Ind J Dermatol Venereol Leprol 1994; 60 : 340-2

CASE REPORTS

SYRINGOCYSTADENOMA PAPILLIFERUM ARISING IN A NAEVUS SEBACEOUS

RM Govalkar*, Gamal Rashed**

A 13-year old girl with pre-existing alopecic plaque of naevus sebaceous over the scalp showed secondary development of syringocystadenoma papilliferum at around puberty, which was confirmed by histopathological study.

Key words : Naevus sebaceous of Jadassohn, Syringocystadenoma papilliferum

Introduction

Syringocystadenoma papilliferum is a sweat gland tumour and occurs most often with naevus sebaceous.[1] Naevus sebaceous of Jadassohn is usually present at birth or may become apparent later in life and around puberty becomes verrucous and nodular. In subsequent stages of development several neoplasms and hamartomas may develop secondarily within naevus sebaceous, the commonest being the syringocystadenoma papilliferum.[2,3] It may be difficult to distinguish both these hamartomas clinically and a biopsy is usually required for diagnosis. A case of syringocystadenoma papilliferum developing secondarily in a pre-existing naevus sebaceous of scalp is reported herein.

Case Report

A 13-year old girl presented with a slowly growing reddish nodule of six months duration developing in a hairless lesion over the scalp existing since birth. She desired removal of this lesion as it used to bleed while combing the hair. Examination revealed a yellowish-brown alopecic plaque measuring 6 x 4 cm, over the left parietal area of the scalp. Near the anterior margin and within the plaque a rose-red, smooth, shiny, dome shaped nodule of 1 cm size (Fig. 1) was noted.

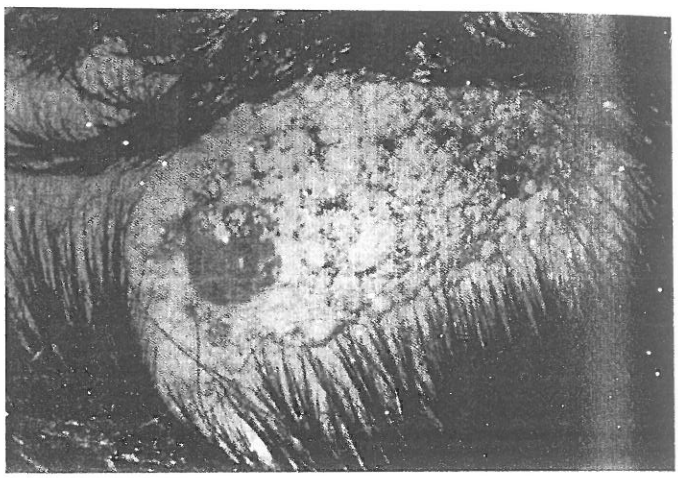

Fig. 1. Syringocystadenoma papilliferum arising within the alopecic plaque of naevus sebaceous over the scalp.

Haemorrhagic crusting was present over the posterior part of the alopecic plaque. Rest of the physical examination revealed no abnormal findings. A clinical diagnosis of naevus sebaceous and / or syringocystadenoma papilliferum was made. A wedge biopsy of the newly developed nodule along with adjacent alopecic plaque was submitted for histopathological study. The light microscopy of the nodule showed multiloculated cystic tumour confined to the

From the Department of Dermatology* (Hail General Hospital) and pathology** (King Khalid Hospital), Hail, Saudi Arabia.

Address correspondence to : Dr Ramesh M. Govalkar, P.O. Box : 2154, Hail, Kingdom of Saudi Arabia.

upper and mid-dermis. The cystic cavities were extending downward from the epidermis (Fig. 2) and were lined by double layer of cells,

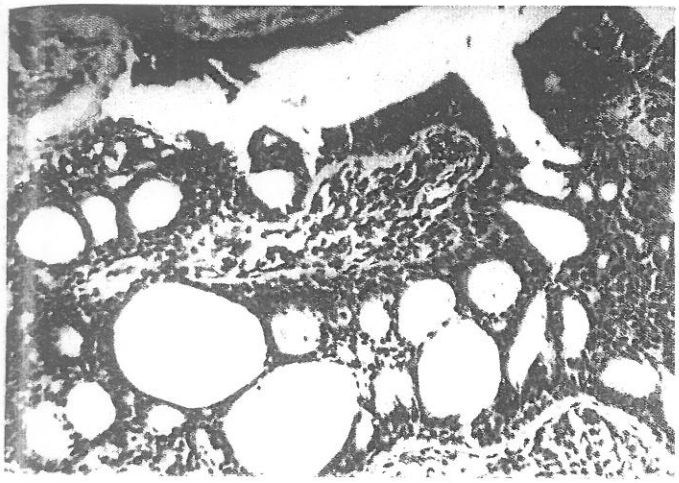

Fig. 2. Photomicrograph showing papillary projections lined by luminal layer of columnar cells and outer layer of cuboidal cells, and a marked plasma cell infiltrate in the stroma of tumour. H & E, X40.

the luminal layer of columnar cells and the outer layer of cuboidal cells. A dense plasma cell infiltrate was present in the stroma of the tumor. Deep dermis beneath the cystic invaginations showed a group of apocrine glands. The histopathology (Fig. 3) of the adjacent alopecic plaque was consistent with the clinical diagnosis of naevus sebaceous.

