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Case Report

Mixed skin adnexal tumour arising from naevus sebaceus: a woman presented with a scalp nodule

皮脂腺痣上發生混合性皮膚附件瘤：女患者頭皮上的小瘤

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This is a case report of a woman who presented with a yellowish scalp nodule. Skin biopsy revealed mixed skin adnexal tumour with components of trichoblastoma, syringocystadenoma papilliferum and sebaceous neoplasm that might arise from a naevus sebaceus. It is an uncommon finding to have different tumours arising from the same naevus sebaceus.

本例報告一女患者頭皮呈現一黃色小瘤。皮膚活檢鑒定為混合性皮膚附件瘤，內含毛母細胞瘤，乳頭狀汗管腺瘤及皮脂腺腫瘤。於同一皮脂腺痣上發生不同腫瘤並不常見。

Keywords: Naevus sebaceus, Syrinocystadenoma papilliferum, Trichoblastoma

關鍵詞：皮脂腺痣，乳頭狀汗管腺瘤，毛母細胞瘤

Introduction

Naevus Sebaceus (NS) is a circumscribed hamartomatous lesion presents at birth. Various adnexal tumours may develop within NS. The following is a case report of a scalp tumour with components of trichoblastoma, syringocystadenoma papilliferum and sebaceous neoplasm. The underlying NS might have been completely replaced by the tumour.

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Case report

A 47-year-old woman presented with a yellowish scalp nodule that had developed from a pre-existing plaque noted at birth. The scalp lesion became painful for a few months before consultation. There was no bleeding or discharge from the lesion. Her past health was unremarkable.

On physical examination, there was a 1 x 0.5 cm yellowish tumour over the vertex of the scalp

(Figure 1). There was no satellite lesion or lymphadenopathy. The differential diagnoses include naevus sebaceus and other skin appendageal tumour.

An excisional biopsy was performed which showed a skin tumour with different components. The major components was composed of lobules of basaloid cells proliferation in the upper dermis, in the form of small islands and closely packed nests. Small foci of keratinization were present occasionally. No artefactual cleft between tumour and the stroma was noted. Adjacent to this component, there were papillary structures with continuation of the epidermis. The papillae were lined by double-layered columnar epithelium. In the invagination between the papillae, small sweat ducts were found. The stroma of papillae contained abundant plasma cells (Figure 2). Another small tumour was found in the upper dermis and composed of a few small islands of basaloid cells with aggregates of sebocytes inside (Figure 3). In addition to the tumour, the lower dermis showed clusters of apocrine glands. These

Figure 1. A 1 x 0.5 cm yellowish tumour over the vertex of the scalp.

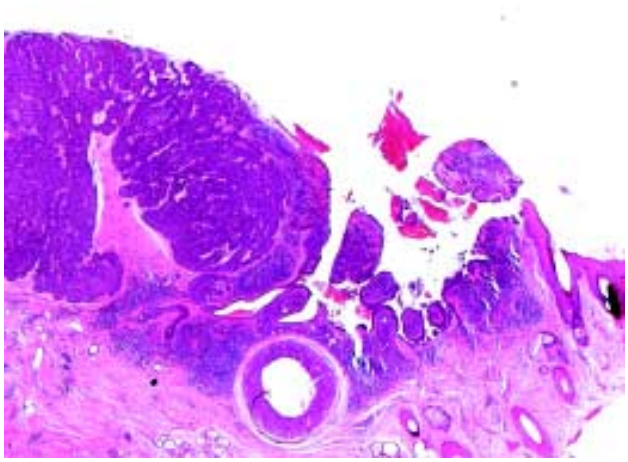

Figure 2. The trichoblastoma component (left) and syringocystoadenoma papilliferum component (right) of the tumour (H&E original magnification x 5).

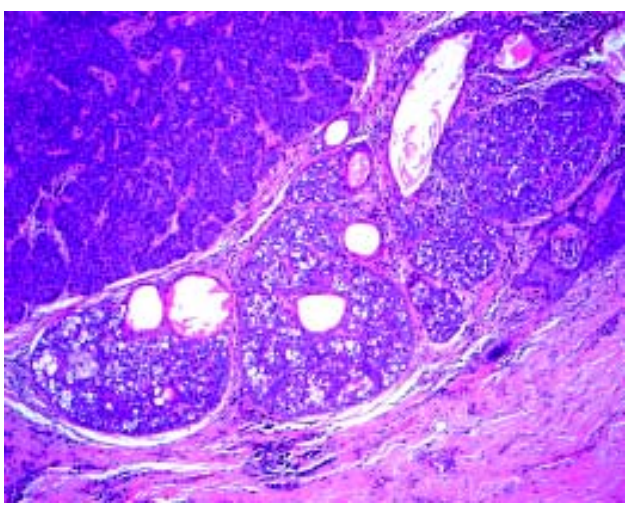

Figure 3. The benign sebaceous tumour component (below) and the trichoblastoma component (above) of the tumour (H&E original magnification x 10).

histological features were compatible with a mixed skin adnexual tumour with components of trichoblastoma, syringocystadenoma papilliferum and sebaceous neoplasm. The clusters of apocrine glands in the lower dermis might represent the residual of a pre-existing naevus sebaceus which was completely replaced by the tumour.

