Congenital dermal sinus with extensive intramedullary expansion and an infected spinal epidermoid cyst in an infant

Saugata Acharyya¹, Sanchari Chakravarty¹

¹Department of Pediatrics, Calcutta Medical Research Institute, Kolkata, West Bengal, India

Abstract

An 11-month-old female child presented with progressive weakness involving both lower limbs and left upper limb. There was hypertonia and hyperreflexia in the affected limbs. This was associated with multiple episodes of urinary tract infection and overflow incontinence of the urinary bladder. Magnetic resonance imaging of the spine revealed extensive intramedullary expansion of a congenital dermal sinus. There was a cystic lesion along the lower end of the sinus tract. Surgical exploration had confirmed the presence of an infected epidermoid cyst along with the dermal sinus tract. The entire tract as well as the abscessed epidermoid cyst was removed. Following this, the child showed gradual improvement of neurological function and bladder dysfunction. This is perhaps the first report in current literature of a congenital dermal sinus tract with such an extensive intramedullary extension (almost the entire spine) and an infected epidermoid tumor in an infant.

Keywords: Congenital dermal sinus, intramedullary extension, spinal epidermoid cyst

Introduction

Congenital dermal sinus tract involving the spine is a rare variety of spinal dysraphism. They may be limited to simple intradermal tract or may have significant intramedullary cystic extension. Rarely, a congenital dermal sinus tract may be associated with inclusion tumors such as spinal epidermoid cysts. A significant intramedullary extension of a congenital dermal sinus along with an enlarging congenital epidermoid tumor may give rise to a variety of neurological symptoms due to pressure effect on the adjoining neural tissue. These may include motor, sensory, and bladder involvement. Rarely, the associated epidermoid cyst gets infected resulting in the formation of spinal abscess.

We report a case of an 11-month-old infant presenting with significant weakness in both lower and left upper extremities.

Address for correspondence: Dr. Saugata Acharyya, 63 Raja Basanta Roy Road, Kolkata, West Bengal, India. E-mail: acharyyasaugata@yahoo.com

The parents had reported that the child finds it difficult to sense the difference between hot and cold water. There was marked bladder involvement with recurrent overflow incontinence. There were upper motor neuron signs in the affected limbs and marked scoliosis. There was no significant abnormality in the back except for an innocuous looking cutaneous mole. Magnetic resonance imaging (MRI) revealed a dermal sinus tract with an intramedullary expansion almost involving the entire spinal cord. There was an associated cystic area along the path of the dermal sinus in the lumbosacral area. The spinal lesion was successfully removed along with the cystic mass. Subsequent histopathological examination confirmed the diagnosis of an infected epidermoid cyst. The pus from the cyst grew Gram-negative bacilli resembling the organism that was isolated in the urine culture. The mass effect from the elongated dermal sinus and the abscess formed by the infected epidermoid cyst led to the neurological symptoms of the child.

Access this article online

Quick Response Code:

Website:

www.jfmpc.com

DOI:

10.4103/jfmpc.jfmpc_158_18

How to cite this article: Acharyya S, Chakravarty S. Congenital dermal sinus with extensive intramedullary expansion and an infected spinal epidermoid cyst in an infant. J Family Med Prim Care 2018;7:1103-5.
Case Details

An 11-month-old female baby had presented with weakness involving both lower limbs and left upper limb. There was a small cutaneous mole at the back of the child just above the buttock, which was present since birth. The parents had noticed that around the age of 7 months she could not sit without support and could barely move the lower limbs. Subsequently, the weakness progressed, gradually involving the left hand. The baby was born by normal vaginal delivery at term weighing 2.7 kg. There was no other birth history available except that the baby had cried immediately following birth. The baby had three attacks of documented urinary tract infection in the previous 4 months and was treated with oral antibiotics as per urine culture sensitivity.

On examination the baby was irritable. Her weight was 7.1 kg and occipitofrontal circumference was 41 cm. There was marked spastic paraparesis of both lower limbs with exaggerated deep tendon reflexes. There was weakness of the left upper limb and some bite marks (self-inflicted as reported by the parents) in both hands.

Investigations revealed a significant (colony count >100,000) pure growth of multiresistant *E. coli* in her urine. This was treated with broad spectrum intravenous antibiotics. MRI of spine revealed extensive hyperintense signals involving the entire spinal cord with inhomogeneous enhancement and diffuse altered signals in lumbar thecal sac [Figure 1]. The cauda equina roots could not be identified and a loculated cystic lesion at the level of L5-S1 was identified [Figure 2]. There was significant intramedullary extension of a dermal sinus tract in the sagittal view [Figures 1 and 2].