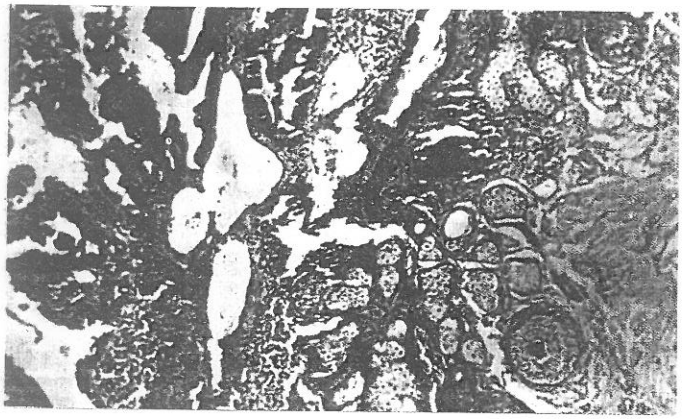

Fig. 3. Photomicrograph showing cystic invaginations and numerous mature sebaceous glands in the upper dermis. H & E, X25.

Comment

Syringocystadenoma papilliferum is a sweat gland tumour. The primary lesion may be present at birth, appear first time in early infancy or at puberty as a solitary plaque or one to several papules.[1,2] The plaques are usually skin coloured or dark brown, flat and smooth, and are devoid of hair when present over the scalp. During adolescence the lesion may become raised with verrucous or papillomatous surface.[2] Usually it is asymptomatic but patients having lesions over the scalp may complain of mild irritation or bleeding while combing the hair or may be tender to even light palpation.[4] Syringocystadenoma papillferum has been associated with several hamartomas or neoplasms. Most often it is seen in association with contiguous naevus sebaceous and at times presents as a single tumour along with naevus sebaceous.[5,6] It may be very difficult to distinguish both these conditions clinically and a biopsy is usually required for diagnosis. Our patient was an adolescent girl with naevus sebaceous of scalp existing since birth. At around puberty syringocystadenoma papilliferum developed secondarily and presented as a single tumour along with naevus sebaceous which was proved by histopathological study (Fig. 3). The tumour itself is free of pilosebaceous units, but dense enlarged sebaceous glands may be seen adjacent to or beneath the\lesion.[3] Although 90% of the cases are observed in anatomic sites normally devoid of apocrine glands, normal or widened apocrine glands may be seen either adjacent to or underlying the tumour.

It is assumed that the benign appendageal tumours present at birth are derived from primary epithelial germ cells and those appearing later in life arise from

pluripotential germinative cells in the epidermis and appendages, either same or unrelated.[7] Syringocystadenoma papilliferum is believed to be derived from apocrine and / or eccrine glands or developed from pluripotential cells in the adult epidermis and appendages stimulated by trauma or by unknown factors.[2] Alternatively, some investigators have suggested that this tumour is derived from the recently described hybrid type of apoeccrine glands.[4]

Before puberty the sebaceous and apocrine glands in the dermis are sparse and underdeveloped and at around puberty these glands try to proliferate. In our patient the proliferating cells of sebaceous or apocrine glands in the pre-existing naevus sebaceous stimulated by repeated trauma while combing the hair might have acted as a multipotential germinative cells giving rise to development of syringocystadenoma papilliferum.

References

1. Atherton DJ. Naevi and other developmental defects. In : Text book of Dermatology. 5th ed. Oxford: Blackwell Scientific Publications, 1992; 456-59.
2. Pinkus H. Life history of naevus syringocystadenomatosus papilliferus. Arch Dermatol Syphilol 1954; 69: 305-22.
3. Helwig EB, Hackney VC. Syringocystadenoma papilliferum lesions with and without naevus sebeceous. Arch Dermatol 1955; 71: 361-72.
4. Mammino JJ, Dennis AV. Syringocystadenoma papilliferum. Int J Dermatol 1991; 30: 763-66 and 828.
5. Greer KE, Bishop GF, Ober WC. Naevus sebaceous and syringocystadenoma papilliferum. Arch Dermatol 1976; 112: 206-8.
6. Grund JL. Syringocystadenoma papilliferum and naevus sebaceous occurring as a single tumor. Arch Dermatol Syphilol 1952; 65: 340-7.
7. Pinkus H. Premalignant fibroepithelial tumors of the skin. Arch Dermatol Syph 1953; 67: 598-615.