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Original Research Paper

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SYRINGOCYSTADENOMA PAPILLIFERUM OF SCALP – A CASE REPORT OF A RARE CUTANEOUS ADNEXAL NEOPLASM

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ABSTRACT Syringocystadenoma papilliferum is a rare benign hamartomatous tumor arising from apocrine or eccrine sweat glands with a predilection for head and neck. Here we report a case of a 30 year old female who presented to our opd with a nodule over scalp since birth which turned into a hairless , exophytic pinkish growth with blood tinged discharge slowly over the last 2 years . A punch biopsy of the lesion revealed syringocystadenoma papilliferum. Excision of the lesion is performed and specimen is sent for (hpe) histopathological examination which revealed syringocystadenoma papilliferum with no malignant transformation. Post operative recovery was uneventful and the patient was followed up till 6 months with no history of recurrence.

KEYWORDS : Syringocystadenoma Papilliferum , Sebaceous Nevus , Cutaneous Adnexal Neoplasm.

INTRODUCTION

Syringocystadenoma papilliferum is a rare, benign hamartomatous adnexal tumor arising from eccrine or apocrine sweat glands . Syringocystadenoma papilliferum usually affects infants and children, presentation in adolescents and adults constitute 15–30% cases. It occurs more commonly in the head and neck ,and very rarely in other areas.

Interestingly, the first case of SCAP (SYRINGOCYSTAD ENOMA PAPILLIFERUM) was described on the thigh of a patient by Stokes in 1917 under the term nevus syringo adenomatosus papilliferus[1]. It usually presents as a solitary or multiple skin coloured nodules or papules in linear arrangement or as a solitary plaque at birth , lesion increases in size at puberty ,becoming papillomatous and often crusted. On the scalp , it frequently arises around puberty within a sebaceous nevus of Jadassohn that has been present since birth.[2]

Syringocystadenocarcinoma papilliferum is extremely rare and may present within a pre existing lesion as insitu and /or invasive adenocarcinoma or rarely squamous cell carcinoma. It spreads by lymphatics and has a favorable prognosis. It can be successfully treated with Mohs micrographic surgery.[3]

CASE REPORT :

A 30 year old female presented to our out patient department with a swelling over scalp since birth , it was initially present as a nodule of 1*1cm . There was a history of gradual increase in size of nodule associated with itching and crusting for the last 2 years with formation of pinkish exophytic growth with serosanguinous discharge . No history of trauma/ radiation exposure . No history of similar swellings noted elsewhere in the body. No history of rapid increase in size/ weight loss/ loss of appetite. No history of cervical lymphadenopathy .

No history of similar complaints in family members. The patient has no comorbid conditions.

On examination, there is single, fleshy, pinkish, 3*3 cm exophytic growth noted in the left temporal region of scalp with serosanguinous discharge. The surrounding skin is normal and there is no growth of hair over the swelling . On palpation the growth is non tender, firm in consistency with regular margins and everted edges. The base of swelling is mobile over the underlying bone. There are no similar swellings noted over the rest of the scalp and there is no cervical lymphade nopathy.

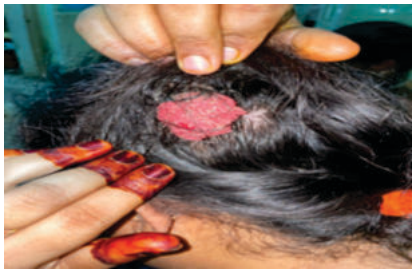

Figure 1: showing preoperative picture of the tumor in left temporal region of scalp

Investigations

BLOOD INVESTI GATI ONS	Hb - 12 gm/dl Wbc - 6800/mm3 Platelets - 2.2 lakhs/mm3 Rbs - 105 mg/dl Serum creatinine - 0.5 mg/dl Blood urea - 20 mg/dl
ULTRASONOGR APHY	Well defined, heterogenous, soft tissue dense lesion measuring 27*14*23 mm, noted in the left posterior parietal region involving skin and subcutaneous tissue extending into the subgaleal plane , There is no extension into the underlying periosteum. IMPRESSION-SOFT TISSUE LESION OF SCALP
PUNCH BIOPSY OF THE LESION	Punch biopsy report showed Epidermal hyperplasia with hyperkeratosis , hyper granulosis , papillomatosis , cystic invagination of infundi bular epithelium projecting into dermis lined with two types of cells , innermost columnar cells with decap itated secretions and outer layer of cuboidal cells with stroma of papillary fronds showing lymphocytes and plenty of plasma cells . S/O: SYRINGOCY STADEN OMA PAPILLIFERUM
CT HEAD AND NECK	2.5*3 cm Soft tissue swelling of scalp with no bony involvement or intracranial extension. No cervical lymphadenopathy.

Patient was posted for surgery and the swelling was excised under general anesthesia. The defect was around 2 cm which was closed primarily without the need of a rotational flap and the excised specimen was sent for histopathological examination (HPE).

