Syringocystadenoma Papilliferum

I Bhuiyan[1], M S Hossain[2], Y Joarder[3], M Alam[4]

Abstract

Syringocystadenoma Papilliferum (SCAP) is a rare papilloma resembles to benign tumour. It is characterized by extensive papillary epithelial elements growing in to dermis. A 9 years old boy presented with slowly growing dome shaped plaque consists of clusters of rose-red like vesicles of firm consistent, smooth-surfaced papules over the occipital region of the scalp. There was no family history of such type of lesions. Histopathological examination showed few cystic invaginations from the epidermis into dermis. Papillary projections are extending from the base of the cyst into lumen of the cystic spaces that consistent with Syringocystadenoma Papilliferum (SCAP).The treatment of choice is surgical excision.

Key words : Syringocystadenoma; Papilliferum.

Introduction

Syringocystadenoma Papilliferum (SCAP) is a rare,benign adnexal neoplasm that may present at birth, appear during infancy or may present during puberty[1,2]. Fifty percent are present at birth or early childhood and another 15-20% develop before puberty [1].

SCAP appear as a solitary nodule, as a nodular plaque or a linear group of nodules[2,3]. At birth or during infancy, the plaque and linear varieties are common which during puberty become more elevated, verrucous or crusted, hyperkeratotic, papiliomatous.[1,2,3] Less common solitary nodular form usually develop during puberty.

The plaque form usually presents as a hairless area in the scalp, while the linear form commoner on the neck and face [2].

A mature lesion comprises clusters of rose- red papules of firm consistency, 2-10 mm in diameters, which are domed, umbilicated or pedunculated, often with a friable or crusted surface[4]. Vesicle like papules pinpoint to pinhead sizes are filled with clear fluid[4]. A small fistula draining clear, bloody or malodorous fluid may develop[2].

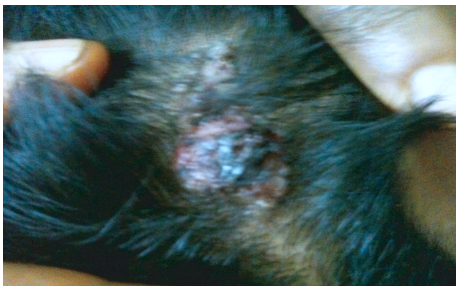
Case Report

In 10th February 2013, a 9 years old boy presented to the Department of Dermatology and Venereology, Shaheed Suhrawardi Medical College & Hospital, Dhaka, Bangladesh with a slowly growing dome shaped broad, bald plaque over the occipital region of the scalp. The patient stated that there was no other cutaneous or systemic abnormality nor any affected family members. The patient's mother stated that the lesion started with few papules, newer papules developed which evolved very slowly over last 4-5 yrs. All of them coalesced to form plaques. There was no itching and surrounding skin was normal.

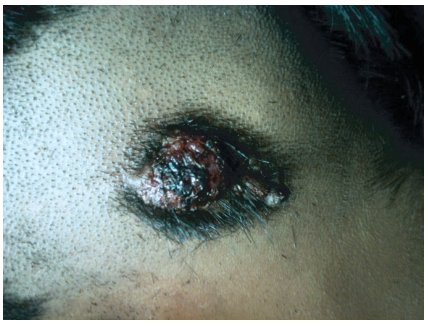
On examination the boy was apparently healthy with average body built and nutritional status.

The plaque was dome shaped consisting clusters of rose-red vesicle like papules of firm consistency, smooth surfaced, bald and few were pigmented.. There was no draining fistula.

Other systemic examination revealed no abnormality.



Single dome shaped plaque consists of cluster of rose red vesicle like lesion situated over the occipital region of scalp with hair



Single dome shaped plaque consists of cluster of rose red vesicle like lesion situated over the occipital region of scalp with out hair

Investigation revealed ESR-08mm in Ist hour, Hb%-12.1 gm/dl, WBC-7200/cc, Neutrophil-58%, Lymphocyte 32%, monocyte-02%. Eosinophil-08%, Platelet-370000, SGPT-21 u/L, Serum creatinine 93mmole/L, Urine R/M/E-normal.

