CASE REPORT

Syringocystadenoma Papilliferum: An Uncommon Diagnosis

Chimdiebele D **NWOSU**[1]
Ochonma A **EGWUONWU**[2]
Gabriel U **CHIANAKWANA**[2]
Ikechukwu R **AZUBUIKE**[1]
Augustine N **NWOBI**[1]
Michael E **CHIEMEKA**[3]

[1]*Department of Surgery*
Nnamdi Azikiwe University Teaching
Hospital Nnewi, Anambra State, NIGERIA

[2]*Department of Surgery*
Nnamdi Azikiwe University Awka/
Nnamdi Azikiwe University Teaching
Hospital Nnewi, NIGERIA

[3]*Department of Histopathology*
Department of Surgery
Nnamdi Azikiwe University, Awka NIGERIA

Author for Correspondence
Dr Chimdiebele **NWOSU**

Department of Surgery
Nnamdi Azikiwe University Teaching
Hospital Nnewi, Anambra State, NIGERIA

Phone: +234 806 439 8386
Email: daisynwo@gmail.com

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ABSTRACT

Syringocystadenoma papilliferum is a benign harmatomatous tumour of apocrine or eccrine sweat gland origin. We report a 30-year-old female who presented with a 24-year history of a slowly growing scalp lesion with a preceding history of trauma to the same region of the scalp as a child. Examination revealed a 2x2 cm tumor in the mid-scalp region, exuding scanty sero-sanguinous fluid on contact. She had an excision biopsy of the lesion following a tentative diagnosis of foreign body granuloma. However, pathologic findings included glandular papillary proliferation connected to the epidermis and lined by columnar cells with oval nuclei with overall impression of Syringocystadenoma papilliferum. She was subsequently reassured and has been on follow-up with no recurrence.

Keywords: *SCAP, Scalp lesion, Harmatomatous tumour, Papillary syringoadenoma*, Nigeria

INTRODUCTION

Syringocystadenoma papilliferum (SCAP), also known as papillary syringoadenoma is an uncommon hamartomatous tumour which is believed to originate from the adnexae of the skin.[1,2] The adnexae includes the sweat and sebaceous glands, hair follicles and other cells located within the epidermal skin layer.[2] Adnexal tumours appear alike clinically since they share a common development and differentiation.[2] It is therefore difficult to make a diagnosis based on clinical grounds alone. Like most

adnexal skin tumours, SCAP is equally benign.[2]

Syringocystadenoma papilliferum was first described more than a century ago in German literature and by Stokes in 1917 in English literature.[3,4] It is considered a childhood tumour and is present at birth in approximately 50% of patients, while it develops before puberty in 15%-30%.[5] SCAP is assumed to occur more in females as was the case in our patient.[6] Seventy-five percent of SCAP lesions occur in the head and neck region and most of these lesions are found on the scalp.[6] This tumour can be found in other areas of the body and has been reported to occur in the lower limb and eye lid in studies in the Western and Northern parts of Nigeria.[7,8] Multiple SCAP lesions are uncommon.[7] SCAP is largely a histopathologic diagnosis and as such may be missed unless the clinician has a high index of suspicion. As only a few cases have been reported in the literature from Nigeria and these being from other regions, we present another interesting case of SCAP in a tertiary health centre in the south eastern part of Nigeria.[2,7,8]

CASE SUMMARY

A healthy, non-smoking 30year old female presented to the general surgery outpatients' clinic with a complaint of a slowly growing scalp swelling of 24 years' duration. She initially noticed an unremarkable nodule in the mid-scalp region when she was six years old. The lesion slowly increased in size over the course of the next twenty-four years with mild pain and alopecia noted over the swelling. She had also noticed a serous/bloody discharge on contact. She had no other swellings in any other part of her body. She reported having sustained a scalp injury on the said area as a child. She had no history of weight loss and no other constitutional symptoms. Other past medical history, family history and review of systems were not significant.

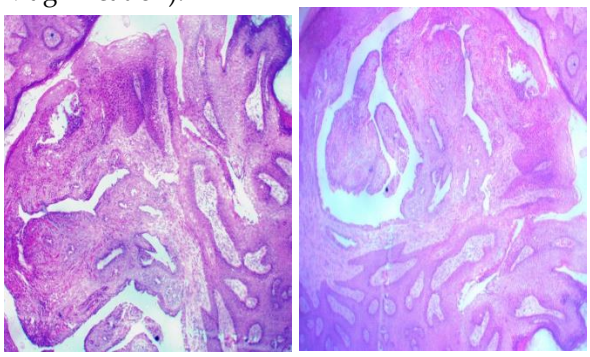
Physical exam revealed a 2x2cm nodule in the right parietal scalp region. This was covered by negroid skin, while the central portion was covered by pink granulation tissue. The area surrounding the lesion was not differentially warm or tender. The lesion was firm in consistency with clearly defined edges. It was not fixed to the deeper structures but was however attached to the scalp and exuded scanty sero-sanguinous fluid on contact. There was no regional lymphadenopathy. Following a tentative diagnosis of a foreign body granuloma, she was appropriately counselled for surgical excision. She subsequently had the lesion and the surrounding 1cm of skin around the lesion completely excised with under local anaesthesia. The wound was closed primarily. She has been followed up at the outpatient clinic for 10 months with no recurrence.

