Faun tail - A skin marker of occult spinal dysraphism
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Abstract : Spinal dysraphism refers to incomplete fusion of midline structures of the embryonal dorsal median region that may affect any combination of somatic ectoderm, neuroectoderm or mesoderm. As skin and neuron tissue are of ectodermal origin, anomalies of both may occur simultaneously. Occult spinal dysraphism is characterised by skin covered lesions without exposed neural tissue. Congenital cutaneous mid line paraspinal lesions,mostly localised in the lumbosacral region are widely recognised as markers of occult spinal dysraphism. Cutaneous changes may include a lipoma, dermal sinus or cyst, hypertrichosis, dimple, hyperpigmentation, capillary hemangioma and aplasia cutis. Abnormal lumbar hypertrichosis may present as a faun tail which is a rare entity and cutaneous marker of occult spinal dysraphism. Here we present an asymptomatic infant with faun tail and split cord malformation type-1 (Diastematomyelia).

INTRODUCTION
Spinal dysraphism refers to incomplete fusion of midline structures of the embryonal dorsal median region that may affect any combination of somatic ectoderm, neuroectoderm or mesoderm. As both skin and nervous tissue are of ectodermal origin, anomalies of these tissues may occur simultaneously. Abnormal lumbar hypertrichosis may present as a 'faun tail' which is a rare entity and cutaneous marker of occult spinal dysraphism. We report an infant with faun tail and split spinal cord malformation type-1 (Diastematomyelia).

CASE REPORT
A 6 month old male infant was brought to our outpatient department with an excessive amount of hair located on his lower back. The lesion had been present since birth. The child had no specific complaints. Antenatal history was uneventful. Antenatal scan was taken at 7th month of gestation but the report was normal study. He was the 2nd born of a non-consanguineous marriage and had one elder sister. He was born by vaginal delivery at term, birth weight-2.5kg. He cried soon after birth and post natal history was uneventful. Developmental assessment revealed normal developmental milestones. He was able to sit without support. He was immunised for the age. Bowel and bladder habits were normal. There was no family history of similar lesion. Growth parameters were normal, including head circumference.

On examination the infant was alert, active, playful and interested in his surroundings. He had no facial dysmorphism and focal neurological deficit. He had full range of movements in all four extremities. Local examination revealed a circumscribed area of hypertrichosis with coarse dark hair of varying length overlying lumbosacral region. Neurological examination was normal. As this entity is a cutaneous marker of underlying spinal anomalies x-ray and MRI spine was taken. The x-ray of dorsolumbar spine showed fusion of D12 and L1 vertebra. The MRI of dorsolumbar spine showed two hemi cords contained with its own dural sheath and separated by osteocartilaginous medial septum extending from D12 to L3 with osseous septum L1,L2 level(split cord malformation type-1). The paediatricians are the first physicians to see such children and should be aware of the potential underlying spinal defects.

Keyword : Faun tail , occult spinal dysraphism, split cord malformation
Faun tail is a cutaneous marker of underlying spinal anomalies x-ray and MRI dorsolumbar spine (including whole spine and brain screening) was taken. The x-ray of dorsolumbar spine showed fusion of D12 and L1 vertebra. The MRI of dorsolumbar spine showed two hemi cords contained with its own dural sheath and separated by osteocartilaginous medial septum extending from D12 to L3 with osseous septum L1, L2 level (split cord malformation type-1). Osseous spur was noted at L1, L2 level. USG abdomen showed two spinal cords in the region of the tuft of hair at back and no other abnormality was detected. Blood chemistry and urine analysis were normal. We diagnosed the child as a case of faun tail with split spinal cord malformation type-1 (diastematomyelia).

Neurosurgeon opinion was obtained. Although the neurological examination of the child was normal, in order to avoid future neurological complications, prophylactic surgery (Detethering of the cord, Excision of osseous septum and bony spur) was advised by Neurosurgeon. However, the child’s family declined surgery.

DISCUSSION
Spinal dysraphism is a distinct group of congenital anomalies characterized by a failure of midline structures of ecto- and mesodermal origin to fuse. A spinal dysraphism can be broadly divided into two different pathological entities

Closed spinal dysraphism: spinal dermal sinus, lipomyelomeningocele, split cord malformations (diastematomyelia), neurenteric cysts, thickened filum terminale.

EMBRYOLOGY
The first 2 months of embryogenesis can be divided into 23 stages. Around day 18 at stage 8, the neural plate is formed, followed by neural folds and their subsequent fusion. Neurepore closure is to follow and completed by stage 12 around day 28. When caudal neurupore fails to close, open dysraphism ensues. From then until day 56, secondary neurulation sets in forming the spinal cord.

