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A Rare Case Report of Syringocystadenoma Papilliferum

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Abstract: ***Background: Syringocystadenoma papilliferum is the rare benign hamartomatous adnexal tumour.50% cases are reportedly present at birth while 15-30% are present during puberty. Case Report :A 70 yr old male patient presented with swelling over scalp region since birth, which was clinically and radiologically diagnosed as infected dermoid cyst. Excision and biopsy was done. Histopathological examination revealed as Syringocystadenoma papilliferum. Discussion: Syringocystadenoma pappiliferum is rare non malignant adenexal sweat gland neoplasm characterized by asymptomatic ,skin coloured to pink papules or plaques of highly variable appearance. Conclusions: Syringocystadenoma papilleferum is an uncommon sweat gland tumor with widely variable clinical appearance. It typically develops as nodular plaque lesion and is reportedly associated with nevus sebaceous in 40%cases.***

Keywords: Syringocystadenoma papilleferum, adnexal tumor, nevus sebaceous.

1. Introduction

Syringocystadenoma papilliferum is a rare benign hamartomatous adnexal tumor. Fifty percent of the cases are reportedly present at birth while 15-30% are present during puberty.The tumor has varied clinical presentation. It presents as a hairless area on the scalp and is said to be associated with sebaceous nevus of Jadassohn. Malignant change is very rare and is indicated by rapid increase of size and/or change in appearance of metastatic lymph nodes. With increasing size, a prominent papillary configuration develops and the surface can become scabbed, which is typical of this tumor.

HPE is confirmatory and immunohistochemistry is required only for further differentiation between apocrine and eccrine tumors.

2. Case Report

A 70 year old male presented with complaints of swelling over the scalp since birth with gradual increase in size,c/o bleeding on touch since 10 days and bleeding on combing hair. The history of pain and itching was also present. O/E : A Swelling of about 4*4cm(figure1) was noted over the scalp slightly towards left of midline in parieto-occipital region, with irregular surface, and ulcerative lesions with blood discharge, firm in consistency, No regional lymphadenopathy .Clinically it was diagnosed as Dermoid cyst, which was confirmed by USG of swelling as infected dermoid cyst. Clinically it was diagnosed as dermoid cyst over the scalp. The patient underwent excision of the lesion under local anesthesia. The lesion was excised completely and was sent for histopathological examination.

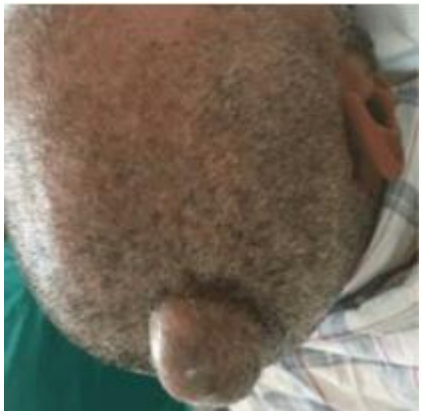

Figure 1

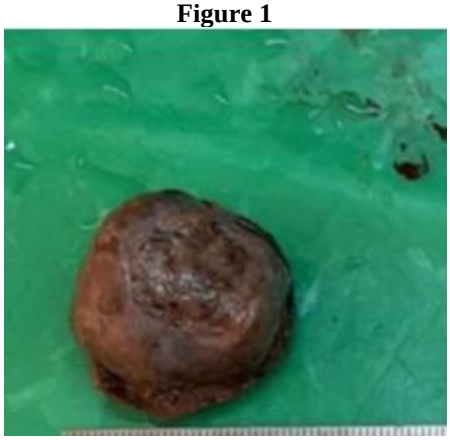

Figure 2

Figure 3

A gross examination showed skin covered gray brown specimen measuring 4.5*4*2 cm(fig 2).Cut section showed grey white to grey brown areas, cystic spaces(fig 3).Sections studied showed ulcerated epidermis with accumulation of acute and chronic inflammatory cells. The main dermal lesion consists of large cystic spaces with prominent papillary infolding. The cells are brightly eosinophilic. At places solid cell nests noted and foci of stromal permeation noted. Concentric calcified bodies resembling psammoma bodies noted. Associated focus of pigmented seborrhoeic keratosis noted. Based on these features the diagnosis of Syringocystadenoma papilliferum was made.The patient was followed up for 6 months and showed no evidence of recurrence.

