CASE REPORT

Scrotal syringocystadenoma papilliferum: case report

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Syringocystadenoma papilliferum (SCAP) is a benign rare adnexal skin neoplasm, which in a third of cases arises from a nevus sebaceous and is most commonly found on the head and neck and in very rare instances found on the genitalia. We report on a 59-year-old man with SCAP on the scrotum. The clinical scenario and histopathological findings are outlined. Following excision and histological confirmation of a noninfiltrative process, the patient remains asymptomatic. To our knowledge, only three other cases of such a lesion on the scrotum have been reported in the literature.

Key Words: syringocystadenoma papilliferum, adnexal, skin neoplasms, scrotum

Introduction

Syringocystadenoma papilliferum (SCAP) is a benign rare adnexal neoplasm occurring at any point in one's lifetime. Three clinical types have been described: plaque type (presents mostly as an area on the scalp devoid of hair); linear type (most commonly seen on the neck or face); and solitary nodular type (mostly seen on the trunk – shoulders, axillae, and genital area).[1] In general, SCAP usually presents as a papular lesion or a plaque on the head and neck, or less commonly on the extremities. Characteristically, SCAP is described as being a solitary grey or dark brown papillary or verrucous, exophytic lesion with a moist appearance that emerges from a flat and smooth skin colored to brown plaque that increases in size at puberty.[1-3] A third of the time SCAP arises within a nevus sebaceous (a circumscribed hamartomatous lesion predominantly composed of sebaceous glands), while the majority of the time it arises de novo. Those lesions arising within a nevus sebaceous are at increased risk for malignant potential. Basal cell carcinoma (BCC) is associated with such lesions in up to 10% of cases. Less commonly, SCAP associated with a nevus sebaceous may progress to squamous cell, verrucous, or ductal carcinoma.[3] The diagnosis of SCAP involves a certain degree of clinical suspicion along with histological confirmation. Due to the aforementioned association with malignant potential, complete surgical excision along with thorough histological examination is the treatment modality of choice.[1-5]

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Case report

A 59-year-old male, with a previous history of bladder cancer, was being followed in our clinic for routine bladder cancer surveillance when he reported changes in a skin lesion on the scrotum. The skin lesion had been present for over 30 years and had been gradually increasing in size over the past 2 years. The lesion was a grey white cystic structure filled with soft yellow grey material. It was located on the inferior pole of the left hemi scrotum, 5 mm x 5 mm in size with a well defined margin. The patient denied any bleeding or discharge from the lesion, but did note that recently he had been experiencing increasing amount of pain from the site, particularly at the end of the day.

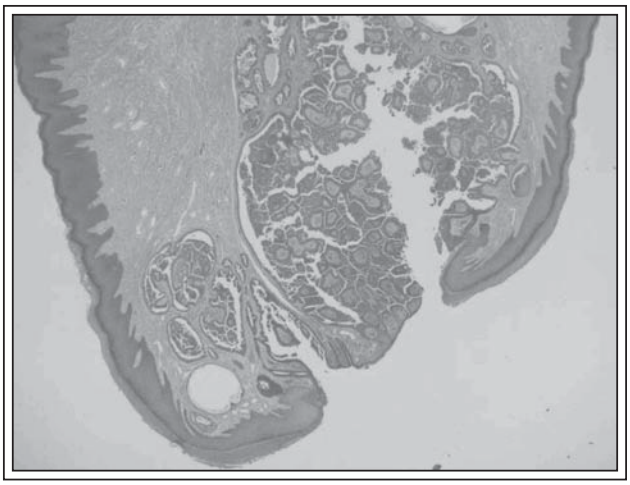

Figure 1. A low power photomicrograph showing the papillary intracystic growth pattern and the opening of the cystic neoplasm to the epidermal surface. (20x)

Histopathology

Microscopic examination shows a cystic skin adnexal neoplasm with an intracystic papillary growth pattern, Figure 1. The neoplasm opens onto the skin surface and the epithelial lining of the neoplasm is in direct continuity with the epidermis in the infundibular area. The epidermis around the opening of the tumor shows mild hyperplasia. The neoplasm has a multinodular or lobulated growth pattern that is consistent with secondary outpouchings from a central cystic space. There is no evidence of an invasive pattern. The epithelial lining is cytologically benign and is composed of two layers – an inner columnar lining and an outer basal cell layer, Figures 2 and 3. The columnar epithelium has eosinophilic cytoplasm and some of this epithelium exhibits apical snouts, consistent with apocrine differentiation. Apocrine differentiation is supported by focal staining with Gross Cystic Disease Fluid Protein, a marker for apocrine differentiation, Figure 4. The epithelial lining focally shows evolving squamous metaplasia, mainly in the deeper part of the neoplasm and quantitatively minor. The luminal secretion stains weakly with both alcian blue and PAS stains. The stroma of the papillary configurations contains plasma cells, especially in the superficial zone. Collectively, these features are considered typical

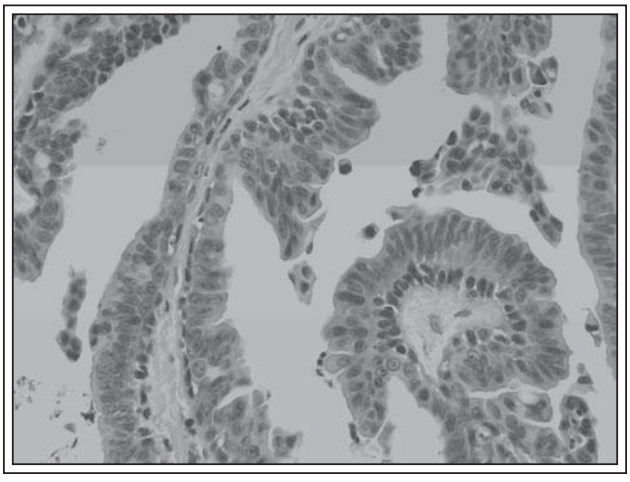

Figure 2. A high magnification of the epithelial lining of the papillary configurations of the neoplasm. It shows the bland double cell lining and the apocrine cytologic features of the inner columnar lining. (400x)

