Faun Tail Naevus: A Cutaneous Marker of Spinal Dysraphism

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We describe three cases (one male and two females) of faun tail nevi, which is one of the most important cutaneous marker of spinal dysraphism. One of the patients presented with acro-osteolysis leading to auto amputation of the toes of the left foot, which required operative intervention. This lays stress on the early recognition of lumbar paraspinal skin lesions and early treatment to avoid irreversible sequelae.

Key words: *Acro-osteolysis, Dysraphism, Faun tail nevus.*

Spinal dysraphism refers to incomplete fusion of midline structures of the embryonal dorsal median region that may affect any combination of somatic ectoderm, neuro-ectoderm or mesoderm(1). As both skin and nervous tissue are of ectodermal origin, anomalies of these tissues may occur simulatenously. We report three cases with faun tail nevi, a marker of spinal dysraphism.

Case Report

Case 1: A 5-year-old boy presented with two years history of spontaneous resorption of digits and painless ulceration of toes of the left foot. Parents noticed decreased sweating over the left foot and leg. There was no associated history of neurological weakness, back pain, gait abnormalities or urinary and bowel incontinence. The child was a product of nonconsanguinous marriage, born through a normal vaginal delivery and had normal developmental milestones. No other family member had similar lesions.

Cutaneous examination revealed a circumscribed area of coarse, dark, terminal hair measuring 25 × 15 mm overlying the lumbosacral area with normal underlying skin (*Fig. l*). There was swelling of left foot and resorption of distal phalanges of all the left toes, except fourth. The plantar arch of the left foot was obliterated. On palpation, spinous processes in the lumbar and sacral segments were absent. Neurological examination revealed reduced sensation to pain and touch in the distribution of L3 -S1 nerves (below knee) on the left side. Left ankle reflex was diminished and plantar reflex was equivocal. Peripheral vascular circulation was normal clinically and confirmed by a normal Doppler study.

A roentgenogram of lumbosacral spinal column showed a posterior fusion defect of

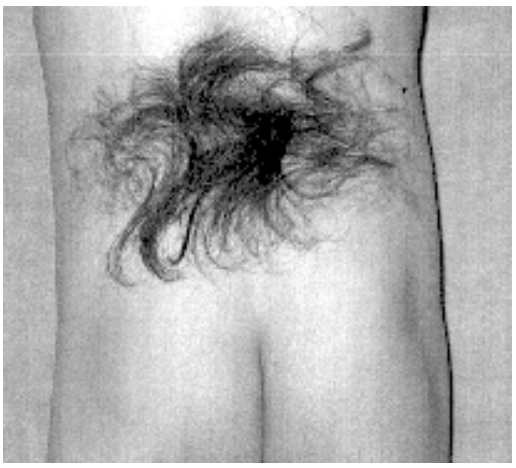

Fig. 1. Circumscribed patch of coarse, dark, terminal hair overlying lumbosacral area.

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lumbar vertebral segments (spina bifida). The MRI of lumbosacral spine showed spina bifida, hydromyelia of distal cord/cones, diastematomyelia with a complete fibro-osseous bony spur at L2 level, thickened and positively tethered filum terminale and filum lipoma. *X*-ray left foot showed resorption of proximal phalanges of 1st and 5th digits, resorption of distal and middle phalanges of 2nd digit, periosteal reaction in 2nd metatarsal, expansion and sclerosis of 1st metatarsal and soft tissue swelling, consistent with acro-osteolysis (*Fig. 2*).

A diagnosis of acro-osteolysis due to spinal dysraphism with faun tail naevus was made. Patient was subjected to L1-L3 laminectomy with excision of bony spur and detethering of the cord. Patient is on regular follow up for assessment of improvement in sensory functions.