3. Discussion

Syringocystadenoma papilliferum is rare 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, tumor in other areas, such as vulva, external ear, lower leg, and scrotum, have also been reported.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.

It reportedly evolves within three stages:

a) Infantile stage: It appears as alopecic orange yellow plaque.
b) Adolescent stage: Under androgenic influence the plaques undergoes various changes such as hyperkeratosis, hyperpigmentation, and sebaceous gland formattion.
c) Adult stage: It is characterized by the presence of large sebaceous glands, ectopic apocrine glands, and epidermal hyperplasia.

During the period of adult stage, variety of benign or malignant lesions can develop.Approximately, one-third of the cases are said to arise in precursor lesions such as organoid nevi. Yamamoto et al. reported the origin of pluripotent cells.

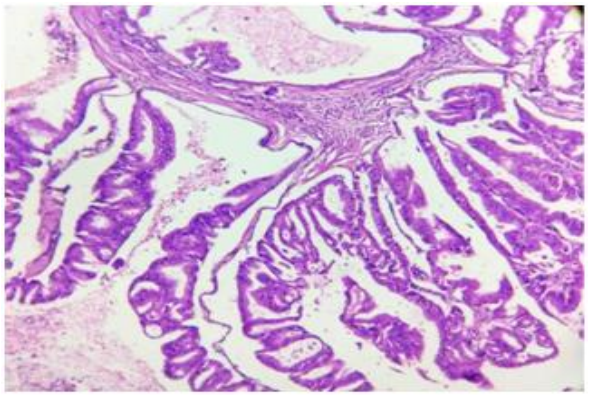

Figure 4

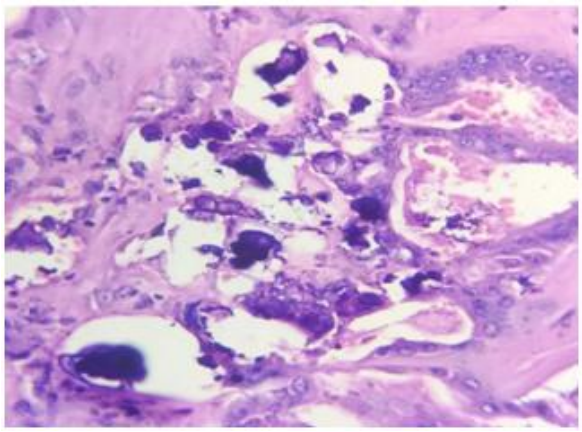

Figure 5

Histopathology (fig.4, 5) typically shows varying degrees of papillomatosis along with cystic invaginations and malformed sebaceous glands. Immunohistochemistry helps in differentiating the origin of the tumor, i.e. either eccrine or apocrine, but is of no clinical significance. Positive immunoreactivity for proteins 15 and 24 and zinc-2 glycoprotein demonstrates evidence of apocrine differentiation, while positivity for CKs demonstrates eccrine differentiation. Immunohistochemistry was not done in the present case. The only treatment for syringocystadenoma papilliferum is excision biopsy that also helps in confirming the diagnosis. Other modalities rarely used for treatment are CO2

4. Conclusion

Syringocystadenoma papilliferum is an uncommon sweat gland tumor with a widely variable clinical appearance. The presentation of tumor may generate multiple differential diagnosis, thus it must be histologically confirmed.Radiotherapy and other destructive procedures are ineffective and should be best avoided. Surgical excision with reconstruction is the treatment of choice.

This is being reported for its rarity.

References

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