Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2015; 3(7):611-613 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)

Cerebriform Nevus Sebaceous of Jadassohn – A Rare Case Report

Dr. N. Ezhil Arasi*[1], Anunayi. J[2], Dr Aravinda. M[3]
[1]Professor and HOD, Upgraded Department of pathology, Osmania Medical College, Telanganaa State, India
[2]Associate professor, [3]Senior Resident, Department of pathology, Osmania Medical College, Telanganaa State, India

***Corresponding author**
Dr. N. Ezhil Arasi
Email: drlotus26@gmail.com

Abstract: Nevus sebaceous also known as Nevus sebaceous of Jadassohn [NS] is an epidermal nevi predominantly congenital sebaceous hamartoma with an estimated incidence of 0.3% in the neonates. It is composed of sebaceous glands, commonly presents as single yellowish plaque over the head and neck. Cerebriformtype is a very rare morphological variant of NS. There is equal sex incidence. This case is presented because of its rarity in a four days old male child.
Keywords: Nevus sebaceous, Sebaceous glands, Plaque, Neoplasms.

INTRODUCTION

Cerebriform NS is an atypical, rare variant of NS, first described in a 20-year-old man by Ramesh et al [1]. Subsequent reports included Bomsztyk et al in a 6-month-old child [2] and by Correale et al in 5 neonates [3]. Causative factors of the cerebriform appearance are still not clearly understood . NS is a hamartomatous composite lesion recognizing epidermal, sebaceous, apocrine, and immature hair follicular elements and diagnosis is usually made clinically based upon its typical appearance and position (most frequently on the scalp, 59.3%; face, 32.6%; preauricular area, 3.8%; neck, 3.2%; locations off the head and neck, 1.3%) [4]. The condition was first described by Josef Jadassohn, a German dermatologist, and now bears his name [5]. Nevus sebaceous occurs in approximately 0.3% of all newborn infants [6].

CASE REPORT

A four days old male child was brought to Dermatology OPD with the complaints of raised pigmented lesion over the scalp on lefttemporo parietal region since birth measuring around 5x4 cms. Pregnancy and delivery were uneventful. No siblings. Parents gave history of a plaque present over the scalp at birth which gradually increased to present size which was cerebriform in appearance. There was no history of trauma. There was no hair growth at the area of interest. There was no significant family history.

On examination a large multilobulated, verrucous lesion was seen on the scalp. The lesion was painless, dry, and soft on palpation. The lesion's appearance was consistent with that of cerebriform nevus sebaceous (NS), a quite rare variant of the more common “classic” NS. A skin biopsy was taken and was sent for the histopathological examination. Grossly a single grey white skin covered soft tissue bit was received.

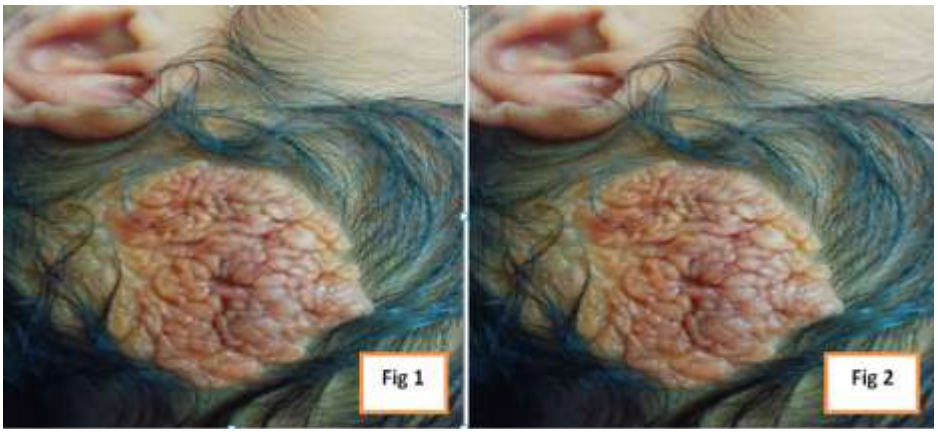

Fig 1 &2: Raised plaque over Left temporo parietal region. Hair growth is absent at the area of interest.

