Syringocystadenoma Papilliferum, Basal Cell Carcinoma and Trichilemmoma Arising from Nevus Sebaceous of Jadassohn

Sir,

Nevus sebaceous of Jadassohn has been referred to as an organoid nevus that classically evolves through several stages and may be associated with a range of skin tumors (1), and of these, syringocystadenoma papilliferum and basal cell carcinoma have commonly been reported to occur, sometimes simultaneously, within the lesion of nevus sebaceous (1-3).

We here report a rare case of nevus sebaceous associated with syringocystadenoma papilliferum, basal cell carcinoma, and trichilemmoma, exhibiting clinically a warty appearance and histologically poor development of the sebaceous glands. This case suggests that secondary tumors arise more frequently from immature nevus sebaceous than from a mature one.

CASE REPORT

A 27-year-old Korean woman had an asymptomatic large fungating mass on the vertex of her scalp. Examination revealed two tumors arising from the same yellowish plaque, which were immediately adjacent to each other (Fig. 1). The larger one, which had developed insidiously for 10 years, had cauliflower-like papillary projections on the surface of 2 × 3 × 3 cm, and the smaller one, which had been growing over the past 2 years, had a smooth and globoid appearance of a papule, measuring 0.7 cm in diameter. In the patient’s history, a pea-sized hairless patch on the vertex of the scalp had appeared at birth and had been slightly elevated during puberty.

On the shave biopsy specimens, the large mass showed marked papillomatosis and acanthosis in the epidermis and clear and pale tumor islands in the dermis connected with broad bands anastomosing to the epidermis. Multiple ductal and glandular structures were also seen in the dermal stroma. The tumor consisted of ductal-like structures, showing the characteristic findings of syringocystadenoma papilliferum, which had papillary projections covered by two layers of epithelial cells with numerous stromal plasma cells. A large pale tumor component extended downward into the dermis by the broad bands, anastomosing with the overlying epidermis, and the tumor cells were mainly composed of clear cells strongly reactive to periodic acid-Schiff stain (figure not shown), which was consistent with the findings of trichilemmoma. However, the adjacent globoid papule was a basal cell carcinoma which was made up of multiple lobules of basoid cells arranged in solid areas. The pulsating arrangement of basoid cells in the periphery of the tumor and peritumoral lacuna was also detected in this section (Fig. 2).

The tumor mass and base were treated with electrodesication and curettage following the shave biopsy, and no recurrence was observed for 6 months.

DISCUSSION

Although nevus sebaceous shows the characteristic features of increased numbers of mature sebaceous glands and hair follicles on histological examination, Mehrregan & Pinkus (1) and Wilson Jones & Heyl (2) reported cases with atypical lesions of warty surfaces and with the immature lesions of diminished sebaceous glands histopathologically, and they suggested the possibility of variations in the potential for secondary tumor development between typical and warty nevi, or histologically mature and immature nevus sebaceous (3). Our case with multiple secondary tumors also exhibited a clinically warty appearance and histologically poor development of sebaceous glands.

Nevus sebaceous has been considered a premalignant lesion, with an incidence of development of carcinomas of 10 to 30%. Most have been basal cell carcinomas, but squamous cell carcinoma and sebaceous or apocrine carcinoma have been occasionally reported (3, 4). Domingo & Helwig (4) reported that 9 cases of nevus sebaceous had malignant neoplasms, including apocrine carcinoma, adenocarcinoma, and squamous carcinoma, and 4 of them showed trichilemmal proliferation in the histologic examination.

Our rare case supports the concept of nevus sebaceous as organoid nevus with diverse differentiations of apocrine, primordial epithelium, and outer root sheath of hair, respectively.

Fig. 1. Large fungating mass and small globoid papule (arrowheads) on the vertex of the scalp.

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Fig. 2. (A) The large tumor shows marked papillomatosis and acanthosis in the epidermis and the portions of syringocystadenoma papilliferum (arrowhead) and trichilemmoma (arrowheads). (B) The histopathologic findings of basal cell carcinoma from the specimen of the small tumor (arrowheads in Fig. 1). (C, D) Characteristic findings of syringocystadenoma papilliferum and trichilemmoma are seen in serial sections, respectively (hematoxylin-eosin stain; A and B: ×20, C: ×100, D: ×40).
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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 salisanopyrine and paracetamol for ankylosing spondylitis. Physical examination revealed localized 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 ankylosis of both sacroiliac joints, extensive lumbrosacral syndesmophytic formation and a large akyphosis 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 variously 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 lumbo-sacrococcygeal 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