Faun Tail

Sang Won Kim, M.D.

Department of Dermatology, Taegu Catholic University School of Medicine, Taegu, Korea

We report herein a rare and grotesque case of a 19-year-old male with faun tail on the lumbosacral area of the back, presented from birth. The lesion was a 10×10cm sized patch with the circumscribed tuft of coarse terminal hairs about 10 to 25 cm in length downward like a horse-tail. In the center of the patch was a 4×3cm sized brown macule in which was a 2×1cm scar. The skin biopsy from the brown macule revealed mild hyperkeratosis with basilar hyperpigmentation. Lumbar radiography showed findings of spina bifida occulta affected from L-4 to total sacral segments. There was mild saddle-hypoesthesia in the neurologic study.

(Ann Dermatol 5:(1) 47–50, 1993)

Key Words: Faun tail, Spina bifida occulta

Faun was a kind of god in ancient Roman mythology. He was half-man and half-sheep, with a ruling power over forestry and stockfarming. The term "faun tail" (FT) was derived from it, denoting a grotesque long, coarse, hairy tuft on the lumbosacral area of the back.

Congenital cutaneous defects occuring in the dorsal midline of the back, is to be found in association with various forms of spinal dysraphism by their common ectodermal origin[1,2]. The skin may be missing, or hypoplastic, but in about half of the cases, there is a dermal sinus or dimple, a lipoma, a tuft of long terminal hair, a pigmented macule, a portwine stain, a skin tag or a dermoid cyst. Spina bifida occulta may occur in as many as twenty percent of affected individuals, of whom only a small percentage will have a significant associated neurologic deficit[1].

In a review of Korean dermatologic literature[3,4], only two cases with FT have been reported in assocation with spina bifida occulta. We describe herein an interesting case of a young adult male with the grotesque FT feature and spina bifida occulta.

Received July 20, 1992
Accepted for publication August 26, 1992
Reprint request to: Sang Won Kim, M.D., Department of Dermatology, Taegu Catholic University School of Medicine, Taegu, Korea

REPORT OF A CASE

A 19-year-old male presented to the Department of Dermatology at the Catholic University Hospital, in November 1990, for the diagnosis and therapy of a hairy tuft on the lumbosacral area. The lesion consisted of a circumscribed hairy patch present since birth. The initial lesion had been a coin-sized small patch, gradually increasing in width and length, which was about 10×10cm at presentation. In the center of the patch was a 4×3cm sized brown macule in which was a 2×1cm scar with an unknown time of development. The tuft of shiny terminal hairs (the so-called FT) from the lesion had grown to about 10 to 25 cm in length downward, like a horse-tail (Fig. 1). The first visit was noted on entry to our department because of the reluctance of the patient to exposure.

Family and past medical history were noncontributory. There was no consanguinity. Physical examination revealed that he was well developed and of normal mentality. The sensation was slightly decreased along S-1, S-2, S-3 dermatomal areas, that is, saddle hypoesthesia, without the changed lower motor function or trophic ulceration on the lower legs. Routine laboratory studies were within normal limits. Simple lumbar radiography showed scoliosis with a decreased lumbar

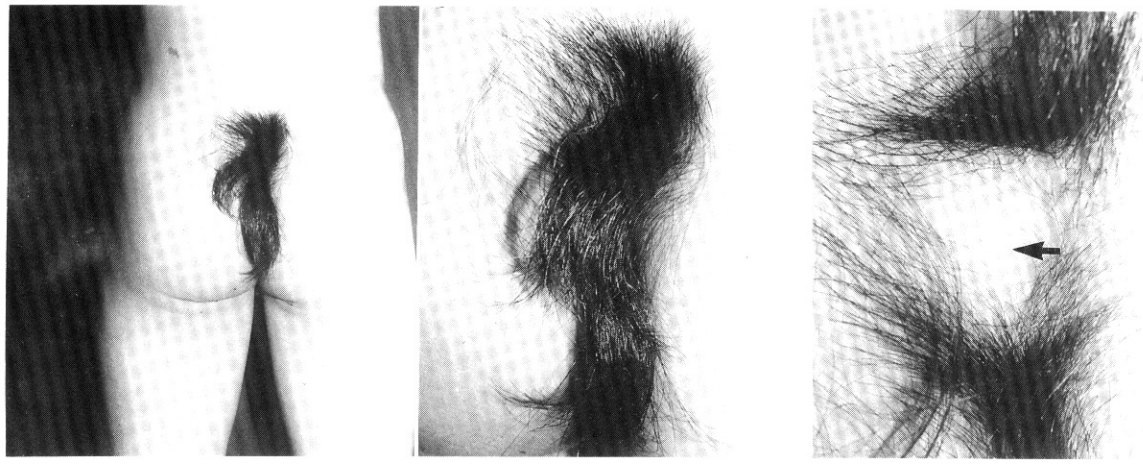

Fig. 1. A circumscribed, tufted hairy patch (left, middle) with small scar (←)in its center (right) on the lumbosacral area of back.

