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ONLINE CASE REPORT

Syringocystadenoma Papilliferum of the Upper Lip

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ورم الغدد العرقية الحليمي في الشفة العليا

حمدان الحبسي، مصطفى الهنائي، أحمد الوائلي، سالم السديري، فيبولا دي سيلفا

ABSTRACT: Syringocystadenoma papilliferum (SCAP) is a rare skin tumour believed to arise from the apocrine or eccrine sweat glands. It appears predominantly in childhood, usually at birth. It is exceedingly rare for it to appear on the upper lip. We report a case of SCAP in a 10-year-old Omani girl who presented to the Sultan Qaboos University Hospital in Muscat, Oman, in February 2012 with a non-tender, non-pruritic, solitary verrucous papule of 4 x 5 mm on the left side of the upper lip. It had been present since birth and had slowly been increasing in size over the years. It was occasionally associated with recurrent ulceration and bleeding and had previously been misdiagnosed and mismanaged. An excisional biopsy was performed and the whole lesion was removed. The surgical site was then sutured and the patient was discharged on the same day.

Keywords: Skin Neoplasms; Sweat Glands; Lip; Misdiagnoses; Basal Cell Carcinoma; Case Report; Oman.

الملخص: ورم الغدد العرقية الحليمي ورم جلدي نادر يصيب الغدد العرقية المفترزة والفارزة في الجلد. غالبا ما يظهر في الطفولة عادة عند الولادة. نادرا جدا ما يظهر هذا الورم على الشفاه العليا. في هذا المقال نقدم تقريرا عن طفلة عمانية تبلغ من العمر 10 سنوات زارت مستشفى جامعة السلطان قابوس في مسقط، عمان في فبراير 2012 وعليها ورم ثؤلولي منفرد (5 × 4 مم) ، غير مؤلم وبغير حكة على الجانب الأيسر من الشفة العليا كان قد ظهر منذ الولادة. ازداد حجم الورم ببطء على مدى السنين مع تقرح ونزيف متكرر. لم تكن محاولات تشخيصه وعلاجه السابقة ناجحة. تم إجراء خزعة استئصالية وتمت إزالة التقرح كله. تم تخييط الموقع الجراحي وخرجت المريضة في اليوم نفسه.

مفتاح الكلمات: الأورام الجلدية؛ غدد العرق؛ الشفة العليا؛ التشخيص الخاطئ؛ سرطان الخلايا القاعدية؛ تقرير حالة؛ عمان.

SYRINGOCYSTADENOMA PAPILLIFERUM (SCAP) is a rare benign skin tumour believed to originate from the pluripotent cells and its histology exhibits either apocrine or eccrine sweat gland differentiation.[1] The tumour appears predominantly in childhood; it appears at birth in approximately 50% of those affected and before puberty in a further 15–30%.[2–4] However, in some cases it may appear at a later age.[5] The tumour is more common in women than men.[6]

The clinical presentation of SCAP is not distinctive and may often be misleading—it can present as a solitary patch, papule, plaque, nodule or as several papules often arranged linearly.[7–9] The plaque type usually presents as a hairless area on the scalp, while the linear type appears on the face and neck region and the solitary nodular type shows a predilection for the trunk.[8,9] The colour of the tumour varies from flesh-coloured to pink, red, brown or grey.[6,10] Its surface may be smooth, hyperkeratotic, verrucous, papillary or moist and fleshy. The tumour tends to grow slowly and is usually less than 4 cm in size.[4]

While the clinical features of SCAP vary widely, its histology is invariably uniform and confirmatory.[7] Although the lesion is benign, transformation to basal cell carcinoma, metastatic adenocarcinoma or ductal carcinoma may occur.[5,8,11] An unusual case of this entity occurring on the upper lip is documented below. To date, only three cases involving the upper lip have been reported in the literature.[5]

Case Report

A 10-year-old girl presented to the Sultan Qaboos University Hospital in Muscat, Oman, in February 2012 with a non-tender, non-pruritic, solitary verrucous papule on the left side of the upper lip. The papule had been present since birth and was associated with recurrent ulceration and bleeding. It had been slowly increasing in size over time. She had consulted many different physicians but no definite diagnosis had been made. Numerous treatments had been attempted without success, including scraping the lesion on a number of occasions and the application of various topical medications, including traditional medicinal treatments. There was no other significant past medical history.