Histopathological examination revealed surface squamous epithelium thrown into folds .There was hyperkeratosis, papillomatosis, and horn cyst formation. Dermis showed few pilo sebaceous units. Consistent with Cerebriform Nevus Sebaceous of Jadassohn.

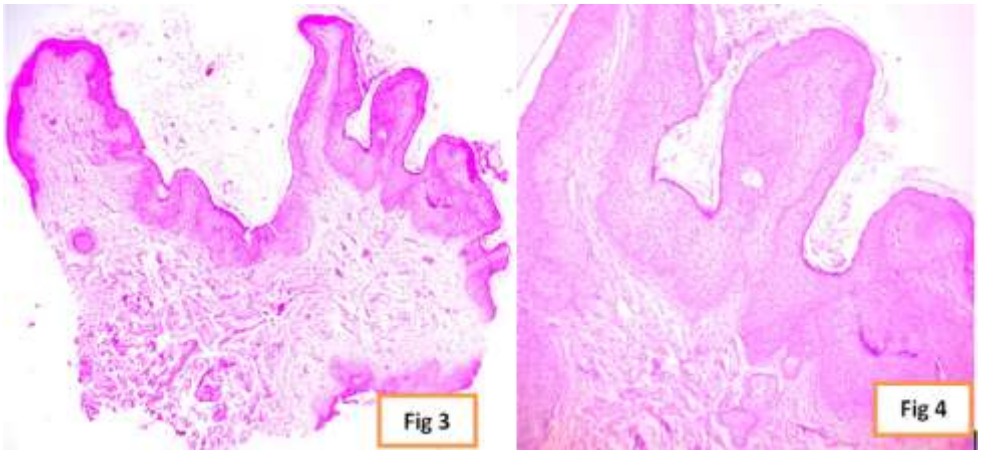

Fig- 3 H&E : Squamous epithelium thrown into folds with horn cyst formation.
Fig- 4(10x) H &E : Papillomatosis with pilosebaceous units.

DISCUSSION

Nevus sebaceous, a hamartoma of the skin and its adnexa, is characterized by epidermal, follicular, sebaceous, and apocrine gland abnormalities. Nevus sebaceous of Cerebriform type is a very rare morphologic variant of NS. These lesions have a predilection for the scalp and characteristically appear early in life as a solitary, hairless patch or small plaque. Often they do not cause problems until the patient reaches adolescence, as hormonal factors induce a verrucous or nodular change and the lesion grows in size, occasionally rather dramatically. Recently, it has been shown nevus sebaceous is caused by post zygotic mosaic mutations in the HRAS and KHAS genes [7].

Presumably, nevus sebaceous develops from pluripotent primary epithelial germ cells which have the potential to differentiate into various neoplasms. Deletions in the PTCH tumor suppressor gene on chromosome 9p22.3 may account for the neoplastic potential of the hamartoma [8]. PTCH gene deletion is proposed mechanism for development of nevus sebaceous.

Nevus sebaceous is usually solitary, although multiple and widespread lesions have been reported [9]. The lesion can be aesthetically unappealing, especially when it occurs on the face. Large or extensive lesions may be associated with developmental deficits [10].

The incidence of these tumors increases with age, particularly after puberty [11]. Majority of these tumors are benign; less than 1% of nevus sebaceous is complicated by malignant tumors [12]. Neoplasms occur mostly in the fourth decade of life in approximately 10 to 30% of lesions [13,14]. Given the size and location of the lesion and the risk of malignant transformation, especially to basal-cell carcinoma, later in life.Other malignant tumors include squamous cell carcinoma, apocrine carcinoma, ductal adenocarcinoma, porocarcinoma, anaplastic adnexal carcinoma, trichilemmal carcinoma, syringomatous carcinoma, and sebaceous carcinoma The differential diagnosis includes epidermal nevus, aplasia cutis congenita, solitary mastocytoma, and juvenile xanthogranuloma.

CONCLUSION

A new phenotype, known as cerebriform nevus sebaceous, has been described. Cerebriform nevus sebaceous is characterized by large, pedunculated or verrucous, pink, hairless, nodules or tumors in newborn. The diagnosis is usually clinical based on the characteristic features. A tissue biopsy or referral to a dermatologist should be considered if the diagnosis is in doubt. Excision of the lesion may be considered at any age for cosmetic reasons.

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