lordotic curve as well as findings of spina bifida occulta affected from L-4 to total sacral segments (Fig. 2). The skin biopsy from the brown macule demonstrated mild hyperkeratosis with acanthosis and basilar hyperpigmentation. Nevus cell nests or smooth muscle hyperplasia were not seen (Fig. 3). A few mature hair follicles in the dermis were transversely sectioned.

He was recommended the more accurate radiologic evaluation and therapeutic management by the departments of plastic surgery and neurosurgery, but refused any approaches including myelographic study.

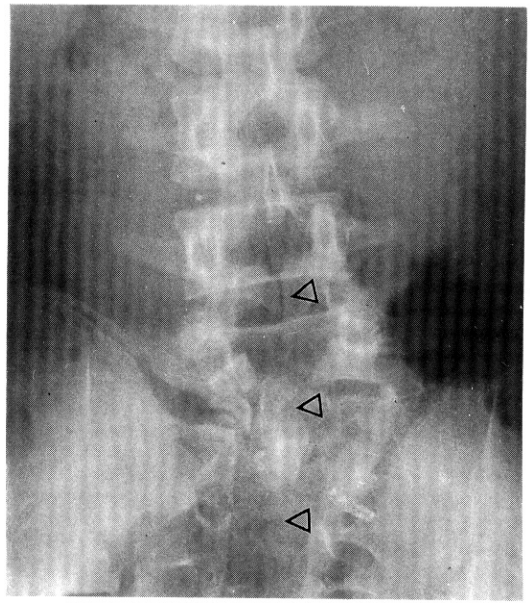

Fig. 2. Lumbar radiography shows scoliosis and defective laminae form L-4 to total sacral segments (◁).

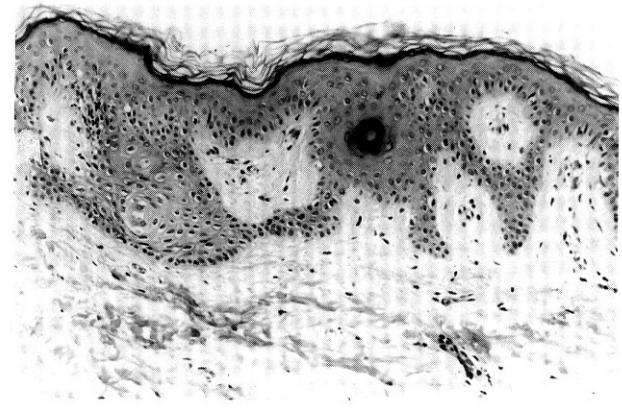

Fig. 3. Biopsy from the brown macule in the lesion shows mild hyperkeratosis and acanthosis with basilar hyperpigmentation (H & E stain, ×200).

DISCUSSION

The term spinal dysraphism was coined in 1940 by Lichenstein[5] to designate incomplete fusion or malformation of structures in the dorsal midline of back, particularly congenital abnormalities of the vertebral column and spinal cord resulting from single or multiple disturbances during embryogenesis. According to such a developmental error, the defects of the overlying skin may be associated with various lesions[1]. In a review of 200 cases with spina bifida in the literature[6,7], 102 cases (51 percent) with spina bifida are associated with single (61%) or multiple (39%) sacrococcygeal cutaneous lesions, of which there are lipomas in 41, hairy patches in 30, pigmented macules in 11, hemangiomas in 7 and bony mass-

es in 2. The dermoid and teratoma are rare[12].

Otherwise, Burrows[8] has reported meanigfully that 46 of 90 cases had an abnormal distribution of hair, frequently a discrete tuft or patch over part of the back.

In 1978, Tavafoghi et al[6] divided abnormal hairy patch into the following: "FT", appearing as a hairy patch that was minimally several centimeters but commonly much wider and lozenge-shaped, and occuring as coarse hair several inches in length in the midline; localized hypertrichosis, consisting of normal-appearing hair or slightly coarse hair over the lower part of the back; "silky down", represented by fine, soft nonterminal or lanugo hairs limited to a discrete midline area. Of these, FT is most serious and problematic.