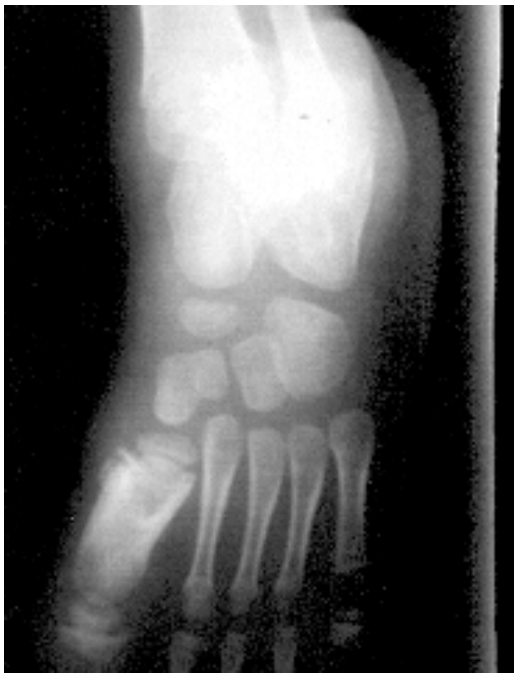

Fig. 2. X-ray left foot: resorption of terminal phalanges of all toes, except fourth. Expansion and sclerosis of first metatarsal. Periosteal reaction in second metatarsal. Soft tissue swelling.

Cases 2 & 3: Two patients (four and six years old girls) presented with tuft of dark, coarse hair over the lower back since birth and were diagnosed to have faun tail nevus. Both of them were product of normal vaginal delivery, with no history of consanguinity in parents, and did not have any other extracutaneous manifestations. *X*-ray lumbar spine of both cases showed spina bifida at the level of L5-S1 and L3-L4, respectively. MRI of spine in second case showed unfused posterior elements of L5 and S1 vertebrae. MRI of third patient did not reveal any features of occult spinal dysraphism.

Discussion

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(2). Other studies have documented an even higher prevalence of cutaneous lesions upto 76% (43-95%)(3). The cutaneous lesions which should raise higher degree of suspicion(4,5) include hypertrichosis, dimples, aplasia cutis, lipoma, hemangioma, dermoid cyst or sinus, acrochordons, true tail, pseudotail and congenital scarring. Lesions with low index of suspicion include telangiectasia, capillary malformations (portwine stain), hyperpigmentation, hypopigmentation, melanocytic nevi, connective tissue nevus, hypertrophic skin, hamartomas, teratoma and neurofibroma. Sacral hypertrichosis (faun tail naevus) is the most common skin lesion evident at birth(6), as was seen in our patients. Hairy patches are most frequently associated with tethered cord(2) and diastematomyelia(7). Similar associations of the hairy patch was seen in our first case.

Acro-osteolysis, as a manifestation of spinal dysraphism has been rarely reported in literature(8). Spinal dysraphism usually first interrupts pain and pressure fibres crossing from one side of spinal cord to the other, leading to a lack of sensation to noxious stimuli in the limbs. The sensory deficit contributes to bone resorption by facilitating repetitive mechanical injuries. Variations in the radiological appearance of acro-osteolysis, depending on the area of terminal phalanx involved can be of help in suggesting different etiologies. Resorption of tips of fungal tufts with variable involvement of shafts and bases is seen in neurological disorder(9). Similar radiological picture was seen in our first case.

MRI is accurate screening modality in the initial diagnosis of dysraphic anomalies, in patients presenting with sensory loss and lower limb anomalies(10). It has been suggested that optimal time for surgery is within the first 3-4 months of life before mechanical traction can be exerted on the conus or cauda equina. Therefore any patient with unilateral or asymmetrical acro-osteolysis in the limbs or well localized paraspinal hypertrichosis should be investigated, as it may suggest evidence of underlying spinal abnormality including spinal dysraphism. Timely intervention is necessary to prevent the occurrence or progression of the neurological deficit once the neurological damage has occurred. Other patients with no apparent neurological deficit should be sensitized for symptoms, which may appear in later life.

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