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Syringocystadenoma papilliferum: report of first case on the pinna

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Abstract

Syringocystadenoma papilliferum is a benign adnexal skin tumour, which, in a third of cases, arises from an organoid nevus on the head and neck. We report on a 17-year-old man with a syringocystadenoma papilliferum on his right pinna of three-years duration. The clinical and histopathological features are described. Following excision and skin grafting, the patient remains asymptomatic four years after surgery. This is the first report of a syringocystadenoma papilliferum on the pinna.

Key words: Adnexal and Skin Appendage Neoplasms; Cysadenoma; External Ear

Introduction

Syringocystadenoma papilliferum is a slow growing benign tumour of epithelial appendages showing differentiation in an apocrine pattern that most often appears on the scalp or on the face.[1] However, in one-quarter of cases, it is seen elsewhere.[2] It is usually first noted at birth or in early childhood and consists of either one papule or several papules in a linear arrangement[3] or a solitary plaque. The lesion increases in size at puberty, becoming papillimatous and often crusted. Histologically the epidermal surface shows papillomatosis and from these areas cystic invaginations are seen. The cystic structures are lined by papillae that have a lining of a double layer of columnar epithelium, which shows an apocrine pattern of secretion.[4,5] No case of syringocystadenoma of the pinna has been reported in world literature. In this study we report the first case of a syringocystadenoma papilliferum of the pinna and also describe the histopathology and immunohistochemical staining.

Case report

A17-year-old male patient presented to us with a papillomatous growth over the lateral surface of the pinna of three years duration. The tumour gradually increased in size, was composed of multiple papules, some of which were translucent and pigmented. There was no pre-existing nevus sebaceous like lesion. There was crusting but no bleeding from the lesion. The tumour was present on the lateral surface of the right pinna, 4 × 3 cm in size with a well-defined margin (Figure 1). A pre-operative biopsy confirmed the diagnosis. The tumour was excised into and the underlying cartilage was free of tumour. The raw area was grafted by a partial thickness skin graft and a specimen was sent for histopathology examination.

Histopathology

The epidermis showed a varying degree of hyperkeratosis, parakeratosis and papillomatosis and several cystic invaginations extend downward from the epidermis (Figure 2). In a lower portion of the cystic invagination, numerous papillary projections extend into the invaginations. The papillary projections and lower portion of the invaginations were lined by glandular epithelium with an apocrine pattern of secretion (Figure 3). Two distinct epithelial cell layers consisting of outer small cuboidal cells and inner columnar cells were present and the underlying stroma was rich in plasma cells (Figure 3). It showed diffuse positive reactivity for gross cystic disease fluid protein (GCDFP-15) on immunohistochemical staining. The patient attends for regular follow up and is asymptomatic four years after surgery.

Discussion

The clinical differential diagnosis of the skin tumour in this case included papilloma, fibroma, basal cell carcinoma, squamous cell carcinoma and malignant melanoma.[6] All were discarded on histologic examination. Rather the typical architecture, lining epithelium and infiltration of underlying stroma by plasma cells[7] indicated that this was a syringocystadenoma papilliferum. The plasma cell's infiltration of the stroma especially in the papillary projections is a highly diagnostic feature of syringocystadenoma papilliferum. These plasma cells are predominately of IgG and IgA classes.[8,9] Frequently, there are malformed sebaceous glands and hair structures in the lesions of syringocystadenoma papilliferum[10] although these were not present in our case. The majority of tumours are seen on the face and scalp of young adults. Our review of the literature uncovered 161 cases of syringocystadenoma papilliferum; 117 (72.7 per cent) were on the head and neck, 34 (21.1 per cent) on the trunk, 1 (0.6 per cent) on the upper arm, and 9 (5.6 per cent) on lower limbs[11–27] (See Table I). It is usually first noticed at birth or in early childhood and consists of either one papule or several papules in a linear arrangement[3] or a solitary plaque. The lesion

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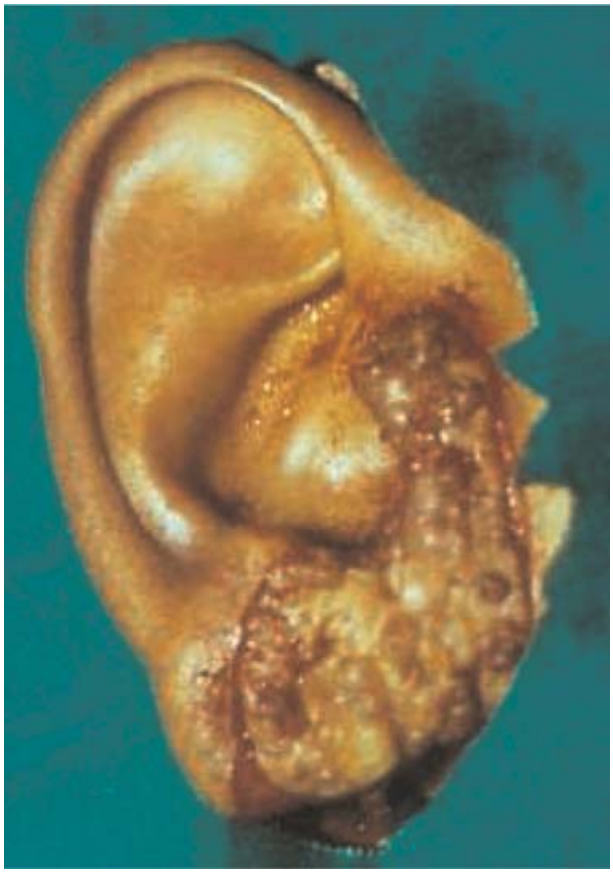