In view of the MRI findings and the presenting clinical signs, neurosurgical opinion was sought. The entire dermal sinus was removed. The histopathological examination of the cystic lesion confirmed the diagnosis of epidermoid cyst, which was infected resulting in an abscess formation.

Discussion

Congenital dermal sinus tracts seem to originate as a result of incomplete separation of the neuroectoderm from cutaneous ectodermal elements.[1-4] The incidence of congenital dermal sinus is 1 in 2500 live births.[5] A rare variety of congenital dermal sinus may have significant intramedullary extension, giving rise to pressure effect on the adjoining neural tissue. This may result in various motor and sensory symptoms along with bladder and bowel involvement. During the formation of the neural tube, inclusion of epithelial elements may occur along the path of a dermal sinus. These may form various inclusion tumors such as spinal epidermoid, dermoid, or teratoma.[6-9] Congenital spinal epidermoid cystic inclusion tumors are uncommon but are more frequent than the iatrogenic variety caused by repeated attempts of lumbar puncture.[10] The usual presentations of congenital dermal sinus include meningitis, mass effect of the associated inclusion tumor, or symptoms caused by tethered cord.[11] At times even a long internal sinus tract may leave innocuous external cutaneous signs at the back along the spine. The pressure symptoms associated with spinal epidermoid cysts are similar to those of any radiculopathy. Gradual worsening of symptoms suggests enlargement of the cyst, whereas sudden deterioration signifies rupture of the cyst and associated meningitis. The recommended treatment is resection of the dermal sinus tract along with removal of the epidermoid inclusion elements.

We report the case of an infant presenting with extensively elongated congenital dermal sinus tract and an associated congenital epidermoid cystic inclusion tumor. These together had resulted in significant pressure effect and neurological symptoms in this infant. The parents had noticed a small mole around the lumbosacral region when the child was born. In view of the insignificant nature of the external cutaneous lesion, no further investigation was done. There was history of recurrent urinary tract infection with significant overflow incontinence. These may also be explained by the spinal lesions. The removal of the dermal sinus tract along with the infected epidermoid cyst resulted in gradual recovery of her neurological function. The urinary bladder dysfunction was cured with no further recurrence.
of urinary tract infection. This case is worth reporting because of the age of presentation and the extent of the sinus. In the study of 23 pediatric case reports, only two children were below 2 years and only one had spinal abscess, which this infant had.[11]

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References
1. Ackerman LL, Menezes AH. Spinal congenital dermal sinuses: A 30 year experience. Pediatrics 2003;112:641-7.
2. Kanev PM, Park TS. Dermoids and dermal sinus tracts of the spine. Neurosurg Clin N Am 1995;6:359-66.
3. Lemire RJ, Loeser JD, Leech RW, Alvord EC. Normal and abnormal development of human nervous system. Hagerstown, MD: Harper & Row; 1975.
4. Wang KC, Yang HJ, Oh CW, Kim HJ, Cho BK. Spinal congenital dermal sinus: Experience of 5 cases over a period of 10 years. J Korean Med Sci 1993;8:341-7.
5. McComb JG. Congenital dermal sinus. Pang D: Disorders of the Pediatric Spine. New York: Raven Press; 1995. p. 349-60.
6. Alafaci C, Salpietro FM, Grasso G, Collufio D, Caruso G, Morabito A, et al. Lumbosacral congenital dermal sinus presenting in a 52 year old man. Case report. J Neurosurg Sci 2000;44:238-42.
7. Black SPW, German WJ. Four congenital tumors found at operation within the vertebral canal; with observations on their incidence. J Neurosurg 1950;7:49-61.
8. French BN. The embryology of spinal dysraphism. Clin Neurosurg 1983;30:295-340.
9. Martinez-Lage JF, Esteban JA, Poza M, Casas C. Congenital dermal sinus associated with an abscessed intramedullary epidermoid cyst in a child: Case report and review of the literature. Childs Nerv Syst 1995;11:301-05.
10. Manno NJ, Uihlein A, Kemohan JW. Intraspinal epidermoid cysts. J Neurosurg 1962;19:754-65.
11. Jindal A, Mahapatra AK. Spinal congenital dermal sinus: An experience of 23 cases over 7 years. Neurol India 2001;49:243-46.