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Case Report

Syringocystadenoma Papilliferum of the Scalp in an Adult Male

Abdullah Al Tarique[1], Nabila Khanduker[2]

ABSTRACT

Syringocystadenoma papilliferum is a rare skin tumour developing from eccrine or apocrine sweat gland. Though it usually appears in early life, it may occur in adult also. The clinical variants are plaques, linear and nodular. Head-neck region is affected most. Associations with naevus sebaceous, basal cell carcinoma or even squamous cell carcinoma have been described. Diagnosis is suspected clinically and confirmed by histopathology. Rapid growth, bleeding and ulceration indicate possible malignant transformation. Excision biopsy followed by reconstruction of the primary defect is the treatment of choice.

Key words: Adult, Scalp, Syringocystadenoma papilliferum

Introduction:

Syringocystadenoma papilliferum is a rare benign hamartomatous skin adenexal tumour, originating from the apocrine or the eccrine sweat glands[1,2,3]. Fifty percent are present at birth or in infancy[4,5].There is no definitive gender predilection[5]. It usually occurs on the head and neck region, may appear on the trunk and extremities also[3]. Abdominal wall, genitalia are considered unusual places for its occurrence[3,6]. In one third cases it is associated with naevus sebaceous although the tumour may emerge de novo[1-4,7,8]. The tumour has three clinical types: plaques, linear, nodular[1,4,7]. The lesion can be solitary or multiple. We present a case of syringocystadenoma papilliferum in a sixty five years old male, which appeared de novo and presented as a rapidly growing ulcerated nodular lesion on the scalp.

1. Associate Professor, Department of Surgery, Green Life Medical College and Hospital.
2. Assistant Professor, Department of Surgery, Green Life Medical College and Hospital.

Correspondence to: Dr. Abdullah Al Tarique, Associate Professor, Department of Surgery, Green Life Medical College and Hospital, 32 Green Road, Dhaka-1205. Phone: 01714496994, Email: tarekawal@yahoo.com

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Case Report:

A sixty five years old male presented with the complaints of a scalp swelling for six months. It appeared as a small nodule, gradually increased in size with itching and occasional bleeding while combing hair. About two weeks back as the tumour started a rapid growth, the surface ulcerated and bled to touch. On local examination an ulcerated swelling was found on the scalp at the left parietal region. It was soft and mobile. Cervical lymph nodes were not palpable. Clinical diagnosis was squamous cell carcinoma of the scalp. Excision biopsy with split skin graft of the primary defect was done under general anesthesia. The histopathology showed scalp tumour with epidermal acanthosis and papillomatosis. There was cyst formation containing papillary structures lined by two layers of cuboidal epithelium. The cores of papillae contained lymphocyte and plasma cell infiltration. There were no features of malignancy. Thus the diagnosis was confirmed. Post operative period was uneventful. The patient is on regular follow up.

