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Dermatology

LINEAR SYRINGOCYSTADENOMA PAPILLIFERUM: RARE TUMOR ON A RARER SITE

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ABSTRACT Syringocystadenoma Papilliferum is a rare entity arising from apocrine or eccrine sweat gland. It is usually associated with sebaceous nevus of Jadassohn. Most common site is the scalp and the head and neck region. Either it is present since birth or presents at puberty. Here we present a rare a case of this rare entity which is present over a rare site, face and at and rarer age.

KEYWORDS : syringocystadenoma papilliferum, hamartoma, sebaceous nevus

INTRODUCTION

- Naevus Syringocystadenomatosus Papilliferus
- SP (Syringocystadenoma Papilliferum)
- Syringadenoma Papilliferum

Syringocystadenoma Papilliferum is a hamartomatous condition of apocrine or eccrine sweat gland. Commonly seen at birth or at puberty.[1] In about 50% of those who are affected, it is present at birth, and in a further 15%-30%, the tumour develops before puberty[2]. It is usually associated with sebaceous nevus of Jadassohn. Most common site is the scalp and the head and neck region. Rarely it may undergo malignant transformation which is seen as rapid increase in size of the lesions or lymph node enlargement or development of ulceration or nodules over the existing lesion. It occasionally co-exists with other tumours such as basal cell carcinomas and verrucous carcinomas[3]

We are presenting a case with rare age and site of presentation which was histologically diagnosed as syringocystadenoma papilliferum.

CASE REPORT:

A 70 years old married male patient who presented with complain of few, asymptomatic raised, soft skin coloured lesions present over the right side of the face near lower eyelid since last 2-3 years. These lesions were insidious in onset that developed over normal looking skin, not preceded by any skin lesions. They gradually increased in size and number initially for a year, after which they remained stagnant, without further change in size or number as well as any morphological characteristics for the last 1.5 years. The patient denied history of any discharge or bleeding, ulceration, pain or itching over the lesions. No other remarkable cutaneous or extracutaneous finding was noted. Patient had no significant past medical history.

On cutaneous examination,

There are a few, well demarcated, discrete, skin to brown coloured, pedunculated nodules arranged in a linear pattern present on the right infraorbital area of face parallel to the nasolabial fold.

On palpation the lesions are firm and non tender. Base is not indurated and lesion not fixed to deeper tissue. There is no regional lymphadenopathy. No secondary changes of the overlying skin. No other skin lesions were noted elsewhere on the body or scalp.

Clinically, the following differential diagnosis were made: syringocystadenoma papilliferum , papillary eccrine adenoma, giant achrocordon, segmental neurofibroma.

Excisional biopsy of the lesions was advised. And patient underwent excision of lesion under local anaesthesia.

The lesion was excised completely with normal 1 cm margin around the lesion and was send for histopathological examination.

Tumor shows a prominent invagination arising from epidermis; the invagination is lined by two layers of cuboidal cells in lower part and squamous epithelium in upper part. Cystic spaces open into this invagination. Lining epithelium of the cystic space shows papillary projections into the cystic spaces. Surrounding dermis shows dense plasma cell infiltrate. No evidence of malignancies was detected.

Based on these features the diagnosis of syringocystadenoma papilliferum was made.

DISCUSSION:

Syringocystadenoma Papilliferum is a rare condition. It was earlier known by the names of Adenoma cystoma intracanaliculare.[4] It is a hamartomatous condition of either the apocrine or eccrine sweat gland. It has been reported due to mutation of PTCH and p16[5]. It presents as asymptomatic skin coloured papules, plaque or nodules. Most common location where it can arise is the head and neck. This condition usually appears at birth or at puberty and is associated with nevus sebaceous.

Clinically, three variants are described. These are: plaque type, linear type, nodular type.The plaque type is seen over scalp. It is associated with nevus sebaceous. It presents as alopecic patch that enlarges during puberty to become verrucous, nodular, or crusted plaques. The linear type presents as multiple reddish, fi rm papules, or umbilicated nodules 1-10 mm in size. Mainly seen in face, head and neck region.The nodular type presents as raised domed pedunculated nodules 5-10 mm in size. They can be solitary or multiple and usually occur on the chest, back, shoulders, and arm pits.[6]

Multiple tumors of adnexal origin, such as trichoblastoma, apocrine adenoma, hidradenoma papilliferum, and trichilemmoma, are being reported to arise along with nevus sebaceous.[7]. Basal Cell Carcinoma (BCC) development has been reported in upto 10% of the cases . Only two cases of squamous carcinoma have been reported. Syringocystadenocarcinoma papilliferum is the malignant counterpart, characterized by solid areas and cytologically malignant cells.[10]

Immunohistochemistry helps in differentiating the origin of the tumor (ie- either eccrine or apocrine), but it has no clinical significance. If it is apocrine in origin, it would be positive for protein15, protein24 and zinc 2 glycoprotein. If it is eccrine in origin, it would be positive for CK.

Treatment is excisional biopsy. It also helps in confirming the diagnosis. Other modalities are CO2 laser.

Salient features of our case include a later age of onset, de novo occurrence without any preceding lesions. Rarer variant- multiple, linear, nodular. Rare site of occurrence- localized to infraorbital fold. Histopathological examination confirmed the diagnosis as well indicated absence of secondary malignant changes.

Pitfall was as follows- IHC was not done due to lack of infrastructure.

CONCLUSION:

Syringocystadenoma Papilliferum is uncommon sweat gland tumour with widely varying clinical appearance. It is called as childhood tumour as it appears at birth, or during infancy or during puberty.[1] It should be histologically confirmed. Surgical excision and reconstruction is the treatment of choice. There is a risk of malignant change.

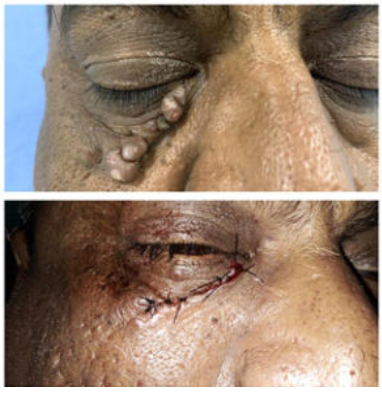

Fig 1 few, well demarcated, discrete, skin to brown coloured, pedunculated nodules arranged in a linear pattern present on the right infraorbital area of face parallel to the nasolabial fold

Fig 2
After excision of the lesion under local anaesthesia.

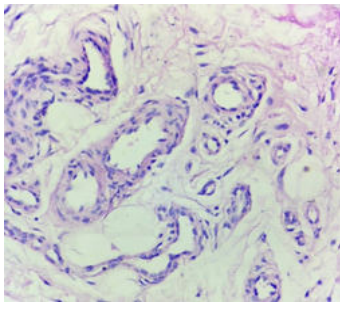

Fig 3 Histopathological examination of the excised lesions

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