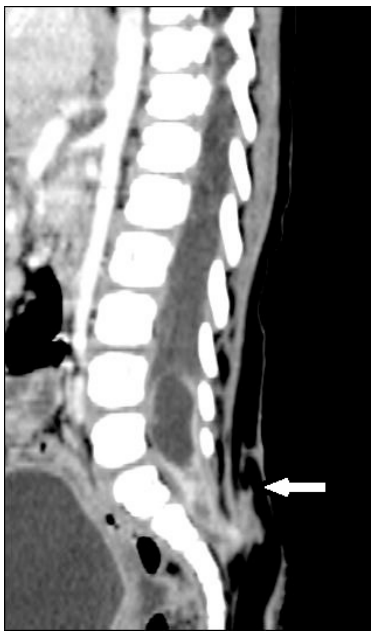

Figure 2. On lumbar CT, dorsal dermal sinus + posterior bone at the lower sacral level was determined as a fusion defect in the spinal canal.

dermal sinus tracts determined with physical examination and imaging methods may prevent the development of progressive and destructive infections in the patient (4). The most frequently seen complication in CSDS patients is meningitis and the least seen but most severe complication is intramedullary abscess (IMA) (1). While IMA may be seen at any age it is most often seen in children below the age of 5 years [6]. Diagnosis is difficult and may be delayed until the development of neurological deficits [4,6]. Progressive sensory and motor losses are the most commonly seen findings of IMA. Depending on the level of abscess involvement, motor deficit is seen. It has been reported that paraplegia develops most in patients [4,7]. In the current case, diagnosis was not made until the age of 18 months and the patient had been given several antibiotic treatments as there was redness, swelling and discharge in the lumbosacral region and the onset of fever 40 days prior to presentation. The complication of

Several authors have recommended early surgery for these patients [1,2,4]. Drainage of the abscess with laminectomy and dorsal midline myelectomy is an effective method for the control of infection and functional recovery [2]. Laminectomy at multiple levels is not recommended as the risk of later stage complications is increased [8]. In some cases reported in literature, full recovery has been reported, while in 20% there has not been full recovery [4]. It has been reported that neurological recovery could be related to the rate and level of intramedullary involvement and the time of diagnosis and surgery [2]. Delays in

diagnosis and incorrect diagnosis may have dramatic neurological results and even cause death [9]. In the current case, early surgery was applied immediately after presentation but the neurological table did not fully recover. Consistent with literature, it can be thought that the incomplete recovery of the current patient was related to the length of time that passed from the onset of fever to the diagnosis.

In conclusion, in neonates and infants, the spinal area must be evaluated well and it must be kept in mind that in patients with colour change, increased hair and especially sacral dimple, there could be midline closure defects. With the diagnosis of CSDS in the early stage with imaging methods and the application of appropriate treatment, the development of more severe and irreversible complications can be prevented.

Yazarlar çıkar ilişkisi olmadığını beyan eder.

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