Discussion

Trichoblastoma was first introduced by Headington in 1970.[1] It is classified as benign neoplasm of follicular germinative cells. It usually presents as asymptomatic, solitary, non-ulcerated, skin-colour to brown or bluish black nodule of one to two centimetres in size. There is no gender predilection. It occurs mostly on scalp or face in adults with pre-existing NS as in our case. It is frequently suspected to be basal cell carcinoma (BCC) though they can co-exist. Histologically, trichoblastoma consists of aggregations of basaloid cells surrounded by variable sclerotic or partly hyalinised stroma. The basaloid cells are monomorphous with dark stainig nuclei, large prominent nucleoli and scanty pale cytoplasm. A few mitotic figures and single necrotic cell may be seen.

Syringocystadenoma papilliferum either develops independently or arises in association with NS. It commonly develops in NS on scalp or face around time of puberty. About 25% arise on trunk or genital area during adolescence or adulthood without pre-existing lesion. Clinically, the lesion presents as rose-red papules of firm consistency. Vesicle-like inclusions may be seen and some papules may be umbilicated. Small fistula discharging clear, bloody or malodorous fluid may be seen on this lesion. The histopathology of syringocystadenoma papilliferum consists of dilated and cystic changes in sweat glands and ducts with numerous papillary projections extending into the lumina. Papillary projections are lined by glandular epithelium consisting of two rows of cells. The tumour stains positively for carcinoembryonic antigen (CEA) and its transition to carcinoma is rare. Syringocystadenoma papilliferum associated with NS sometimes co-exist with trichoblastoma as described in this case.

Naevus sebaceus is a circumscribed hamartomatous lesion arises secondary to disordered development of epithelial, pilar, sebaceous and apocrine structures. The pathogenesis is unknown but the lesion involves both epithelial and non-epithelial elements reflecting malformation of both ectoderm and mesoderm. It usually presents as a solitary patch, slightly elevated pinkish, yellowish or orange to tan plaque with alopecia. It has no sex or racial predilection and is usually found at birth on scalp.

Complications of NS include the development of secondary benign and malignant tumours. Early reports indicate higher risk of BCC ranging from 6.5% to 50%.[2-4] However, two recent studies found that the incidence of BCC was low at around 0.8%.[5,6] Most lesions previously diagnosed as BCC were most likely trichoblastoma. In a study of a series of 155 samples of NS, a total of 33 neoplasms were found in 30 patients. No authentic BCC was found.[5] The most common neoplasm was trichoblastoma which accounted for 36.6% of all neoplasm found. Syringocystadenoma papilliferum accounted for another 30.3%. Another study of 596 cases of NS with differentiation of BCC from trichoblastoma by silhouette analysis and examination of stroma showed only 5 cases (0.8%) of BCC.[6] Benign tumours were found in less than 2% of NS excised in children and no BCC was found in children specimens. Trichoblastoma and syringocystadenoma papilliferum were most commonly encountered. Almost all trichoblastoma had been classified as BCC initially. Since the rate of malignant tumours in

NS was very low based on recent studies, the classical recommendation to excise small NS during childhood prophylactically for risk of malignant change is probably of uncertain benefit. Careful clinical follow up is recommended for NS in children.

The development of mixed tumour in NS is known but rare.[7,8] In most cases, these lesions represent the association of neoplasm with follicular differentiation (i.e. trichoblastoma or BCC) and with proliferations exhibiting apocrine differentiation (e.g. syringocystadenoma papilliferum). In our case, the combination of trichoblastoma, syringocystadenoma papilliferum and sebaceous neoplasm is peculiar. Clinically in view of a yellowish scalp plaque since birth and histologically the presence of ectopic apocrine glands, we believed that the mixed adnexal tumour might have arisen from a pre-existing NS.

References

1. Headington JT. Differentiating neoplasms of hair germ. J Clin Pathol 1970;23:464-71.
2. Mehregan AH, Pinkus H. Life history of organoid nevi. Special reference to nevus sebaceous of Jadassohn. Arch Dermatol 1965;91:574-88.
3. Jones EW, Heyl T. Naevus sebaceus. A report of 140 cases with special regard to the development of secondary malignant tumours. Br J Dermatol 1970; 82:99-117.
4. Morioka S. The natural history of nevus sebaceus. J Cutan Pathol 1985;12(3-4):200-13.
5. Jaqueti G, Requena L, Sanchez Yus E. Trichoblastoma is the most common neoplasm developed in nevus sebaceus of Jadassohn: a clinicopathologic study of a series of 155 cases. Am J Dermatopathol 2000;22: 108-18.
6. Cribier B, Scrivener Y, Grosshans E. Tumors arising in nevus sebaceus: A study of 596 cases. J Am Acad Dermatol 2000;42(2 Pt 1):263-8.
7. Lillis PJ, Ceilley RI. Multiple tumors arising in nevus sebaceus. Cutis 1979;23:310-4.
8. Kaddu S, Schappi H, Kerl H, Soyer HP. Trichoblastoma and sebaceoma in nevus sebaceus. Am J Dermatopathol 1999;21:552-6.