The accompanied deficit in spina bifida varies from birth to 76 years at age of onset. No symptoms appear until adolescence or even adult life, well past the growth period in a small percentage[9]. The speed of the patient's worsening condition varies considerably and symptoms differ widely[6,9,11]. The serious deficit is not present in dimple or sinus. However, FT serves as a warning sign for the progressive deficit that may subsequently develop[6,10].

The deficit may show mild asymmetry or paresis of the legs, reduced ankle reflex, and shortening and trophic ulceration of one or both feet. Sensation may be impaired over the areas innervated by the lower several segments, leading to the saddle-shaped area of analgesia over the buttocks and posterior surface of thighs. The severest sign is that of a flaccid paraparesis with sphincter paralysis[1,9,10]. Our case demonstrated that FT, presented from birth, was diagnosed at 19 years of age, when first seen on entry. The neurologic evaluation showed saddle hypoesthesia involving S-1, S-2, S-3 dermatomes, without any other deficit or trophic changes. The formation of a small scar in the lesion was assumed to be related to the healing of a congenital defect or post-inflammatory process of dermal sinus. It was similar to Choi et al's case[3] in Korean literature.

The early recognition of such lesions needs the cooperation of the patient and the thoroughness of examination. Therefore, a careful study including a comprehensive diagnostic approach and close follow-up, is necessary[7,10,11]. Radiography shows defective fusion of the laminae in the affected region, usually the first sacral and fifth lumbar[1]. Especially, myelography is considerably reliable to afford valuable assistance when confronted with this condition[7,8]. Spina bifida occulta in our case was discovered in the simple lumbar radiography. The myelography was not done by patient's refusal.

Treatment is surgical. It's main purpose is to explore the spinal cord for prevention of neurologic deterioration[6,9,10]. Correction of the cutaneous lesion is by no means an adequate method. When dealing with a small lesion, excision can be done at any time. In a large lesion, such as an extensive hairy patch, the resulting defect from excision requires resurfacing with a skin graft[10].

Therefore, it is usually in the domain of the pediatrician and neurologist. Dermatologic aspects in this field have recieved scant attention and have not been studied in detail. Accordingly, we should be concerned about midline sacrococcygeal cutaneous lesions and the spinal association, based on the congental defect.

REFERENCES

1. Atherton AJ, Rook A: *Naevi and developmental defects. In Rook A, Wilkinson DS, Ebling FRG et al (eds): Textbook of Dermatology, 4th ed, Blackwell Scientific Publications, Oxford, 1986, pp223-225.*
2. Adams RD: *Neurocutaneous diseases. In Fitzpatrick TB, Eisen AZ, Wolff K et al (eds): Dermatology in General Medicine, 3rd ed, McGraw-Hill Book Co, New York, 1987, p2039.*
3. Choi JS, Lee WH, Lee JB: *Circumscribed hypertrichosis and blue nevus with spina bifida occulta. Kor J Dermatol 19:365-368, 1981.*
4. Suhr GB, Lee JS, Lee JH et al: *A case of pina bifida occulta with faun-tail nevus. Kor J Dermatol 26:759-763, 1988.*
5. Lichenstein BW: *"Spinal dysraphism", spina bifida and myelodysplasia. Arch Neurol Psych 44:792-809, 1940. Cited from reference 6.*
6. Tavafoghi V, Ghandchi A, Hambrick GW et al: *Cutaneous signs of spinal dysraphism. Report of a patient with a tail-like lipoma and review of 200 cases in the literature. Arch Dermatol 114:573-577, 1978.*
7. Harris TJ, Gang DL, Kleinman GM et al: *Unusual sacrococygeal embryologic malformations with cutaneous manifestations. Arch Dermatol 118:643-648, 1978.*

8. Burrows FGO: *Some aspects of occult spinal dysraphism: a study of 90 cases. Br J Radiol 41:496-507, 1968.*
9. Anderson FM: *Occult spinal dysraphism. Diagnosis and management. J Pediat 73:163-177, 1968.*
10. Eid K, Hochberg J, Saunders DE: *Skin abnormalites of the back in diastematomeylia. Plastic Reconstr Surg 63:534-539, 1979.*
11. Grysprecht GL: *Myelographic assessment of occult forms of spinal dysraphism. Acta Radiologica 1:702-717, 1963.*
12. Pantoja E, Rodriguez-Ibanez E: *Sacrococcygeal dermoids and teratomas. Historical review. Amer J Surg 132:377-383, 1976.*