[1] Dr. Ishrat Bhuiyan,
MBBS; DDV, FCPS
Assistant Professor
Dept. of Dermatology & Venereology
Shaheed Suhrawardi Medical College
Dhaka

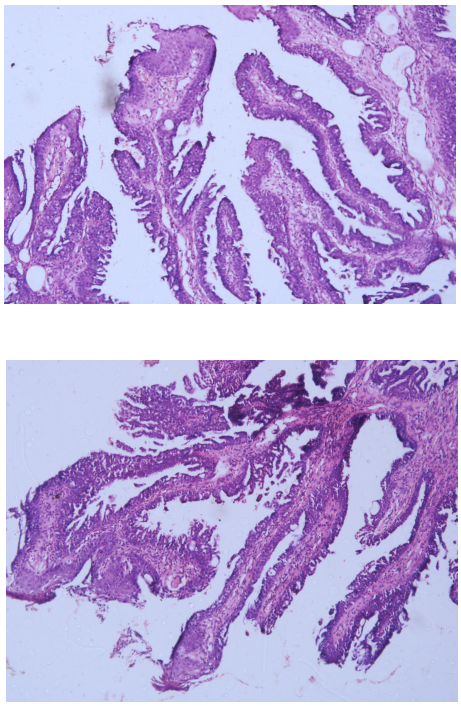
[2] Dr. Md. Shahadat Hossain
MBBS; DDV, FCPS
Associate Professor
Dept. of Dermatology & Venereology,
Shaheed Suhrawardi Medical College
Dhaka

[3] Dr. Yasmin Joarder
MBBS; DDV
Assistant Professor
Dept. of Dermatology & Venereology,
Northern International Medical College

[4] Brig. Gen. Dr. Mahbubul Alam
Classified Specialist &
Professor of Pathology
Armed Forces Medical College (AFMC)

Correspondence
Dr. Ishrat Bhuiyan,
Assistant Professor
Dept. of Dermatology & Venereology
Shaheed Suhrawardi Medical College
Dhaka
Email: ishratskin@yahoo.com

Histopathological examination showed few cystic invaginations from the epidermis into dermis. Papillary projections are extending from the base of the cyst into lumen of the cystic spaces that consistent with Syringocystadenoma Papilliferum (SCAP).



Histopathological features of the lesion (Syringocystadenoma Papilliferum) of the patient

Discussion

The SCAP, a rare, benign epidermal adanexal neoplasm presents as a solitary nodule or nodular plaque or linear group of nodules.

The commonest site is scalp and face. Less frequently involved sites are neck, face, trunk, arms or genital areas. At birth found on scalp and face. Approximately 25% arise on trunk and genital area or inguinal region during adolescence.[1,2,5]

The site of origin of Syringocystadenoma Papilliferum (SCAP) is quite controversial. Some suggest that Syringocystadenoma Papilliferum (SCAP) has an apocrine gland origin, but multiple studies have found that 90% of these lesions occur in areas devoid of apocrine glands. Lever and Schaumburg –Lever postulated the presence of a pluripotential cell which may give rise to these lesions. Helwig and Hackney postulated the presence of an intermediate cell between the two types. So, all of above findings explain the features of both gland types in Syringocystadenoma Papilliferum (SCAP) [4].

For evoluation of Syringocyst adenoma Papilliferum(SCAP) mutations in PTCH or P16 tumour suppressor gene may play role.[1,2,3].

Syringocystadenoma Papilliferum (SCAP) has been seen in conjunction with a wide variety of adnexal tumours [1].

In about one third of cases Syringocystadenoma Papilliferum (SCAP) is associated with nevus sebaceous. Basaloid epithelial proliferation resembling basal cell carcinoma has been found in association with SCAP about 10% of time, but noted only in lesions that also exhibit a nevus sebaceous.Few instances of transition of a (SCAP)into adeno carcinoma with regional lymph node metastases have been reported. [2, 3, 4]

Histopathologocally, SCAP is characterized by one or several endophytic invagination of the glandular epithelium into the dermis. The upper portion of the invagination is lined by squamous keratinizing cells similar to those of the surface epidermis. Numerous papillary projections extend into the lumina of the lower portion of invagination, both of which lined by glandular epithelium often consisting of two rows of cells, outer portion with cuboidal cells with round nuclei, scanty cytoplasm and Luminal high columnar cells with oval nuclei,faintly eosinophilic cytoplasm. Beneath the cystic invagination, deep in the dermis groups of tubular glands with large lumina have a typical connection with the above invagination. Almost invariable presence of a fairly dense cellular infiltrate composed nearly entirely of plasma cells in the stroma of this tumour, specially in the papillary projections,is a highly diagnostic feature. These plasma cells(which are in the glandular stroma) are predominantly of Immunoglobulin G(IgG), IgA classes [1,3,4,5,]

Tumour cells stain positively for carcinoembryogenic antigen.[1,2,3].

Treatment of this lesion usually consists of complete surgical excision, as it has malignant potentials. The anatomic areas unfavourable for excision or grafting, CO_2 Laser may be an alternative option[4].

Conclusion

Though rare, SCAP has chances into malignant transformation (rapid growth, ulceration, pain & pruritus). Therefore early detection and proper curative measures should be taken.

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