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MULTIPLE SYRINGOCYSTADENOMA PAPILLIFERUM ARISING FROM AN EXTENSIVE NEVUS SEBACEOUS OF JADASSOHN: CASE REPORT
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MULTIPLE SYRINGOCYSTADENOMA PAPILLIFERUM ARISING FROM AN EXTENSIVE NEVUS SEBACEOUS OF JADASSOHN: CASE REPORT

S. K. KIPRONO, S. M. ALI and A. M. EMARAH

SUMMARY

We present an extensive nevus sebaceous of Jadassohn which was present at birth and had a rapid growth during puberty. Multiple nodules of syringocystadenoma papilliferum developed on the plaque of nevus sebaceous. The extensive nature of nevus sebaceous and numerous benign tumours is a therapeutic challenge on the face. Currently there is no adequate evidence to support prophylactic excision of nevus sebaceous of Jadassohn, especially in people with skin of color

INTRODUCTION

Nevus sabaceus of Jadassohn (NS) is cutaneous hamartoma that involve the epidermis and adnexial structures (1). The skin lesions are usually present at birth or may occur early in childhood. Majority of the NS occur in the head and neck region, particularly the scalp were it presents as flat or mamillated patch of alopecia (2). NS is round to oval, well circumscribed patch measuring 1-6 cm in diameter (2). Syringocystadenoma papilliferum (SCAP) is usually a solitary, benign adnexial tumour of predominantly apocrine or eccrine differentiation (3). It may present at birth or in childhood. Commonly it occurs on the scalp, in which it is associated with NS (4).

CASE REPORT

A 23 year old female presented to the outpatient clinic with a skin lesion on the left side of the face extending from the vertex to the anterior neck as shown on figure 1A and B. The skin lesion was present at birth and has been growing with age. Rapid growth was observed during puberty. In the last five years the patient noticed an eruption of numerous papillomatous nodules within the plaque. These nodules were painful,had foul smelly discharge and bled easily on touch. On local examination there were 32 discrete flesh coloured papillomatous nodules covered by haemorrhagic crusts. On the scalp the plaque was associated with scarring alopecia. The extension on the neck was hyperpigmented verrucous plaque with small pedunculated nodules as shown on Figure 1C. No other developmental abnormalities were noted. The HIV status was negative and a CT scan of the head was normal. Baseline investigations were within normal ranges. Biopsy done showed a tumour composed of duck-like structures that extend to the surface epithelium (Figure 3). The papillomatous tumour is lined by squamous epithelium on the surface and double columnar layer on deeper part. Decapitation secretions are noted arising from the columnar cells. The papillomas have a fibrovascular corewith lymphocytes and plasma cell infiltrate. Skin surrounding the tumour showedpapillomatous epidermal hyperplasia and an increase in sebaceous glands which are placed abnormally higher in the papillary dermis (Figure 2A and B). There were no histological features of Human papilloma virus infection. The final diagnosis was SCAP arising from NS. Tumour was excised and full thickness skin grafting harvested from the lower abdominal wall was done in stages. The patient is being followed up in our clinic and at three months the wound was healed and no features of recurrence.

Figure 1A,B,C,D
A verrucous facial lesion extending to the neck. The is associated alopecia in 1B and involvement of the eyebrows and nasal labial fold in 1D

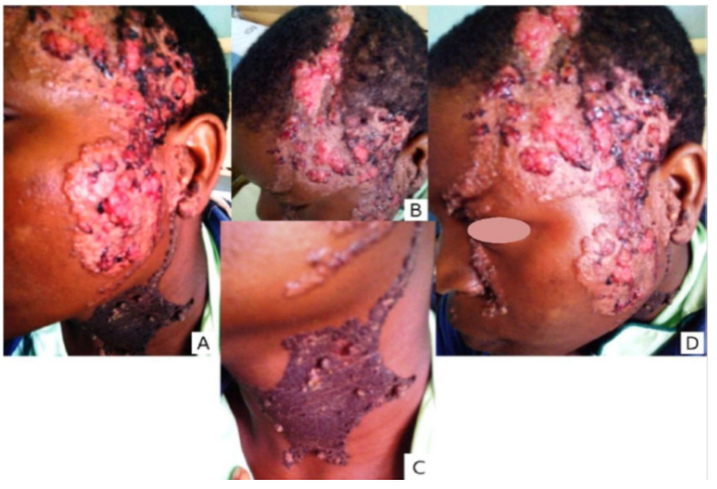


Figure 2A (X5) and B (X10)
Hematoxylin and eosin stain of the backgroundlesion. Superficially located sebaceous gland hyperplasia with conspicuous apocrine glands

