Faun Tail Nevus Associated with Myelomeningocele

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Dear Editors

Hypertrichosis is the presence of excessive growth of terminal hair on the non-androgen dependent areas of the body. A faun tail is abnormal lumbosacral hypertrichosis characterized by a patch of coarse terminal hair which is usually several inches long [1]. Faun tail nevus at the lumbar area can be a cutaneous marker for underlying spinal abnormalities like spinal dysraphism [2]. In this article the author reports an eight year old female with a tuft of terminal hair at the lower back associated with myelomeningocele.

An eight year old female presented with a history of a patch of hair and swelling on her lower back since infancy. The antenatal and birth history was unremarkable. Developmental milestones achieved were normal for her age. There was no history of urinary or fecal incontinence however she complained of lower limb weakness. There was no such history in family. She was born of a non-consanguineous marriage. On examination, a well-defined patch of terminal, dark hair of size around 7 cm × 15 cm was present on the lumbosacral region in the midline (Figure 1). At the centre of the patch, there was a swelling of size around 5 cm × 6 cm, cystic in consistency (Figure 2). The skin overlying swelling was normal with no visible veins, punctum, sinus, or pulsations. There was a large ill-defined slaty grey macular pigmentation involving lower back and upper medial quadrants of both buttocks which persisted on diascopy. No other cutaneous or mucosal abnormality was noted in the patient. Neurological examination revealed grade 4 power in muscles of lower limbs. However, no sensory deficit was elicited. Her gait was normal. MRI spine and brain showed spina bifida at L2, L3 and L4 vertebrae. A cystic swelling enclosing the spinal cord, its meninges, cerebrospinal fluid, and parts of nerve roots (Myelomeningocele) was seen protruding through the posterior spinal defect. Based on history, suggestive clinical examination and further supported by radiology, a diagnosis of faun tail with myelomeningocele and Mongolian spot was made. The patient was referred to department of Neurosurgery for further management.

Spina bifida is a birth defect characterized by the incomplete closure of the spine and membranes around the spinal cord. It is divided into three types: viz., spina bifida occulta, meningocele, and myelomeningocele. Occulta is the mildest form of spina bifida in which the outer part of some of the vertebrae have not completely fused with each other. However, the splits in the vertebrae are so small that the spinal cord does not protrude through it. The skin at the site of the lesion may be normal, or having a dimple in the skin, or a birthmark or it may have some hair growing from it. In case of posterior meningocele, a single defect in union allows the spinal meninges to herniate between the vertebral defect forming a meningeal cyst. Since the nervous system remains unaffected, patients with meningocele are unlikely to suffer long-term neurological problems. Myelomeningocele, also called as ‘open spina bifida’, is considered the most severe form. In this type, the unfused portion of the vertebral spine allows the spinal cord and covering meninges to protrude through the defect forming a sac enclosing the spinal elements, such as meninges, cerebrospinal fluid, and parts of the spinal cord and nerve roots [3]. It is usually associated with other problems like lower limb muscle weakness, bladder or bowel incontinence, hydrocephalus, tethered spinal cord, and latex allergy. Our patient also had myelomeningocele and demonstrated lower limb weakness however there was no bowel or bladder incontinence.

Spinal dysraphism refers to a group of pathologic conditions related to improper closure of the caudal neuropore and comprises all the conditions associated with spina bifida. Other cutaneous markers which can be associated include dermal sinuses, dimples, subcutaneous lipoma, aplasia cutis, port wine stain, acrochordons, hemangiomas, telangiectasia, etc. [4]. In our case, there was associated Mongolian spot, a type of dermal melanocytic nevus.

Regarding the management of hypertrichosis, acquiring a long-term reduction of unwanted hair is a challenge to the dermatologist. The various treatment modalities available for removal of excessive hair include trimming, bleaching, waxing, electrolysis, physical and chemical depilatories, intense pulsed light therapy and laser hair removal [5].

Figure 1: A patch of terminal black hair present over back (faun tail). There is associated slaty grey macular pigmentation over buttocks (Mongolian spot).
The author wants to stress upon that faun tail at the posterior midline site should be evaluated to recognize any associated neuroskeletal anomalies so that appropriate early treatment can be administered for the possible underlying anomalies before any irreversible sequelae develop in the patient.

References