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Nevus Sebaceous On Shoulder: An Unusual Location

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Abstract*: Sebaceous nevi are epidermal hamartomas commonly seen over scalp and face. We report a case of sebaceous nevus with unusual presentation at an unusual site. This form of nevus sebaceous is quite rare in contrast to relatively common linear variant.*

Conclusion: *Sebaceous nevus is rare on shoulder. Unusual location, nodular papillomatous presentation with linear extension lead to misdiagnosis. Undifferentiated hair follicles, sometimes with dilated infundibula are typical of sebaceous nevus. The divergent differentiation observed in NS is consistent with the common embryologic origin of the folliculo sebaceous apocrine unit.*

Keywords: *Nevus Sebaceous (NS), Shoulder, Nodular presentation*

Key Message: *Nevus Sebaceous of Jadassohn can also occur on sites other than face and scalp, and presentations can be varied like cerebriform, nodular or along the blaschko lines. So Histopathological examination aids in the confirmation of diagnosis.*

I. Introduction

Nevus sebaceous, also known as nevus sebaceous of Jadassohn, commonly thought of as a sebaceous malformation, But in truth it is a hamartoma of the skin and its adnexa,. It Exhibits varying degrees of abnormalities in epidermal, follicular, sebaceous, and apocrine glands. The condition was first described by Josef Jadassohn, a German dermatologist. As the lesion involves more than a single component it is also called as organoid nevus. It is sporadic and seen in 3/1,000 neonates. NS usually presents as a well demarcated, yellow orange, alopecic, velvety plaque. Most commonly on scalp(59.4%) and face(32.6%). Other common sites are preauricular area(3.8%) & neck(3.2%). Sebaceous nevi are rare on trunk and extremities, and only three cases are reported so far (leg). In cases where it is associated with other developmental defects; it is called as epidermal nevus syndrome. Here we report a case of nevus sebaceous with rare nodular presentation on a rare site (shoulder).

II. Case Report

A 10 year old female child Radhika, resident of Garika village came with a chief complaint of a mass over right shoulder since 4 years of age.
The present complaint started as a tiny skin coloured flat lesion on right shoulder at birth.
It gradually increased in size since 4 years of age.
Similar lesions developed in the adjacent area and gradually extended linearly towards the elbow in due course.
Mass was asymptomatic except for occasional foul smelling watery discharge on manipulation which was associated with itching and subsided with medication.
No h/o seizures.
No delayed milestones
Patient's performance at school is normal
Patient did not attain menarche at the time of presentation.

On Examination
Multiple skin coloured papillomatous growths of size ranging from 4x3 cm to 1x1 cm present on right shoulder.
Yellowish crusting seen on the surface.
With surrounding extensions on shoulder & linear extension towards elbow

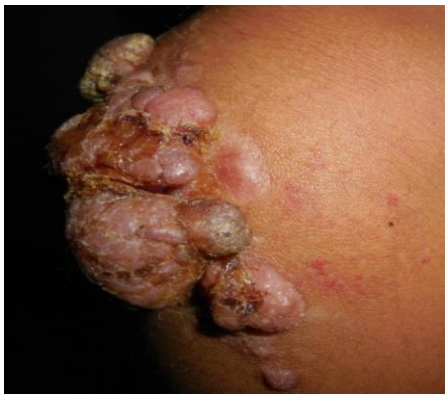

Figure 1a

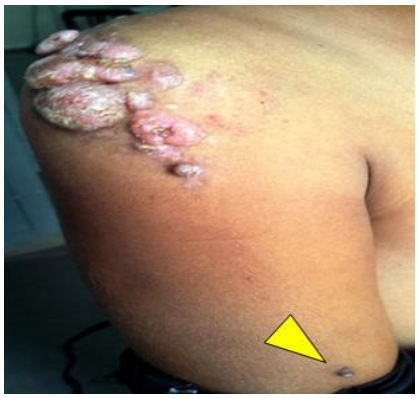

Figure 1b

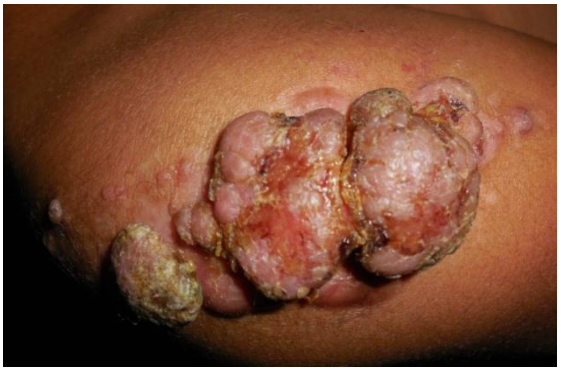

Figure 1c

On basis of linear pattern and papillomatous growths with discharge, we diagnosed it to be Syringo cystadenoma papilliferum (or)
Lymphangioma circumscriptum

Investigations:
Routine investigations were within normal limits.
Systemic examination revealed no significant abnormality.