A physical examination revealed a solitary painless raised papule of 4 x 5 mm on the left side of the upper lip, with an area of surrounding blood clots [Figure 1].

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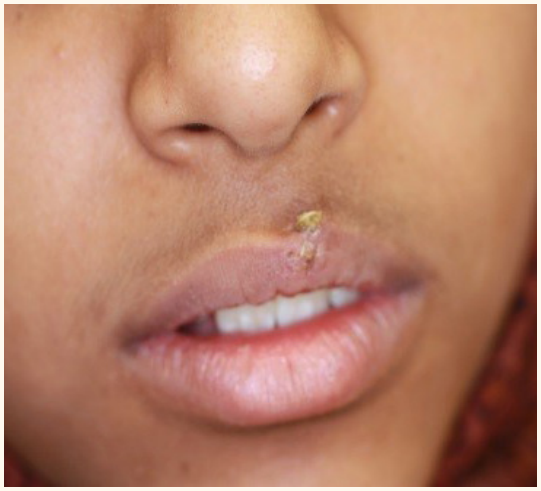

Figure 1: Photograph of a non-tender, non-pruritic solitary verrucous papule on the left side of the upper lip of a 10-year-old girl. This papule had been present since birth.

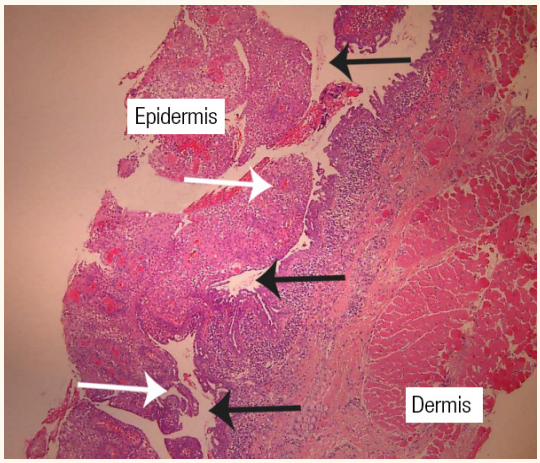

Figure 2: Histopathology of the skin lesion showing the cystic invagination of the epidermis with papillary projections (black arrows) and duct-like structures (white arrows). The specimen was processed using Hematoxylin and Eosin stain at x5 magnification.

No other abnormalities were detected around the lesion or on any other parts of the body.

An initial diagnosis of a granuloma was made. An excisional biopsy was performed and the whole lesion was removed and sent for a histopathology examination. The defect was then closed with a suture and the patient was discharged on the same day.

The pathological findings revealed a gross specimen measuring 15 x 5 x 3 mm, consisting of multiple fragments of soft tissue. The specimen was processed using a Haematoxylin and Eosin stain. Microscopically, a biopsy determined that the specimen was comprised of epidermal, dermal and skeletal muscle cells. The epidermis was hyperplastic, showing cystic invagination into the dermis, with infiltration of the lymphocytes and neutrophils [Figure 2]. In the lower part of the epidermis, a lesion was observed. The lesion was composed of a few papillary projections and ducts/gland-like structures lined by luminal and cuboidal cells. The papillary projections were lined by double epithelial cells with an abundance of plasma cell infiltrates in their cores [Figures 3 and 4]. No *atypia* or mitosis was seen. The overall architecture of the lesion revealed an invagination from the epidermis with overlying squamous hyperplasia, papillary projections lined by double epithelial cells and infiltration of the plasma cells. This morphology was consistent with a diagnosis of SCAP.

