Although the association of lumbosacral spinal dysraphism and congenital spinal dermoid cysts is well known, the association of craniocervical spinal anomalies and posterior fossa dermoid cysts has only been recognized recently. Advances in imaging technology and awareness of the association likely contribute to an increase in recently reported cases [1].

In 1936, the relationship between posterior fossa dermoid cysts with cervical fusion anomalies such as the Klippel-Feil syndrome was first recognized [2]

Since then, approximately 18 other cases of this rare association have been reported [3] [4] [5] [6].

Several hypotheses have been proposed to explain the em- bryologic association of a posterior fossa dermoid and KFS. Failure of segmentation of the cervical sclerotomes leads to the Klippel-Feil anomaly and occurs after the formation of the entire neuraxis. The related failure of cleavage of ectoderm from neuroectoderm resulting in entrapment of dermal ele- ments within the closing neural tube may contribute to the association of Klippel-Feil anomalies and dermoid cysts [7].

Other proposed theories include overdistention of the neural tube resulting in distortion of the somites and reduced expres- sivity of the Hox or Pax genes, the highly conserved DNA sequences that control the development of the intervertebral disks [8].

A mechanical basis to explain the relationship between these abnormalities is that during the formation of the ce- phalic and cervical brain flexures, a shortening of the cervical spine because of a reduction or fusion in the number of somites may result in altered tissue tension, which could lead to entrapment of dermal elements [9].

Patients with KFS and posterior fossa dermoid cysts present with a variety of signs and symptoms, which may be attributed to both craniovertebral bony anomalies and in- creased intracranial pressure secondary to mass effect. It is interesting to note that, though seen in a minority of patients,the presence of mirror movements is more likely to be seen in children with Klippel-Feil anomalies and neuroschisis.

The exact anatomic basis of mirror movements remains in ques- tion, but this sign may be a clue to an occult posterior fossa dermoid in children with Klippel-Feil deformity [10].

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