Histo pathological examination
Epidermis showed epidermal hyperplasia with papillomatosis,
Acanthosis and follicular plugging
Dermis showed numerous eccrine glands with few apocrine glands
And an undifferentiated hair follicle, which is typical of a nevus sebaceous

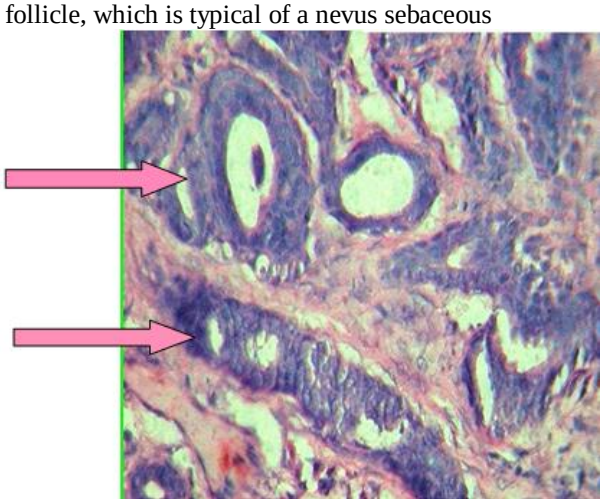

Figure 2

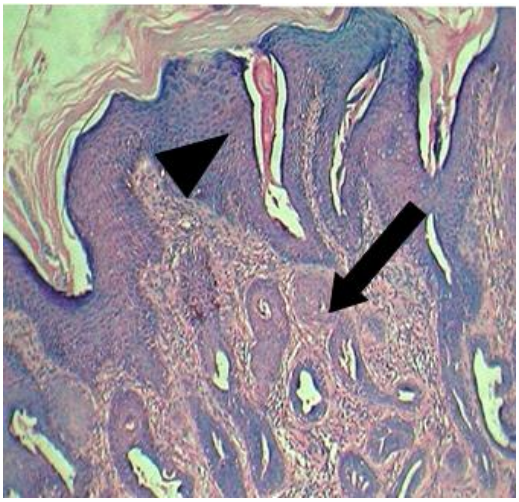

Figure 3

Figure 1 a, 1b and 1c. multiple skin coloured papillomatous growths on shoulder with linear extension towards elbow
Figure 2. shows undifferentiated hair follicle and an abortive hair follicle
Figure 3. epidermal hyperplasia and papillomatosis and follicular plugging
Dermis shows numerous sweat glands and apocrine glands

III. Discussion

Nevus of Jadassohn also called organoid nevi are hamartomatous proliferations. They are usually sporadic, with an incidence of 0.3%. [(1)] There is no sex predilection and no racial difference. Sporadic incidence is postulated to be due to post zygotic somatic mutations.[(2)] It usually appear at birth or early in life as a pinkish, yellow or orange tan plaque and around puberty becomes verrucous and nodular., common sites are face and scalp. There may be deviations from this classical description, Less commonly it can present on rare sites or with a atypical morphology.

Kanekura et al., first reported a case of sebaceous nevus on leg. [(3)]

Ugras et al., reported a case of sebaceous nevus on vulva, [(4)]

Dr.Iffat hassain reported a case of nevus sebaceous in nasal cavity. [(5)]

Cerebriform sebaceous nevi are reported for the first time in 1998 by Ramesh et al., [(11)] later by Bomztyk et al., [(11)] Correale et al reported 5 case of large, papillomatous, and pedunculated sebaceous nevi. All of their patients presented at birth with large exophytic lesions, all but one were confined to the scalp and the face. [(11)] Only one patient had extensive NS extending from scalp to the elbow. None of the patients showed any associated abnormalities. They concluded that whereas the patients presenting with large cerebriform and papillomatous Sebaceous Nevi over the scalp should be evaluated for neurologic and ophthalmic abnormalities, they do not usually reveal any underlying abnormality. The reason for nodular and papillomatous presentation of Organoid Nevus is not known, but as Nevus Sebaceous has some neoplastic potential, Various tumor genes have been implicated in the aetiology,

Xin et al., suggested constitutive activation of patched-hedgehog pathway and a loss of heterozygosity at PTCH locus 9q22.3., [(5)] which is the gene implicated in BCC (basal cell carcinoma) or GORLINS SYNDROME. Presumably nevus sebaceous develops from pleuripotent primary epithelial germ cells. Our patient presented with multiple papillomatous nodules progressing gradually. On basis of clinical presentation, we made a diagnosis of syringo cystadenoma papilliferum, the commonest secondary neoplasm in NS, and a punch biopsy specimen was sent for Histo Pathological Examination. HPE revealed the typical features of a nevus sebaceous, the abortive hair follicles, and some apocrine glands. Microscopic features of sebaceous nevi varies with age of the patient. During childhood, sebaceous glands are under developed and reduced in size and number. However, presence of cords and buds of poorly differentiated epithelial cells which represent primordial hair follicles is typical. Mature sebaceous glands appear during puberty or sometimes at an earlier age. Before puberty the sebaceous and apocrine glands in the dermis are sparse and under developed and at around puberty these glands try to proliferate. In our patient, the proliferating cells of sebaceous or apocrine glands in the pre-existing lesion stimulated by repeated minor trauma might have acted as a multi potential germinative cells giving rise to development of nodular proliferations.[(9)] The patient was referred to the department of plastic surgery where she underwent wide excision under general anesthesia with skin grafting,

and followed up for 1 year. The present case was peculiar because of the morphology of the lesions and their pattern of distribution.

IV. Conclusion

Nodular presentation may represent a rare variant of Nevus sebaceous, although the factors that govern morphology of nevi are not yet known. It should also be emphasized that Histo pathological examination should be done in all suspected cases to confirm the diagnosis.

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