Defective secondary neurulation results in occult dysraphism in which the caudal part of the spinal cord remains connected with the epidermis by tissues of mesenchymal origin and this is the ultimate cause for tethering later on in life.

OCCULT SPINAL DYSRAPHISM
Congenital spinal defects covered by intact skin are called as occult spinal dysraphism. A midline cutaneous posterior anomaly is often a clue for an underlying occult spinal dysraphism. The cutaneous signs of spinal dysraphism are seen in 50% of cases of occult spinal dysraphism. (3) Other studies have documented an even higher prevalence of cutaneous lesions up to 76% (43-95%). (4) The cutaneous lesions which should raise higher degree of suspicion include hypertrichosis, dimples, aplasia cutis, lipoma, hemangioma, dermoid cyst or sinus, acrochordons, true tail and congenital scarring. Lesions with low index of suspicion include telangiectasia, capillary malformations (portwine stain), hyper-pigmentation, hypopigmentation, melanocytic nevi, connective tissue nevus, hypertrophic skin, hamartomas, teratoma and neurofibroma. Lumbosacral hypertrichosis (faun tail naevus) is the most common skin lesion evident at birth, as was seen in our patient. Hairy patches are most frequently associated with tethered cord and split cord malformation (diastematomyelia). (5)(2) Similar association of the hairy patch was seen in our case.

Many patients are initially asymptomatic but may become symptomatic later in life. The cutaneous markers should be evaluated with complete history and physical examination, especially in older children. History taking should include questions regarding additional congenital malformations, family history of neural tube defects, weakness or pain in the lower extremities, abnormal gait, scoliosis, difficulties with toilet training or incontinence, recurrent urinary tract infections, and recurrent meningitis. If the tethering is undetected, sensory and motor deficits, bladder and bowel dysfunction may occur. So, early untethering for secondary tethered cord syndrome is essential. Progressive foot deformities, scoliosis or kyphosis are seen in approximately one fourth of children with increasing age. When neurological deterioration starts, complete recovery may not be guaranteed even when technically successful untethering is performed. So, any patient with abnormal lumbar hypertrichosis should be investigated for occult spinal dysraphism. For demonstrating the spinal cord anomalies, MRI is the best radiological study. It is not advisable to remove the skin lesion prior to neurosurgery consultation,because there may be a second defect such as a dermal sinus and probing could lead to meningitis. Hair reduction with laser is recommended for treatment of hypertrichosis of faun tail. (6) SPLIT SPINAL CORD MALFORMATION

Split spinal cord malformations refers to a group of malformations where the spinal cord is split or clef ted over a portion of its length. Several terminologies were used in literature like ‘Diastematomyelia’, ‘Diplomyelia’ with poor distinction. Pang et al. have proposed unified theory and recoined them as ‘Split cord malformations’. The basic error appears to be development and persistence of ‘Accessory Neurenteric Canal’ (ANC). The persistence of anterior end of this canal will result in intestinal malformations and persistence of posterior end causes cutaneous malformations like angiomas, hypertrichosis, dermal

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**RADIOLOGICAL DIAGNOSIS**

Plain X-ray, CT scan, CT myelography, and MRI are the imaging modalities used to diagnose this anomaly. MRI has become the procedure of choice, to demonstrate spinal cord, dural sac, the spur, location of filum terminale and other associated abnormalities. The CT and MRI can be complimentary in increasing the diagnostic accuracy. (7)

**SURGERY**

Neurological deterioration is believed to result from the tethering phenomena. The symptoms and their manifestation may vary depending on the type and associated anomalies. The onset of neurological deficits seems to be early in type-1 malformation in comparison to type-2. Since majority become symptomatic by second decade, and eventually deteriorate in their neurological function. So Neurosurgeon recommends prophylactic surgery. Prophylactic surgery is logical as neurological deficits are not completely reversible once they occur, on the other hand the children with no neurological deficits maintain steady neurological state postoperatively. The ideal time of surgery is around 6 months of age when anaesthesia and blood loss is better tolerated. (8)

**CONCLUSION**

Cutaneous lesions have an important role in the detection and diagnosis of occult spinal dysraphism. Recognition of these stigmata followed by appropriate radiological imaging and prophylactic surgery decreases the long-term morbidity of this condition. The Paediatricians are the first physicians to see such patients and should be aware of the potential underlying spinal defects.

**REFERENCES**