Histopathology Report :

Soft tissue mass measuring 4*3*2 cm with cut section showing multiple cystic spaces of gray white , gray yellow, gray brown material with sections showing superficial stratified squamous epithelium with hyperkeratosis, acanthosis, hypergranulosis (figure 2) with cystic invaginations of epithelium projectiong as papillary folds with fibrovascular cores into the dermis. These papillae are lined by inner columnar and outer cuboidal epithelium. Dermis shows fibrocollagenous tissue , adnexae and dilated ducts along with perivascular and periadnexal chronic inflammatory infiltration. One foci shows adenosis of ducts and some others showing secretions with cholesterol clefts , adipose tissue and congested blood vessels. (figure 3)

Impression :

Scalp swelling showing Syringocystadenoma papilliferum with no malignant features.

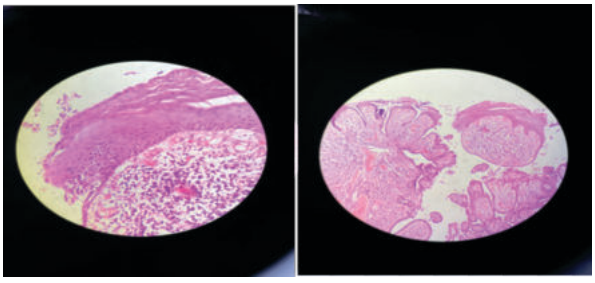

Figure 2 (40x) showing surface stratified squamous epithelium with hyperkeratosis and hypergranulosis.

Figure 3 (10x) showing papillary folds with dilated capillaries and plasma cell infiltration.

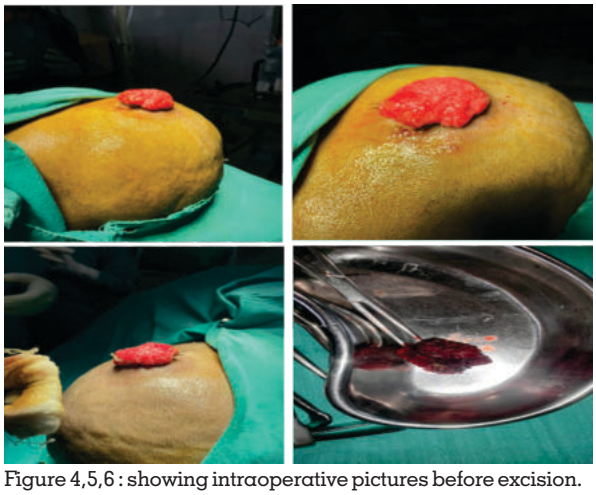

Figure 4,5,6 : showing intraoperative pictures before excision.

Figure 7 : showing an excised specimen which was sent for HPE.

DISCUSSION :

SCAP is a rare benign hamartomatous adnexal neoplasm that originates from either apocrine or eccrine sweat glands. These benign adnexal tumors developing at birth are believed to be arising from primary epithelial germ cells in contrast to those appearing later in life which arise from pluripotent germ cells in adult epidermis and skin appendages either spontaneously or after trivial trauma like combing hair or by unknown factors.[4] Histologically, SCAP is characterized by endophytic invaginations of the epithelium into the dermis. These are duct-like structures leading into the dermal cystic spaces. Papillary projections of variable shape and size protrude into the lumen of these spaces. These are lined by double-layered outer cuboidal and luminal high columnar epithelium. Dilated capillaries and a dense infiltrate of plasma cells are noted in the stroma of these papillary projections.[5]

The main differential diagnosis includes basal cell carcinoma, cutaneous lymphoma, factitious dermatitis, and pyogenic granuloma. However, slowly growing fleshy plaque that has been growing for years and sometimes oozes fluid and sometimes bleeds is characteristic for SCAP. Syringo cystadenocarcinoma papilliferum (SCACP), which is the malignant counterpart of SCAP, should be ruled out when evaluating the histopathology of SCAP.[6] . SCACP is often positive for carcinoembryonic antigen, gross cystic disease fluid, and p63. Additionally, p63 expression favors a primary sweat gland neoplasm of the skin rather than a cutaneous metastasis of a visceral adenocarcinoma.[7]

Treatment options include excision of the growth and primary closure of the defect in the skin or flaps may be needed in few cases where large defect is present post excision. The treatment of SCACP is wide local excision of the swelling with lymph node dissection if positive. Other methods include CO2 laser excision and Mohs micrographic surgery.[8,9]

CONCLUSION :

We are presenting this case owing to its rarity and common differentials such as pyogenic granuloma, basal cell carcinoma and SCACP should be kept in mind while evaluating such cases. Our patient has been diagnosed with SCAP and was managed with surgical excision and primary closure of the defect. Post operative recovery was uneventful and the patient was followed up for upto 6 months with no complaints of any local complications/recurrence.

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