Syringocystadenoma Papilliferum – A Rare Case Report and Review of Literature

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ABSTRACT

Syringocystadenoma papilliferum (SCAP) is very rare skin adnexal tumour of apocrine gland origin located primarily on the scalp and appearing as a hairless nodular or plaque lesion. It is said to be common at birth or during puberty but cases in adults have also been reported. We have reported a case of scalp swelling in 22-year-young female. She was first clinically diagnosed with nevus sebaceous but later histopathologically confirmed to have syringocystadenoma papilliferum.

Key words: Adnexal tumor, nevus sebaceous, syringocystadenoma papilliferum

INTRODUCTION

Syringocystadenoma papilliferum is a rare benign adnexal tumour which originates from the apocrine or the eccrine sweat glands. It is relatively a rare neoplasm, predominantly a childhood tumour. In about 50% of those who are affected, it is present at birth, and in a further 15%-30%, the tumour develops before puberty [1]. The presentation with multiple lesions is rare, and in those lesions which arise outside the head and neck region, it is even more uncommon. Syringocystadenoma papilliferum (SCAP) may occur de-novo or within a nevus sebaceous. It occasionally co-exists with other tumours such as basal cell carcinomas and verrucous carcinomas [2].It presents as a hairless area on the scalp and is said to be associated with sebaceous nevus of Jadassohn [3]. Malignant change is very rare and if any is indicated by rapid increase of size and/or change in appearance of metastatic lymph nodes [4].

We are here presenting a very rare case in young female, she was initially diagnosed as nevus sebaceous of the scalp but on histologically confirmed as syringocystadenoma papilliferum.

CASE REPORT

A 22-year young female presented with complaints of ulcerative lesion on the scalp since two years which is gradually increasing in size. It was initially a minimally raised lesion of size about one cm. It was painless, later on some nodular, pus and bloody discharge was seen. The patient is otherwise known to be generally in good health, with no comorbidities. Local examination revealed a 3 cm × 2.5 cm firm, immobile, non-tender, non-fluctuant, verrucous swelling on the vertex of the scalp and slightly favoring the right side. Sanguinopurulent discharge was evident along with crusting surrounding the lesion [Figure 1]. No other lesions were present, nor were regional lymph nodes palpable. The patient was otherwise clinically normal on systemic examination. Routine laboratory tests inclusive of a haemogram and a complete metabolic profile were within normal limits. The patient also had complaints of bleeding on touch sometimes and bleeding on combing hair. The history of occasionally tender and itching was present. The clinical diagnosis of nevus sebaceous was made and the patient was advised incision biopsy of this ulcerated lesion. On histopathological examination Syringocystadenoma papilliferum was found, which is a benign skin adnexial tumour is.
Syringocystadenoma papilliferum is rare hamartomatous proliferative malformation derived from apocrine sweat glands of skin. About 50% are present at birth or appear during infancy and tend to proliferate around puberty [5]. It is nonmalignant adnexal sweat gland neoplasm characterized by asymptomatic, skin colored to pink papules, or plaques of highly variable appearance. Most common sites are head and neck region; however, tumors in other areas, such as vulva, external ear, lower leg, and scrotum have also been reported [6].

Three clinical types of Syringocystadenoma papilliferum have been described: [7] [8]

1. **Plaque type:** presenting as a alopecic patch on scalp and may enlarge during puberty to become nodular, verrucous or crusted plaques commonly tend to be associated with nevus Sebaceous of Jadassohn in one third of the cases[9].

2. **Linear type:** Consists of multiple reddish pink firm papules or umbilicated nodules 1-10 mm in size commonly occurring over face and neck.

3. **Solitary nodular type:** they are domed pedunculated nodules 5-10mm in size with predilection for trunk shoulder and axillae.

Syringocystadenoma papilliferum is commonly associated with hamartomas of follicular or sebaceous gland origin in one third of cases syringocyatadenoma papilliferum is associated with Naves sebaceous of Jadassohn.[10] In our case, the lesion was in the scalp. It usually appears at birth or during infancy and around the time of puberty. In about one-third of cases, syringocystadenoma papilliferum is reportedly associated with nevus sebaceous. Multiple
tumors of adnexal origin, such as trichoblastoma, apocrine adenoma, hidradenoma papilliferum, and trichilemmoma, are being reported to arise along with nevus sebaceous [11]. In this case, the tumor was neither present since birth nor associated with other tumors. Ulceration or a rapid enlargement is indicative of a malignant transformation. Syringocystadenocarcinoma papilliferum is a malignant counterpart of syringocystadenoma papilliferum [12]. The diagnosis is clinically suspected and histologically confirmed. Due to the risk of a malignant change, a prophylactic surgical excision, followed by a detailed histological examination, is the treatment of choice.

Kaddu et al.[13] in his study of 316 cases neoplasm arising in Naevus sebaceous of Jadassohn found 7.6% of benign and 2 cases of malignant cases all occurring in adulthood. Munoz-Perez et al. [14] in their series of 226 cases found 18% of benign tumors in subjects all above the age of 14 years. There are very few case reports or references of Syringocystadenoma papilliferum in Indian literature; courtesy the extreme rarity, under reporting and ignorance. Kumar V. et al. [15] in 1991 reported a single case of Syringocystadenoma papilliferum in a 10 year old girl. Golvalkar R.M. et al. [16] in 1994 reported a single case of Syringocystadenoma papilliferum arising in backdrop of Naevus sebaceous of Jadassohn in a 13 year old girl. Sood A. et al. [17] in 2000 reported 2 cases of Syringocystadenoma papilliferum in a 5 year old boy and 20 year old man with lesions in the axilla and neck respectively.

Regardless of the type, the histopathology of the lesion is usually the same and renders the same prognosis. Malignant transformation of this lesion is possible, usually to basal cell carcinoma in 10% of cases, uncommonly to squamous cell carcinoma and rarely to its malignant counterpart: Syringocystadenocarcinoma papilliferum[18].

Clinical diagnosis is, on most counts, not feasible. Due to the various presentations and possible differential diagnoses, histopathology would yield the most practical, as well as accurate option. Knowledge of this diagnosis would add a supplementary alternative to our clinical suspicions. Treatment for SCAP is surgical excision, which has a two-pronged benefit. Should this lesion undergo future malignant degeneration, excision sufficiently eliminates that risk. Secondarily, excision allows cosmetic relief for the patient, should it be a concern.

CONCLUSION

Syringocystadenoma papilliferum is a rare neoplasm, usually appears at birth, during infancy or around the time of puberty. It rarely appears in adults. In the present case, it was clinically diagnosed at first as nevus sebaceous of the scalp, but later, it was histological confirmed as syringocystadenoma papilliferum. Awareness of the various possible presentations of SCAP can prevent misdiagnoses and overtreatment. Presentation of this tumour may generate multiple differential diagnoses and it must be sent for a histopathological examination. Radiotherapy and other destructive procedures are ineffective and should be best avoided. Surgical excision is the only treatment of choice.

REFERENCES


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