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SYRINGOCYSTADENOCARCINOMA PAPILLIFERUM OF THE SCALP ARISING FROM NAEVUS SEBACEOUS OF JADASSOHN IN ADULT MALE FROM RURAL SARAWAK: A CASE REPORT

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INTRODUCTION

Syringocystadenocarcinoma Papilliferum (SCACP) is an exceptionally rare cutaneous adnexal skin neoplasm of the apocrine or eccrine sweat glands or both. SCACP is regarded by World Health Organization classification of skin tumours as the malignant form of a more common adnexal skin tumour, Syringocystadenoma Papilliferum (SCAP)[1].

One recent study revealed only 50 cases of SCACP were reported over 34 years in English literature in which 27 out of the 50 cases occur at head and neck region[1]. Other locations included buttock, eyelid, axilla and pinna[1,2]. SCACP may progress from pre-existing naevus of Jadassohn since birth but can also be de novo[1].

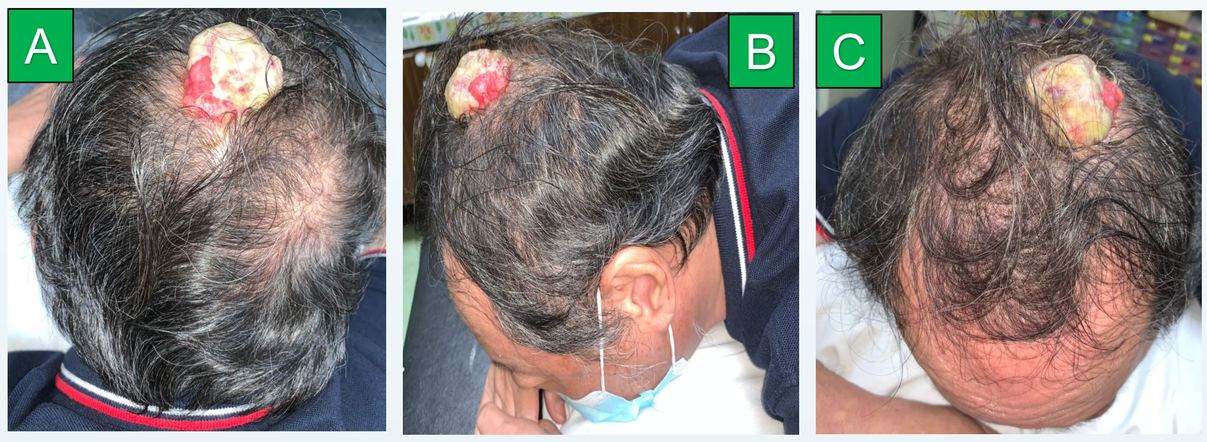
We hereby present a 68-year-old Chinese male who was referred to Neurosurgical department for wide local excision of scalp malignancy due to lack of plastic surgery service in rural Sarawak.

CASE PRESENTATION

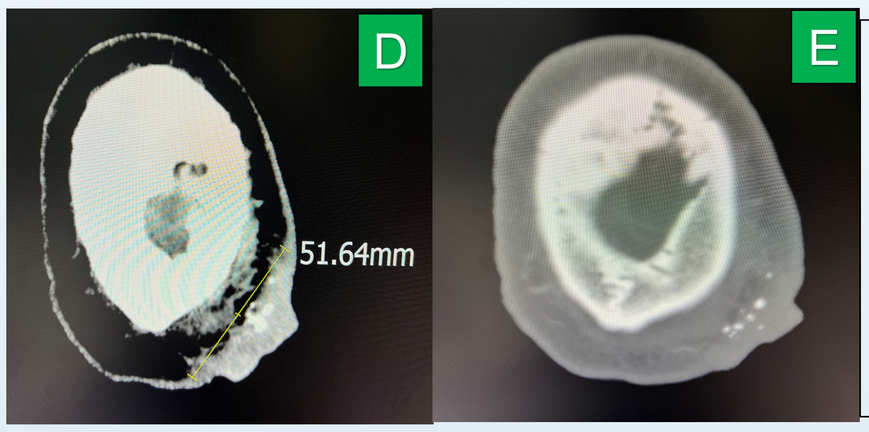
A 68-year-old man with underlying hypertension presented with 6-month history of progressively increasing in size of vertex scalp swelling, associated with occasional foul-smelling pus discharge. Upon further history taking, he claimed that he had a lesion at the same site since birth. The lesion was initially small, flat with reddish brown pigmentation and constant in size and shape. Otherwise, he denied any constitutional symptoms nor family history of malignancy.