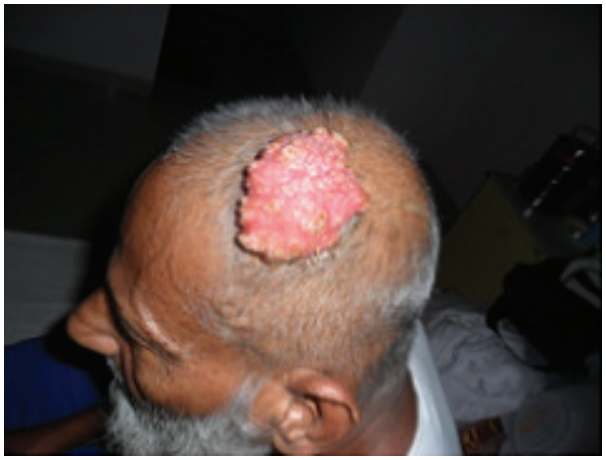

Figure 1. *The tumour on the scalp*

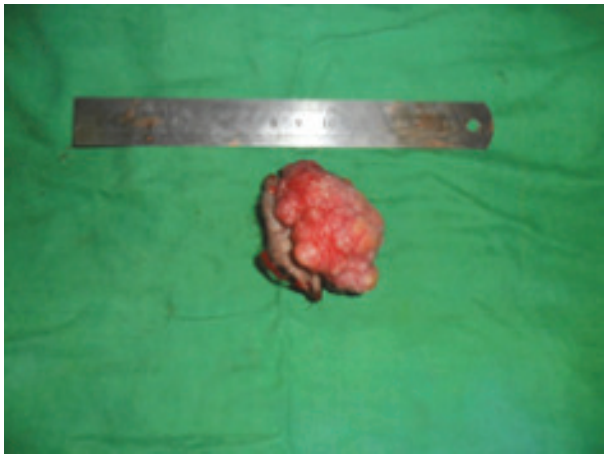

Figure 2. *Excised tumour*

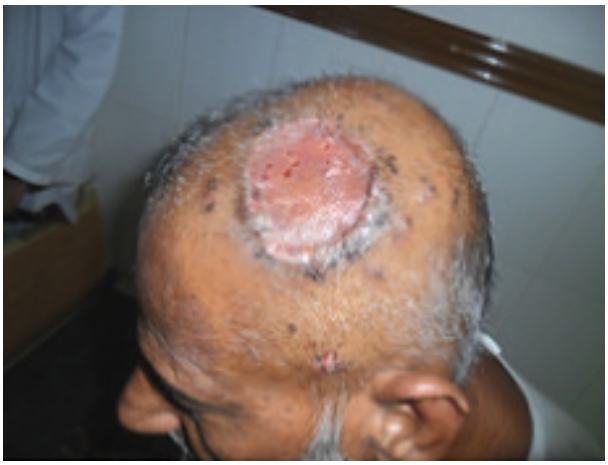

Figure 3. *Skin graft at the primary defect of the scalp.*

Discussion:

Syringocystadenoma papilliferum is a hamartomatous malformation of the skin, derived from apocrine or eccrine sweat glands and originating from pluripotent cells[1,2]. The tumour was first described by Stokes in 1917[3]. It emerges de novo or from preexisting skin lesion [7]. In one third cases it is associated with naevus sebaceous[2,4,7]. The malignant counterpart of this tumour is syringocystadenocarcinoma papilliferum[1,3]. Three clinical types of syringocystadenoma papilliferum have been described[1, 4]. The plaque type is more common on the scalp and appears as a hairless area. It may become larger, nodular, verrucous or crusted at puberty and often associated with naevus sebaceous[1,4]. The linear type commonly occurs on face or neck[1,4]. These lesions, consisting of multiple, pink to red to brown coloured firm papules or umbilicated nodules, are usually1-10 mm in diameter[1,4]. The solitary nodular type is common on trunk, shoulder and axilla and often 5-10 mm in diameter. They appear as domed, umbilicated or pedunculated swelling and may have friable or crusted surface[1,4]. Our patient had a nodular type of lesion on the scalp, which was unusual. The characteristic microscopic feature of syringocystadenoma papilliferum are epidermal hyperkeratosis and papillomatosis, cystic invagination from the epidermis, villus papillary projections into the cysts lined by two layers of cells and dense plasma cell infiltration into the core of the papillary projections[1,4,8]. About fifty percent of the tumour are present at birth or appear during infancy[1,2,5]. Our patient presented at an age of sixty five. No definitive gender predilection is described for this tumour[5]. Changes such as rapid growth or ulceration are signs of malignant transformation[2]. The patient of this case report gave a similar type of history, which biased our clinical diagnosis in favor of possible malignancy. Diagnosis of syringocystadenoma papilliferum depends on clinical suspicion from history and examination. The confirmation of clinical diagnosis is by histopathology[2,9]. Dermoscopy or surface microscopy was described as a noninvasive tool for early diagnosis and for monitoring the prognosis of this tumour[9]. Due to risk of malignant transformation, the treatment of choice is prophylactic surgical excision biopsy[1,2,8]. Laser excision and Mohs' micrographic surgery have also been mentioned[8].

Conclusion:

Syringocystadenoma papilliferum is a rare benign skin adenexal tumour. Though more common in early life, it can appear in the elderly. The tumour may emerge de novo or from preexisting skin lesion specially

naevus sebaceous. It has three clinical subtypes which can appear in unusual places. Rapid growth and ulceration leads to suspicion of malignant skin cancer as clinical diagnosis. Surgical excision is the treatment of choice and histopathology confirms the diagnosis.

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