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Case Report

Nevus sebaceous of Jadassohn of face with infundibular and keratinous cyst in an adolescent Arab male-a rare case report

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ABSTRACT

Nevus Sebaceous of Jadassohn is rare hamartomatous skin lesion. We report an interesting case of an Adolescent Arab male 18yr old, who presented to us with a velvety plaque of 7cm length in his left preauricular area. Excision biopsy revealed Nevus sebaceous of Jadassohn with infundibular and keratinous cyst. The lesion is present since birth started as a tiny lesion and grown to the current size. No such large sized lesion on the face has ever been reported in the medical literature till date. We advise an early excision as they have strong potential for malignant change.

Keywords: Congenital skin lesion, Hamartomatous skin lesion, Nevus sebaceous

INTRODUCTION

Josef Jadassohn in 1895 first described Nevus sebaceous as a circumscribed hamartomatous lesion, predominantly composed of sebaceous glands.[1] Pinkus designated this disease organoid nevus, as the changes are not confined only to the sebaceous glands but also involve proliferative changes of sweat glands and hair follicles.[2] NSJ is a rare skin lesion, associated with cosmetic deformity ,malignancy and has incidence of 0.3% in newborns.[3] There was profound proliferation and hyperplasia of the lesion happens at puberty and this leads to larger and verrucous appearance of the lesion.[3]

METHODS

Arab male of 18yr old, Oman nationality, body mass index of 45kg/m^2 presented to us with a tear drop shaped velvety plaque like lesion in the left preauricular area. The lesion was present since birth, started as a small lesion, enlarged slowly and reached the current size of 7cm in length. On examination, the lesion was linear, velvety plaque like lesion situated in the left preauricular area and narrows down with a ‘’ rat tail” like appearance behind the external ear. The lesion was cosmetically disfiguring one.

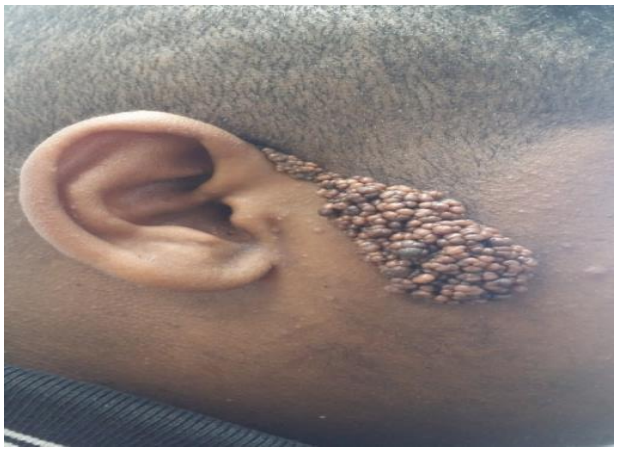

Figure 1: External appearance of the lesion.

The lesion was freely mobile and was soft in consistency in nature. There was no evidence of ulceration, bleeding

or regional lymph node enlargement (Figure 1). He underwent Excision under Local Anesthesia. Histopathology revealed epidermis showing marked epithelial hyperplasia, hyperkeratosis, and florid papillomatosis and dilated keratin filled Infundibular cyst (Figure 2).

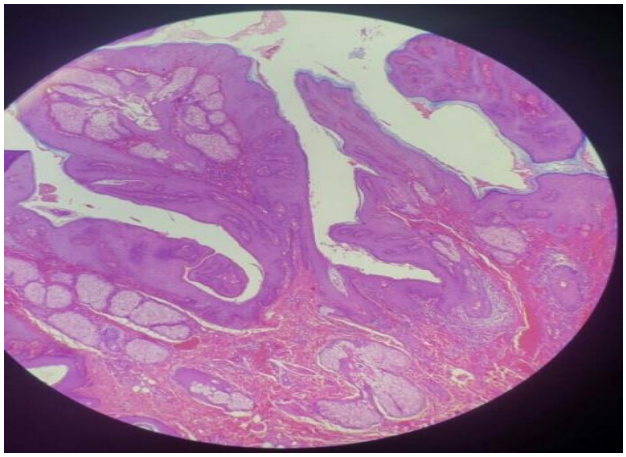

Figure 2: HPE picture shows epidermis showing marked epithelial hyperplasia, hyperkeratosis and florid papillomatosis.

Stroma showed sebaceous gland hyperplasia, immature hair follicles, ectopic apocrine glands and lymphoplasmocytic infiltrates. Keratinous cysts were also present but there was no evidence of any malignancy (Figure 3).

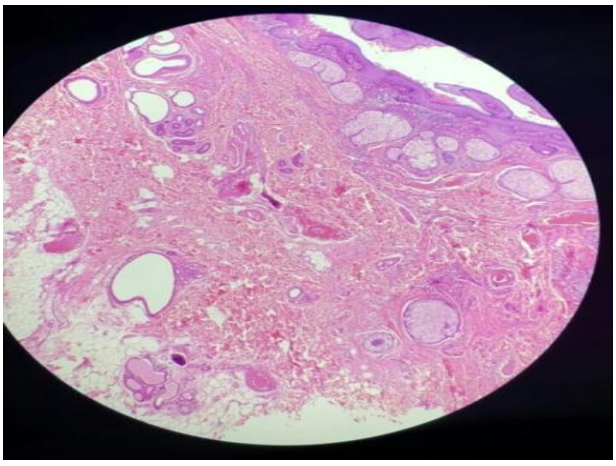

Figure 3: HPE picture shows hair follicles in apocrine glands.

DISCUSSION

Our specimen is fairly large size of 7x2.5 cm and such large sized sessile NSJ of face is not reported in the past. Moreover, an eight-month-old infant with a pedunculated NSJ 10x8 cm lesion in face and underwent excision has been reported.[4] Our lesion is sessile and patient is in adolescent age group.

Trichoblastoma and syringocystadenoma papilliferum are the commonest benign tumors associated with NSJ but in our case, only infundibular and keratinous cysts were present which is rare.[5] Malignant changes like basal cell carcinoma and squamous cell carcinoma are possible with a lifetime risk of 5-22%.[6] The youngest case report of malignant change to basal cell carcinoma was reported at the age of seven.[7] There are several benign and malignant tumors can be associated with NSJ. Few are sebaceous carcinoma, basal cell carcinoma, trichoadenoma, sebomatricoma, apocrine gland cyst, trichoblastoma and syringocystadenoma papilliferum.[8]

CONCLUSION

Our case is unique since such a large lesion of NSJ of the face has not been reported in the literature till date. Moreover, the association of benign lesions like infundibular cysts and keratinous cysts are also specific. We also emphasize the medical fraternity for an early excision of NSJ as there is evidence of malignant changes occurring even in childhood.

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