On examination, the patient was well built and alert. He had frontal and bitemporal fibrosing alopecia. No lymphadenopathy. The solitary scalp lesion was located at the vertex, more towards the left high parietal region, measuring 4x5x2.5cm. It was asymmetrical, fungating, firm, non-fluctuant, immobile, non-tender and verrucous with foul-smelling sanguinopurulent discharge. The surrounding of the lesion was crusted. His systemic examination was unremarkable.

Routine laboratory tests were all within the normal range. Electrocardiogram and chest radiograph were normal as well. Contrast-enhanced computed tomography (CECT) brain showed no bone erosion and intracranial extension. CT thorax, abdomen and pelvis showed no distant metastasis.



Figures A, B and C showed the vertex scalp lesion from posterior, side and frontal view respectively.



Figures D and E showed vertex scalp lesion, more towards the left high parietal region with calcification within the lesion. Skull was spared.

The patient then underwent a wide local excision under local anaesthesia with 1cm clear margin. Z-incision was made to maximise closure.

Intraoperatively, the scalp tumour was poorly circumscribed and moderately vascularised. Pericranium at the affected area was excised to achieve maximal marginal resection. Outer table of skull at the affected area was drilled and bone dust was collected and sent for histopathological examination.

Extensive undermining in subgaleal plane was performed to allow tissue movement for closure. Galea was approximated with Vicryl 2/0 and skin was closed with Nylon 3/0 – both in simple interrupted manner. However the scalp laxity was rather stiff and immobile partly due to age and dermal fibrosis as evidenced by alopecia over frontal and temple regions. Thus there was a 3x1cm defect left behind as galea approximation was not possible and the wound remains very tense despite maximal undermining of subgaleal plane.

Postoperatively, daily ribbon gauze soaked with normal saline dressing was applied. At post surgery day 3, the defect was clean and remained pink. At week 1, the base of defect started to fill up. Sutures were removed with no gapping except the defect. At week 2, the wound including the defect had fully healed but the previous defect area was inverted and spread.

The patient had been well without relapse for 8 months thus far.

Figures F, G and H showed the wound at post surgery day 3, day 7 and day 14 respectively.

DISCUSSION

SCACP is an extremely rare adnexal skin tumour. There was no gender predilection but some reports revealed male to female ratio of 1.5:1[1].

Clinically, this lesion could generate various differential diagnosis. Thus, histopathological diagnosis is crucial.

Histopathologically, the tumour resembles its benign counterpart, syringocystadenoma papilleferum (SCAP)[1,2,3]. The tumour usually has both solid and cystic structures and composed of two-tiered or multi-layered epithelium[1,3]. However, SCACP has asymmetrical structure and poorly circumscribed margin. SCACP usually invades dermis and its dermal component are commonly of varying shapes and sizes such as cribriform, angulated glandular and cords[1,3]. SCACP also exhibits variable degree of nuclear atypia and pleomorphism as well as numerous mitotic figures[3]. SCACP has higher nuclear to cytoplasmic ratio, nuclear irregularity and coarse chromatin[1,3].

Immunohistochemistry usually showed different immunoreactivities[1]. In this case, it showed positive reaction to gross cystic disease fluid protein (GCDFP)-15.

Complete surgical excision with 1cm clear margin is the preferred treatment but chemotherapy or radiotherapy can be offered in patients who refuse surgery or in inoperable cases[1]. In this case, the margin was clean histopathologically.

CONCLUSION

SCACP is a rare skin neoplasm in which its pathophysiology and histogenesis remain unclear. It can be associated with naevus sebaceous of Jadassohn. Histopathological diagnosis is needed. Wide local excision is the mainstay of treatment.

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