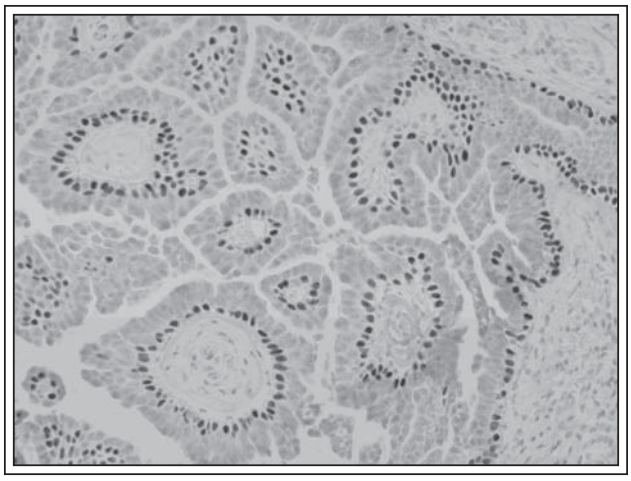

Figure 3. A p63 immunohistochemical evaluation of the papillary tumour growth. It highlights the basal cell layer of the neoplasm. (200x)

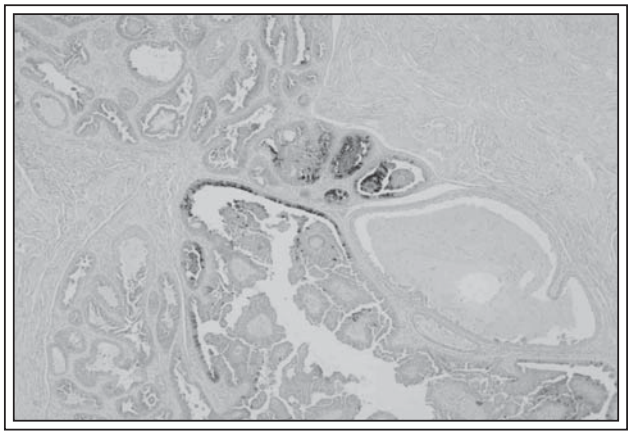

Figure 4. A gross cystic disease fluid protein (GCDFP) immunohistochemical evaluation showing focal staining. This finding supports apocrine type differentiation in these areas.

for syringocystadenoma papilliferum – a benign skin tumor that most experts think exhibits apocrine differentiation.

Discussion

SCAP was first described by Stokes in 1917.[6] In reviewing the literature there appears to be 165 documented cases of SCAP. The majority of these cases (118) appeared on the head and neck, followed by trunk (34), lower limb (9), scrotum (3) and upper limb (1).[2,4,7] A case of SCAP arising on the scrotum is exceedingly rare. There have only been three cases previously reported in the literature.[3,4,8] The clinical differential diagnosis of the lesion in our case includes: basal cell carcinoma, squamous cell carcinoma, fibroma and infected sebaceous cyst.[1,7] Histological examination made the diagnosis and ruled out the aforementioned clinical considerations.

Histopathologically, SCAP has characteristic features that lead to a reproducible diagnosis in typical examples. Typical examples have an endophytic cystic configuration that is lined by bland columnar epithelium with an outer myoepithelial layer. Well formed papillary structures protrude into the cystic space and may protrude above the skin surface. Plasma cells are consistently present in the stroma of the papillary structures. The columnar lining typically shows apical snouts, at least focally. This feature is thought to reflect apocrine differentiation and there is ultrastructural and immunohistochemical evidence in the literature to support this interpretation. The main consideration in the histologic differential diagnosis (in a tumor not associated with nevus sebaceous) is hidradenoma papilliferum. The opening onto the skin surface and presence of prominent numbers of plasma cells are features that favor the diagnosis of SCAP.

There is still debate about the cellular origin of SCAP. Findings with light and electron microscopy show features consistent with both apocrine and eccrine differentiation as do findings with immunochemistry. Of particular interest is the anatomical distribution of such lesions. Greater than 90% of SCAP are found in areas lacking apocrine structures (scalp, head, neck) and few examples are reported from sites rich in apocrine elements (scrotum, axilla, breast).[2,3,11] In our case, the scrotal location is a plausible site for a neoplasm arising from an apocrine gland.

Another theory proposes that these neoplasms arise from pluripotent appendageal cells.[8,11,12] This line of thinking would explain the apparent mixed differentiation that is suspected in many cases of SCAP. That said, the consensus seems to be that the majority of SCAP exhibit apocrine differentiation, as seen in our case. It seems likely that the apocrine differentiation expressed in many of these neoplasms is not reliable evidence that the neoplasm arose from a mature apocrine gland.

Conclusion

SCAP, not associated with nevus sebaceous, is an uncommon skin tumor. It is especially rare in a scrotal location making this tumor of special interest. The debate as to the origin of such neoplasms is likely to continue for some time, especially with so few cases being reported in the literature. The theory postulating the evolution of such neoplasms from pluripotent appendageal cells appears to be most logical in view of the suspected mixed apocrine and eccrine differentiation in many of the reported examples. SCAP is reliably diagnosed by histologic examination and it behaves as a benign skin adnexal tumor. The recommended treatment is simple excision and this tumor does not have a reputation for frequent local recurrence. □

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