FIG. 1
Operative photograph showing the lesion.

increases in size at puberty, becoming papillomatous and often crusted.[10] On the scalp, a syringocystadenoma papilliferum frequently arises at puberty within a nevus sebaceous that has been present at birth. In about one third of the cases, syringocystadenoma papilliferum is associated with nevus sebaceous. In our case the patient noticed the tumour at 14 years of age and there had been no pre-existing lesion at the site. In about 10 per cent of the cases a basal cell carcinoma develops but only in the context of a nevus sebaceous.[2] A few instances of the transition of a syringocystadenoma papilliferum into an syringocystadenocarcinoma papilliferum with regional lymph node metastasis have been reported.[28,29]

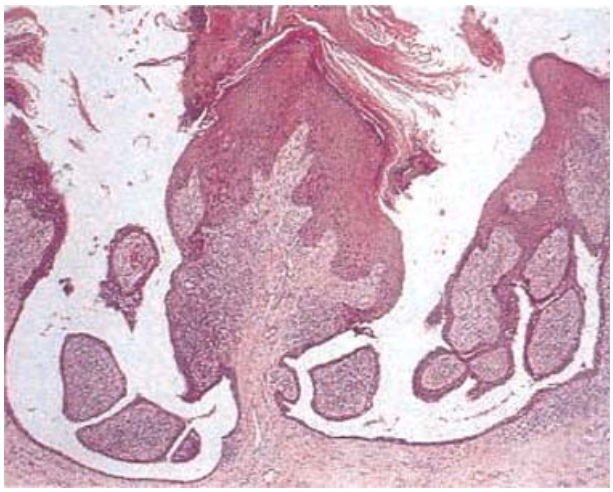

FIG. 2
Cystic invaginations extend downward from the epidermis.

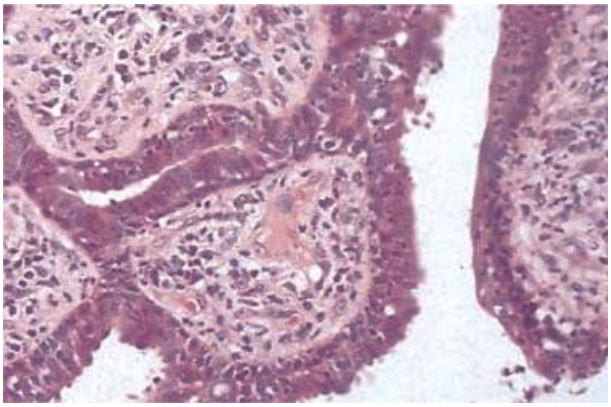

FIG. 3
The papillary projections lined by two rows of cells. The stroma of the papillae contains plasma cells.

TABLE I
DISTRIBUTION OF SYRINGOCYSTADENOMA PAPILLIFERUM

Site	Number of cases
Head and neck	117
Trunk	34
Lower limb	9
Upper limb	1
Pinna*	1
Total cases	162

* = our case.

Histogenesis

There is no unanimity about the direction of differentiation in syringocystadenoma papilliferum. Features of both apocrine and eccrine differerentiation can be seen in syringocystadenoma papilliferum. Positive immunoreactivity for gross cystic disease fluid protein (GCDFP-15) supports an apocrine genesis[29] in our case. On other hand, light and electron microscopic features of some lesions show evidence of eccrine differentiation.[30] It is likely that, rather than arising from mature structures; syringocystadenoma papilliferum arises from pluripotential germ structures of a variety of different types. In conclusion, the view expressed by Pinkus probably is correct: although most of syringocystadenoma papilliferum are apocrine in differentiation, some are eccrine.[10]

- **This paper reports the first case of a syringocystadenoma of the pinna**
- **It is a slow growing, benign tumour that produces symptoms mainly due to its cosmetic effect**
- **Surgical excision is the treatment of choice**

Our case was treated by excision and skin grafting of the raw surface. In a patient with a syringocystadenoma papilliferum we recommend surgical excision both to avoid the potential for development of secondary malignant tumours

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and for cosmetic reasons. Histopathological examination of the excised specimen is required to rule out underlying malignancy.

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