Histopathological Examination

Gross findings as illustrated in the Figure, were those of an irregularly shaped yellowish firm tissue covered by negroid skin with a nodular outgrowth. It measured 4.5x2.0x1.3cm and weighed about 20grams. The cut surface was yellowish.

Microscopy of sections of the specimen showed a benign neoplasm composed of glandular papillary proliferation connected to the epidermis. These projections were lined by columnar cells with oval nuclei. The stroma was infiltrated by dense plasma cells. However, no atypical cells were seen. The overall histopathological features were diagnostic of Syringocystadenoma Papilliferum.

Figure. Photomicrographs of the lesion illustrating glandular papillary proliferations and lining cells. (Haematoxylin and eosin stain. X10 Magnification).



DISCUSSION

The rare nature of the SCAP is evident in the retrospective review of adnexal skin tumours over a fifteen-year period in a Northern Nigerian tertiary hospital by Samaila.[2] Adnexal skin tumours accounted for less than 0.9% of all cutaneous tumours in this review. Of the fifty-two (52) adnexal tumours reported in the patients over this period, only 2 (3. 8%) tumours were identified as SCAP.[2]

There is some controversy as to the exact origin of the harmatomatous SCAP tumour. Although it offered a better insight, immunohistochemical and ultrastructural studies such as that by Yamamoto *et al.* to determine the exact origin of the tumour failed to resolve this controversy.[3] The cells comprising the SCAP tumour were found to react positively to several anti-cytokeratin antibodies including CK7, CK19 and CK5 in a pattern similar to that observed in ductal/secretory part of adult sweat glands in this study.[3] Ultra-structurally, three cell types were identified and these include the basal cells, luminal cells and clear cells.[3] These cells were observed to be in different stages of development. Although ultrastructural studies seem to be more in keeping with apocrine origin, immunohistochemistry did not contribute significantly in determining the eccrine or apocrine origin of SCAP.[3] Immunohistochemistry was however not done for the index patient.

SCAP develops either independently or in association with the nevus sebaceous of Jadassohn in seventy-five percent of cases.[9] It has also been associated with benign lesions such as nevus comedonicus, apocrine poroma and hidradenoma papilliferum.[8] Although a benign hamartomatous tumour, it can transform to its malignant variant; Syringocystadenocarcinoma papilliferum or to basal cell carcinoma in 10% of cases.[6,10] SCAP may co-exist with other malignant tumours such as verrucous carcinomas (a variant of squamous cell carcinoma).[6] It has equally been reported to co-exist with two other histologically different cell types including the nodular variant of basal cell carcinoma and a verrucous carcinoma in a single tumour on a patient's scalp.[6] No other lesions were reported to co-exist with SCAP found in the index patient.

Three clinical variants of SCAP have been described and the clinical presentation depends on these various types. They include; the plaque variety which is usually seen in the scalp as a flat hairless patch which may enlarge.[7] This variant is usually associated with the sebaceous nevus of Jadassohn. [7] It may also present on the face and neck region in the linear type which consists of multiple papules or nodules with a punctum/umblication and on the trunk preferentially in the solitary nodular type.[7]

SCAP usually occurs in childhood and this was the case in our patient who had observed this lesion since she was six years old. Although the plaque variety occurs more in the scalp region, this was not the case in our patient as she had a scalp nodule. Histopathology of the patient's specimen was consistent with the known description of

SCAP although no mention was made as to the presence or absence of decapitation secretions.

We entertained a diagnosis of foreign body granuloma because our patient gave a history of injury to that region of the scalp, however the absence of fibroblasts, endothelial cells and inflammatory cell infiltrates ruled out foreign body granuloma.[11] Some other differential diagnosis of SCAP include hidradenoma papilliferum, tubular apocrine adenoma, warty dyskeratosis and inverted follicular keratosis.[12] Hidradenoma papilliferum is differentiated from SCAP by the presence of basaloid cells surrounded by proteinaceous fluid and mucoid aspirates. Tubular apocrine adenoma differs histologically by the absence of background plasma cell infiltrate while warty dyskeratosis and inverted follicular keratosis contain keratinous material which is not seen in SCAP.[12]

The only treatment for SCAP is surgical excision which is also used to confirm the diagnosis in most occasions. Surgical excision is necessary because of the small risk of transformation to the malignant variant.[13] Diagnosis of SCAP on clinical basis is not reliable due to its different clinical presentations, several possible differential diagnosis and its rare nature. This makes histopathological diagnosis a necessity. Awareness of this condition and a high index of suspicion can therefore prevent misdiagnoses as was the case in the index patient. The prognosis for the lesion is the same regardless of the type and malignant transformation is possible though rare. This is usually to basal cell carcinoma then squamous cell carcinoma and very rarely to its malignant variety Syringocystadenocarcinoma papilliferum.[5]

CONCLUSION

As Syringocystadenoma papilliferum is a rare tumour, it is important to have a high index of suspicion when evaluating such scalp lesion with a history dating back to childhood to prevent a misdiagnoses and possible over treatment at surgery.

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