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Naevus Comedonicus as Dermatologic Hallmark of Occult Spinal Dysraphism

Sir,
Naevus comedonicus is a developmental anomaly of the pilosebaceous apparatus, clinically characterized by papules and comedones arranged in bands. Spinal dysraphism refers to a group of malformations of any or all the midline tissues of the back and spine. Cutaneous manifestations are frequently associated stigmata in more than 50% of cases of occult spinal dysraphism (1). We describe a case of naevus comedonicus associated with hypertrichosis of the sacral region, which gave a hint of occult spinal dysraphism.

CASE REPORT
A 40-year-old man was evaluated for a midline cutaneous sacral hairy patch, which had been present since birth. He was taking salicylate and paracetamol for ankylosing spondylitis. Physical examination revealed hypertrichosis associated with grouped, skin-coloured and erythematous papules, some containing a central dark hyperkeratotic plug on the sacral region. A biopsy specimen showed large numbers of atrophic cystically dilated hair follicles, containing abundant lamellated keratin aligned perpendicularly to the skin. Radiologic evaluation revealed dysgenesis of both sacral joints, extensive lumbar spine syndesmophylic formation and a large skylith of the sacrum. Magnetic resonance imaging scans demonstrated a wide vertebral fusion defect of the sacrum and a hyperintense lesion in the distal portion of the filum terminale, interpreted as an intradural lipoma.

DISCUSSION
Cutaneous stigmata of occult spinal dysraphism have been variably described (2). Localized hypertrichosis overlying the spinal defect occurs in about 30% of the cases as a single or combined skin lesion (1). Naevus comedonicus has never been reported in association with dysraphic conditions, even though common congenital skeletal and central nervous system abnormalities have been described in the naevus comedonicus syndrome, a disorder related to the group of the epidermal nevus syndrome (3, 4).
In our case we believe naevus comedonicus to be not a fortuitous condition, because of the close topographic relationship between cutaneous and skeletal lesions. Naevus comedonicus is a hamartomatous lesion of the follicular infundibulum, which further supports the common origin of ecto-mesodermal anomalies of the skin, bone and nervous system of the lumbosacrococcygeal region.

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Alopecia Syphilitica in a Prepubertal Girl

Sir,
Acute hair loss is a common manifestation of secondary syphilis, but it tends to be overlooked by patients and physicians. It may be confused with alopecia areata, trichotillomania or other alopecias, especially in children. It is important to consider the possibility of alopecia syphilitica in patients with acute patchy or diffuse hair loss. Clinicians who see teenagers should routinely take a sexual history and be prepared to offer counseling and care (1).

CASE REPORT
An 11-year-old girl with no contributory medical history presented with a rapid increase in scalp hair loss for 3 months. The lesion failed
to respond to topical antibacterial or antifungal agents or topical corticosteroids. Physical examination revealed diffuse patchy alopecia of the entire scalp, without evidence of scarring (Fig 1). Vertical sections of a scalp biopsy specimen demonstrated several terminal anagen hairs, one of which had a peribulbar lymphocytic infiltrate. Only a few plasma cells were present in the infiltrate. Thyroid function test results, a complete blood cell count, dehydroepiandrosterone sulfate level, and free serum testosterone level were all normal, but the rapid plasma reagent (RPR) test was reactive. Confirmatory tests for syphilis demonstrated a Venereal Disease Research Laboratory (VDRL) titer of 1:5120 and a reactive fluorescent treponemal antibody absorbed (FTA-ABS) test result. Additional details of the patient’s history were obtained, and the patient admitted to a single, unprotected sexual encounter approximately 6 months before the onset of alopecia. The patient was successfully treated with oral penicillin 1,000 mg daily (30 mg/kg) for 4 weeks. Within 4 months there was dramatic hair regrowth, and 6 months after treatment the patient’s VDRL titer was 1/2.

**DISCUSSION**

Very little is written about alopecia syphilitica in children, but it progresses in much the same manner as in an adult case. It can be transmitted from adults to children in sexual or nonssexual ways. Transmission by intimate contact is required, usually through infected oral or genital lesions.

Hair loss does not occur in primary syphilis, except in association with a primary chancre of the scalp. Hair involvement in secondary syphilis may assume several forms. The first is an uncommon symmetric cutaneous type associated with an actual secondary papulosquamous lesion on the scalp. The second type is essential syphilitic alopecia, which results in hair loss without visible syphilitic skin lesions, including a moth-eaten alopecia, a generalized thinning of the hair, and a moth-eaten type combined with general thinning of the scalp hair (2). Moth-eaten alopecia is usually a manifestation of late secondary syphilis, but it may occur at earlier stages.

When an associated eruptive skin rash or lymphadenopathy is present, the diagnosis may readily be suggested and confirmed by a positive serologic test. When such findings are absent or overlooked, a biopsy may be obtained to differentiate from other alopecias such as trichotillomania, traction alopecia, or alopecia areata. Recently, Jordaan & Louw (3) reported the histopathological features in 12 patients with moth-eaten alopecia. The main findings were a normal epidermis; follicular plugging; a mild perifollicular and perivascular cellular infiltrate composed of lymphocytes, macrophages, plasma cells, eosinophils, mast cells, and melanophages; telogenization; and follicle-oriented melanin clumping. The Warthin-Starry stain failed to confirm the presence of Treponema pallidum in all cases.

Since moth-eaten alopecia and early lesions of alopecia areata have considerable microscopic resemblance, serologic testing for syphilis is recommended in patients with unexplained rapid hair loss or sudden worsening of a pre-existing patterned alopecia. Non-treponemal tests, including the microscopic VDRL test and the microscopic RPR test, detect IgM and IgG antibody to certain antigens. Rising titers indicate new infection, reinfection, reactivation of old infection, or treatment failure. Titers will fall with appropriate therapy. Treponemal tests such as the FTA-ABS test and the microhemagglutination assay for antibody to Treponema pallidum (TPHA) are used to confirm positive non-treponemal tests. Since 95% of syphilis in children is acquired as a result of sexual abuse, some experts recommend routine testing of all sexually abused children (4, 5).

Treatment for primary and secondary syphilis, including alopecia syphilitica, is benzathine penicillin G 50,000 U/kg intramuscularly up to a maximum dose of 2.4 million units (maximal adult dose) in one dose. Children with acquired syphilis beyond the new-born period should be examined at 3 months and again at 6 months for falling titers (1).

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