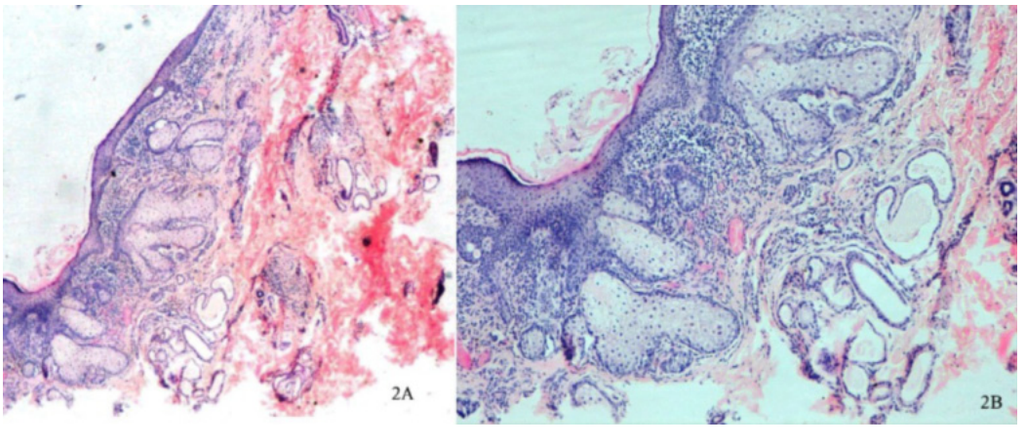
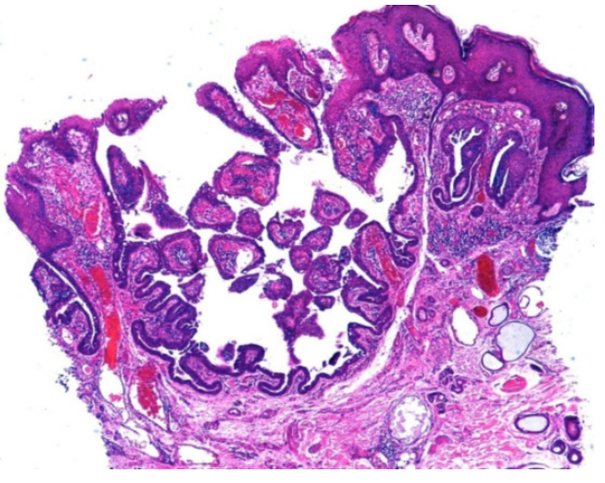


Figure 3 (X5)
Hematoxylin and eosin staining of the nodules. Duct-like structures extend as invagination from the surface of the epithelium into the dermis. Higher magnification shows the ducts are covered by columnar epithelium with decapitation secretions



DISCUSSION

This is a rare case of extensive NS with multiple SCAP tumours. No other type of tumour was seen within the NS plaque. The SCAP arising from NS has been widely reported in English literature however in this patient, the extensive nature of NS and numerous SCAP tumoursis a therapeutic challenge in cosmetically sensitive areas. According to recentreview (2), 16% (0-51%) and 8% (0-21%) of NS develop benign and malignant tumours respectively. Among these benign tumours are trichoblastoma, trichilemoma, sebaceous adenoma, hidradenoma and syringocystadenomapapilliferum (2,5). Majority ofthe malignant lesion are basal cell carcinoma (BCC) and few apocrine adenocarcinomas (5). A retrospective review of 596 cases by Cribier *et al* (6),found a low (0.8%)incidence of BCC while majority (90%) of the tumours previously reported has BCC were trichoblastomas.The incidence of BCC is generally low in people with skin of colour (7). Similarly, in African albinos the prevalence of BCC is lower than squamous cell carcinoma (SCC) (8). It remains to be determined whether NS increases the risk of BCC in African population. Multiple tumours that include SCAP, tumour of follicular infindicular,sebaceoma, trichoblastoma,trichoadenoma and trichilemoma have been reported to coexist within the same lesion of NS4 (9,10,11). The multiplicity of these tumours is thought to arise from heterogenic mechanisms of pluripontential primary epithelial germ cells of NS (12). A genetic defect at 9q22.3 has been suspected in development of multiple tumours (13). The gene mutation could also explain the development of multicentric SCAP in this patient.

The treatment of NS is by excision to the subcutis: This is usually done for cosmetic reasons (2). Currently there is no consensus on the role of prophylactic excision of NS to prevent malignancies (2). The risk of malignancy within NS in African patients is unknown. The risk could be lower than reported in Caucasians hence prophylactic excision will not be justified. There is lack of data on the risk of a second tumour arising from the remaining NS after previous excision of a tumour on the same lesion. The role of secondary prophylactic excision in patients with few tumours arising from extensive NS is unknown. In this patient the NS lesion on the nasal area and the neck were tumour free. There is no sufficient evidence for removable of tumour free lesions in cosmetically sensitive areas.

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