Neuroradiology (2000) 42: 51–53

W. C. Shen
T.-L. Chiou
T. Y. Lin

Dermal sinus with dermoid cyst in the upper cervical spine: case note

Received: 28 March 1999
Accepted: 4 May 1999

W. C. Shen (✉)
Department of Radiology, and School of Medicine, China Medical College, China Medical College Hospital, No. 2 Yuh-Der Road, 407 Taichung, Taiwan, Republic of China
Tel.: + 886-4-2052121
Fax: + 886-4-2034174
e-mail: wcshen@ficnet.net

T.-L. Chiou
Department of Neurosurgery, China Medical College Hospital, No. 2 Yuh-Der Road, 407 Taichung, Taiwan, Republic of China

T. Y. Lin
Department of Pathology, China Medical College Hospital, No. 2 Yuh-Der Road, 407 Taichung, Taiwan, Republic of China

Abstract We describe a 5 year-old girl who had a skin dimple of the back of her upper neck. MRI showed a dermal sinus tract in the upper cervical spine, associated with an intramedullary dermoid cyst at C2–3, and spina bifida. A laminectomy was performed, the dermoid cyst and the sinus tract were completely removed. This congenital complex is very rare.

Key words Sinus dermal · Cyst dermoid · Magnetic resonance imaging

Introduction

Spinal congenital dermal sinus is rare, and supposedly results from failure of the neuroectoderm to separate from the cutaneous ectoderm during neurulation [1]. It is found most frequently in the lumbosacral followed by the occipital regions. In the cervical region it is very uncommon; Wright [2] collected 127 cases of congenital dermal sinus from the literature, only two of which were in the cervical spine. Peter et al. [3] reported 15 children with spinal dermal inclusions, 13 in the lumbosacral area, and one was in the cervical spine. We report a very rare case of cervical dermal sinus associated with an intramedullary dermoid cyst, which severely compressed the spinal cord.

Case report

A 5-year-old girl had a dimple in the midline of her upper neck posteriorly at birth. She occasionally complained of neck pain, but 2 months before admission had gait disturbance and easily fell down. There had been no discharge from this dimple, and no history of meningitis.

Examination revealed mild weakness of both legs and both hands, with increased deep tendon reflexes at the knees, and a Babinski sign in both feet.

Plain radiographs of the cervical spine were interpreted as normal. MRI showed a round lesion in the spinal canal of C2–3, and a tract connecting it to the skin dimple. The lesion gave heterogeneous low signal on T1-, and high signal on T2-weighted images (T2WI), and was thought to be a cyst-like tumour (Fig. 1a, b). The spinal cord was seen to be stretched around the intramedullary cyst. On axial images, spina bifida of C3 was noted (Fig. 1c).

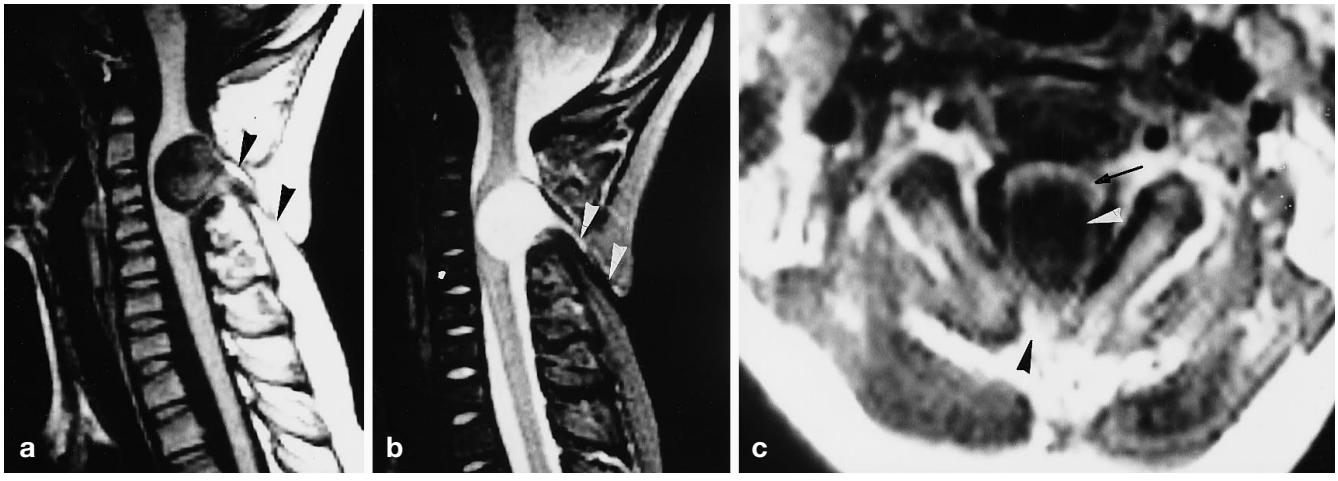

Fig. 1 **a** T1-weighted sagittal image. There is a round cyst with heterogeneous signal in the spinal cord at C2–3. A tract *(arrowheads),* the dermal sinus, connects this a skin dimple. **b** T2-weighted image. The cyst contains high-signal material, communicating with the dermoid sinus *(arrowheads).* **c** Axial T1-weighted image. There is a cyst *(white arrowhead)* in the spinal cord, which appears paper-thin *(arrow).* There is spina bifida *(black arrowhead)*

A C2–3 laminectomy level was performed and a cyst was found in the upper cervical spinal cord. It was removed completely, and the subcutaneous tract was resected. The postoperative course was uneventful, and power in all four limbs gradually returned to normal.

