



Spectrum of Dermal sinus and its embryological implication

KEY WORDS

spinal dermal sinus- spinal dysraphism-congenital malformation-skin dimple-disjunction of surface ectoderm

Dr.J.Srisaravanan

Prof Department of Neurosurgery Madurai Medical College Madurai

ABSTRACT

Dermal sinus is one of the developmental anomalies involving the spinal cord. It is mainly due to failure of disjunction of surface ectoderm from the neuroectoderm. We analyzed 6 cases of dermal sinus admitted in our department. All the patients had varying internal abnormalities. The similarity in them is the presence of dermal sinus. The aim of study is to highlight the various types of anomalies associated with the spinal dermal sinus.

Materials and Methods

We have analyzed case admitted with the complaints of dermal sinus in our department. All the patient underwent relevant clinical and radiological examinations. They were presented with varying spectrum of clinical presentation with one common feature of spinal dermal sinus

The neurological deficits are also of varying degrees according to the internal pathology. Their clinical features and findings are summarized in the following table

Table 1 Clinical and Radiological Features

s.no	Age	Sex	External feature	Clinical feature	Location of Dermal sinus	MRI
Case 1	23	m	dermal sinus	Pain in the sinus area	lumbar spine	Tethering of cord and low ending of spinal cord. Spina bifida
Case 2	23	f	Dermal sinus	Spastic paraparesis with bladder involvement	Dorsal spine	Intra medullary epidermoid with lipomatous elements
Case 3	12	f	Dermal sinus	Restricted neck movement	Cervical spine	Anteriorly placed neuro enteric cyst
Case 4	5	m	Dermal sinus	Bladder and bowel incontinence, Motor weakness	Lumbosacral	Tethered cord, terminal lipoma
Case 5		m	Dermal sinus	No neurological deficit	Dorsal d3level	Cord is tethered to the dermal sinus
Case 6	7	m	Dermal sinus	Bladder and bowel incontinence	Lumbar	Tethered cord

Table 2 intra operative finding of dermal sinus with possible embryological explanation

S.No	Intra operative	Possible Embryology	Outcome
Case 1	Simple tethering of cord dura and thickened filum terminale	Failure of disjunction of surface ectoderm from the neuroectoderm. Spina bifida is part of Neural tube defect	Same as preoperative

Case 2	Sinus tract is connected to the spinal cord and it is associated with intramedullary dermoid	Between the 3rd and 5th weeks of fetal development, cells fated for cutaneous ectoderm somehow become trapped within neural ectoderm during neural tube closure and eventually form tumors.	Spastic paraparesis and bladder involvement improved
Case 3	Anteriorly placed cystic lesion which is tapped and biopsy is taken from the wall	At the end of the third embryonic week, the development of the notochord is intimately related to endodermal cells. If the notochord fails to detach itself from the endodermal layer, endodermal cells can be dragged forwards and upwards. This may lead to the formation of a cyst in front of spinal cord. Persistent accessory neuroenteric canal	Same as preop
Case 4	Dermal sinus ended in the terminal part of blind end of dural sac with tethering of cord	Lipoma of the filum terminale is probably due to persistence of caudal cells that differentiate to fat cells. Tight filum is due to deranged canalization and retrogressive differentiation disorder	CSF leak and wound gapping needed secondary suturing. Neurological status same as preop
Case 5	Dermal sinus extended into the dura and attached to the underlying spinal cord	Failure of disjunction of surface ectoderm from the neuroectoderm	Neurological status same as preop
Case 6	Dermal sinus extended to dura and associated with tethered cord	Failure of disjunction of surface ectoderm from the neuroectoderm	Same as preop

All cases were operated by standard laminectomy and excision of dermal sinus. Underlying abnormalities were dealt according to the pathology. In cases with intra medullary lesion standard myelotomy and excision of the lesion was done. All the cases were followed up till date.

Discussion

Incidence of dermal sinus is about 1 in 2500 live birth. 1% of all tracts along the spine were cervical, 10% were thoracic, 41% were lumbar

and 35% were lumbosacral. In Our observation of 6 cases, one case was cervical and two cases were thoracic and three cases were lumbosacral. In three cases failure of disjunction will explain the dermal sinus. The presence of neuroenteric cyst, intramedullary dermoid and lipoma in the tethered cord in association with dermal sinus cannot be explained by the simple disjunction theory. We tried to explain these anomalies with dermal sinus At the end of the third embryonic week, the notochord is intimately related to endodermal cells. If the notochord fails to detach itself from the endodermal layer, endodermal cells can be dragged forwards and upwards. This may lead to the formation of a cyst in front of spinal cord. This persistent accessory neuroenteric canal associated with ectodermal disjunction leads to the formation of neuroenteric cyst in the dermal sinus.

In case 4 Lipoma of the filum terminale is probably due to persistence of caudal cells that differentiate to fat cells. Tight filum and dermal sinus is due to deranged canalization and retrogressive differentiation.

In case 2 between the 3rd and 5th weeks of fetal development, cells fated for cutaneous ectoderm somehow become trapped within neural ectoderm during neural tube closure and eventually form dermoid and it is connected to skin as dermal sinus.

Conclusion

All spinal dermal sinus patients have to be evaluated completely for the presence of neurological abnormality. Simple non disjunction of surface ectoderm from the neuroectoderm theory cannot explain the spectrum of anomalies present in dermal sinus. It needs a detailed study and large number of cases for statistical signification

References

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