Discussion

SCAP is a rare benign tumour of the skin, usually seen in patients at birth or during infancy or early childhood. Most lesions are not distinctive and require a biopsy for the diagnosis; however, the tumour is usually described as ranging from skin-coloured to pink, and taking the form of either a hairless firm plaque, grouped nodules or a solitary nodule.[12]

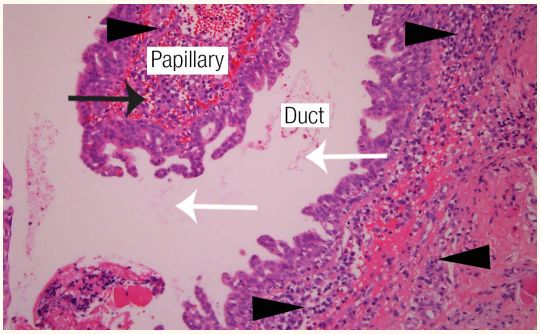

Figure 3: Duct-like structures (white arrows) and papillary projections (black arrows) lined by double epithelial cells can be observed. Plasma cells are visible in the cores (arrow heads). The specimen was processed using hematoxylin and eosin stain at x20 magnification.

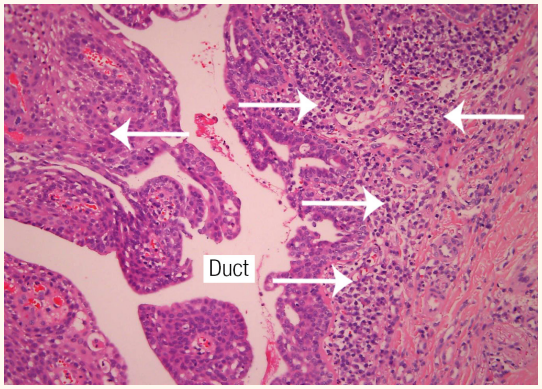

Figure 4: Histopathology indicated an abundance of plasma cells (white arrows). The specimen was processed using hematoxylin and eosin stain at x20 magnification.

The differential diagnosis of SCAP is broad. It can include *nevus* sebaceous, hidradenoma papilliferum, *molluscum contagiosum*, verruca *vulgaris*, squamous cell carcinoma, keratoacanthoma and basal cell carcinoma.[4,13]

The clinical presentation of the tumour is non-specific and can resemble any of the aforementioned conditions;[7,8] however, the histopathological features are specific and diagnostic. The tumour histology shows characteristic epidermal cystic invaginations into the dermis, papillary projections and abundant plasma cell infiltration.[2,8,10] The invaginations are lined by a double layer of epithelial cells, whereas the papillary projections are lined by glandular epithelium which often exhibits decapitation secretion.[4,8,10] Immunohistochemical studies using epithelial membrane antigen and cytokeratin markers show that the epithelium is composed of several cell types at different stages of development, with a tendency to differentiate toward the basal cells in the apocrine lineage or luminal cells in the secretory epithelium.[1,11,14] The results of these studies support the theory that SCAP is a tumour originating from the pluripotent cells.

Cases of SCAP are sporadic and most commonly seen in the scalp region. They are only rarely described in other parts of the body.[4,10,13] It is hoped that this report will add to the available information on SCAP involving the upper lip. An extensive search of the literature has shown that only three similar cases have been documented so far.

Although this lesion is benign, a transformation to basal cell carcinoma, metastatic adenocarcinoma or ductal carcinoma may still occur.[15,16] SCAP has been associated with basal cell carcinoma in about 10% of cases.[7,10] Squamous cell carcinoma may also develop, but much less frequently. Other malignant tumours associated with SCAP include verrucous and ductal carcinomas.[16] Rapid growth, pain, ulceration or bleeding in a long-standing lesion may indicate a malignant transformation to syringocystadenocarci noma papilliferum.[6,16]

Conclusion

SCAP is an uncommon tumour and its appearance on the upper lip is very rare. The clinical presentation of this tumour resembles many other skin lesions. It should be considered in the differential diagnosis of any long-standing plaque or nodular lesion, particularly if there is an increase in size or the occurrence of ulceration and/or bleeding. The tumour has characteristic histological features which can be used as the basis for diagnosis. Malignant transformation can occur; a complete excision of the lesion is therefore the treatment of choice.

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