Pathological examination of the cyst showed it composed of squamous epithelium and skin appendages, typical of dermoid cyst (Fig. 2).

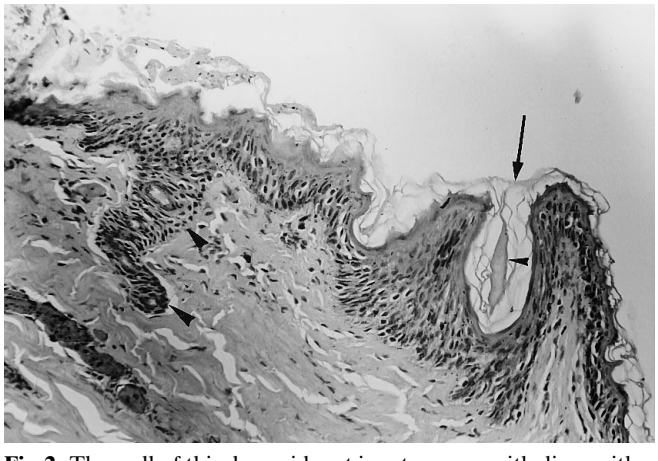

Fig. 2 The wall of this dermoid cyst is cutaneous epithelium with a hair follicle *(arrow),* hair shaft *(small arrowhead),* and acrosyringia *(arrowheads)*

Discussion

Congenital dermal sinuses occur most commonly in the lumbosacral region, are usually associated with a spina bifida, and may connect the skin directly to the spinal canal [4]. Epidermoid and dermoid cysts may form at any point along a dermal sinus. Cutaneous or subcutaneous abnormalities commonly seen with occult spinal disorders are abnormal hair, angioma, lipoma and dimple. Our patient showed a very rare complex: a dermal sinus at C2–3, associated with a dermoid cyst, and with skin dimple.

French [5] reviewed intracranial congenital dermal sinuses, finding 89 % to be associated with an inclusion tumour. The latter was extradural in 18 %, and deeper in 82 %; most were dermoid cysts, epidermoid cysts being rare. However, Shikata et al. [6] reported seven cases of intraspinal inclusion cysts, four epidermoid and three dermoid. In the series of Barkovich et al. [7], five of seven dorsal spinal dermal sinuses, three were associated with epidermoids, two with dermoids.

Patients with spinal dermal sinus may present with meningitis and/or mass effect from the associated inclusion tumour [1, 8]. Kanev and Park [9] said if the dermal sinus ends blindly and never extend intraspinally, there is no infectious or mechanical risk to the developing nervous system. The midline must be carefully inspected when a child of any age suffers meningitis, especially when an unusual organism is cultured [9]. If an inclusion tumour is intramedullary, there is a high risk of intramedullary abscess [10]. Our patient had an intramedullary dermoid cyst, but had no history of meningitis.

Cervical spinal bifida below C1, either occult or overt, is relatively rare [11]. Such high spinal dysraphism usually involves a meningocele or dermal sinus tract [12], the Klippel-Feil syndrome, or a Chiari II malformation [11].

References

1. Wang KC, Yang HJ, Oh CW, Kim HJ, Cho BK (1993) Spinal congenital dermal sinus – experience of 5 cases over a period of 10 years. J Korean Med Sci 8: 341–347
2. Wright RL (1971) Congenital dermal sinuses. Prog Neurol Surg 4: 175–191
3. Peter JC, Sinclair-Smith C, de Villiers JC (1991) Midline dermal sinuses and cysts and their relationship to the central nervous system. Eur J Pediatr Surg 1: 73–79
4. Harris HW, Miller OF (1976) Midline cutaneous and spinal defects. Midline cutaneous abnormalities associated with occult spinal disorders. Arch Dermatol 112: 1724–1728
5. French BN (1990) Midline fusion defects and defects of formation. In: Youmans JR (ed) Neurological surgery. Saunders, Philadelphia, pp 1164–1169
6. Shikata J, Yamamuro T, Mikawa Y, Kotoura Y (1988) Intraspinal epidermoid and dermoid cysts. Surgical results of seven cases. Arch Orthop Trauma Surg 107: 105–109
7. Barkovich AJ, Edwards MS, Cogen PH (1991) MR evaluation of spinal dermal sinus tracts in children. AJNR 12: 123–129
8. Gurbani SG, Cho CT, Lee KR (1994) Staphylococcus epidermidis meningitis and an intraspinal abscess associated with a midthoracic dermal sinus tract. Clin Infect Dis 19: 1138–1140
9. Kanev PM, Park TS (1995) Dermoids and dermal sinus tracts of the spine. Neurosurg Clin N Am 6: 359–366
10. Martinez-Lage JF, Esteban JA, Poza M, Casas C (1995) Congenital dermal sinus associated with an abscessed intramedullary epidermoid cyst in a child: case report and review of the literature. Childs Nerv Syst 11: 301–305
11. Steinbok P (1995) Dysraphic lesions of the cervical spinal cord. Neurosurg Clin N Am 6: 367–376
12. Vogter DM, Culberson JL, Schochet SS, Gabriele OF, Kaufman HH (1987) "High spinal" dysraphism. Case report of a complex cervical meningocele. Acta Neurochir (Wien) 84: 136–139
13. Crols R, Appel B, Klaes R (1993) Extensive cervical intradural and intramedullary lipoma and spina bifida of C1: a case report. Clin Neurol Neurosurg 95: 39–43