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Original Research Paper

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NEVUS SEBACEOUS : A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT Nevus sebaceous (aka. Nevus Sebaceous of Jadassohn or Organoid nevus) is a hamartomatous disorder of the skin and is an epidermal appendageal nevus disorder with predominant sebaceous gland involvement .The Diagnosis of which is mainly based on the clinical presentation ,histopathology and dermoscopy can be useful when the diagnosis is in doubt .Careful clinical follow up followed by a therapeutic excision might suffice but a prophylactic full thickness complete excision is recommended as there are rare chances of malignant transformation with chances of recurrence , metastasis and fatal outcome .In this article we report a case of nevus sebaceous along with a review of literature.

KEYWORDS : Nevus Sebaceous, Hamartomatous, Epidermal appendageal nevus, Sebaceous gland, Prophylactic full thickness excision.

INTRODUCTION :

The term sebaceous nevus was first coined by Jadassohn in 1980's. Nevus sebaceous (syn: Nevus sebaceous of Jadassohn/ Organoid nevus) is an epidermal appendageal nevi.

The term "nevus sebaceous" is considered a misnomer by some, who instead prefer to call it an "organoid nevus"(the term first coined by Mehregan and Pinkus) as it is considered to be a hamartoma with congenital malformations of pilo sebaceous unit with predominant sebaceous gland involvement .It most commonly affects the scalp (more than 50 % involvement) ,than the face ,the preauricular area ,neck, trunk ,oral or genital mucosa .

Case Report :

A 14-year-old boy born of non- consanguineous marriage came with the complaints of asymptomatic raised lesion over the left parietal region since birth. The lesion was static in progression until 6 months back when it started expanding. There was no history of other cutaneous or systemic involvement. There was no history of similar complaints in the family. His developmental milestones were normal.

On examination: A single well defined, orange-brown 4 X 2 cm elliptical plaque with irregular margins and mamillations on surface with loss of hair was seen over the left parietal region on the scalp (figure 1). General, ocular, musculoskeletal and CNS examination was within normal limits. A 3 mm punch biopsy was taken from the affected site and it showed epidermal hyperkeratosis, acanthosis and papillomatosis, with numerous admixed mature and immature sebaceous glands with rudimentary hair follicles that was consistent *with "nevus sebaceous of Jadassohn"*.

Since, it is a benign, asymptomatic plaque with minimal malignant potential. Patient's parents were counselled regarding the nature of the disease and its progression during puberty and the various treatment modalities available and their potential adverse effects. They opted not to go for destructive procedures and hence patient was reassured and asked to come for follow up in case of new lesions or rapid change in texture, size, nodulation or ulceration.

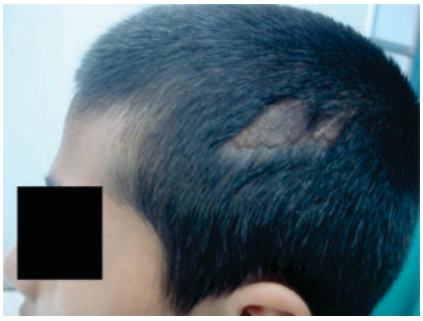

Figure 1 depicts a single well defined, orange-brown 4 X 2 cm elliptical plaque with irregular margins and mamillations on surface with loss of hair over the left parietal region on the scalp.

DISCUSSION:

Nevus sebaceous is a hamartomatous disorder of the skin and is an epidermal appendageal nevus disorder .Nevus sebaceous is a very rare condition approximately affecting 0.3% of all neonates. Both genders have a more or less equal predisposition.

The cases are usually sporadic and rarely familial[1]. Recently some studies have demonstrated that nevus sebaceous is caused due to post zygotic mosaic mutations in the HFAS and KHAS gene. It is being hypothesized that nevus sebaceous develops from pluripotent primary epithelial germ cells which have potential to differentiate into various malignancies. Certain deletions in the PTCH tumour suppressor genes on chromosome gp22.3 may lead to the neoplastic potential of the nevus. Certain studies have noticed increased expression of analogous receptors in the nevus sebaceus. The increase in the size of nevus in teenage or adolescence is a result of production of androgens which increases at the time of puberty and its effect on the pilosebaceous and apocrine elements .Broadly natural history of nevus sebaceous can be divided into the following two stages as shown below in Table 1:

Table 1 depicts the differences between the prepubertal and postpubertal stages of nevus sebaceous.

PREPUBERTAL	POSTPUBERTAL
1. Usually present since birth, sometimes may be present little later. 2. Clinically it presents as a solitary asymptomatic. Well defined waxy, yellow to orange, verrucous or mamillated (cobble stone texture), glabrous(complete or partial) plaque over the head and neck, sometimes the face(honeycombed pattern) 2 it can present in oval, linear or blaschkoid pattern. 3. The lesions are usually stable and show no increase in size from birth until puberty.	1. Presents after attaining puberty. 2. Clinically there is expansion and increase in the size, verrucosity and mamillated due to hormonal influence on the pilosebaceuos unit at puberty. 3. Nevus sebaceus might appear for the first time at this phase.

Usually, the lesions in nevus sebaceous are solitary but sometimes multiple or widespread lesions have been reported. The lesion are usually devoid of hair .Nevus sebaceous is associated with certain syndromes like Epidermal nevus syndrome which denotes the presence of epidermal nevi with developmental abnormalities in other extra cutaneous systems [3]. The epidermis, its appendages, CNS, part of eye and bones are all derived from the ectoderm. Hence in epidermal nevus syndrome, the extracutaneous involvements pertaining to the above mentioned structures can occur as shown in the figure 2:

Figure 2: Shows the extracutaneous involvement in nevus sebaceous syndrome. Not necessary that all features need to be present. * Malignant tumors are very rarely associated with nevus sebaceous (<5%).

Schimmelpenning-Feuerstein-Mims (SFM) syndrome or otherwise called the linear nevus syndrome in which large lesions in blaschkoid distribution are observed with extracutaneous manifestations mostly commonly involving the neural , ocular and skeletal systems [4] and Phakomatosis pigmentokeratotica (PPK) which is a rare epidermal nevus syndrome with nevus sebaceous and speckled lentiginous nevus [5].Nevus sebaceous sometimes leads to complications such as developmental defects which can take place if the lesions are large ,mental retardation, seizures, hemiparesis when there is neurological involvement in centrofacial lesions and benign and malignant changes which can occur in the original nevus. Neoplasms are most common in the fourth decade of life in 10 to 30% of individuals. Most of these lesions are benign with chances of malignancy being less than 5%. Usually in malignancy there is appearance of large, discrete, ulcerating nodule within the lesion. The commonest forms of benign appendageal tumors are syringocystadenoma papilliferum and trichilemmoma. The commonest form of malignant tumor is basal cell carcinoma. Also squamous, sebaceous, apocrine, eccrine carcinomas can occur .The diagnosis is predominantly based on clinical presentation of the lesion. A tissue biopsy or dermoscopy can be opted for confirmation of the diagnosis. Ultrasonography is used for prenatal diagnosis .Histopathology in prepubertal stage shows papillomatous epithelial hyperplasia. The epidermis may show slight acanthosis. The hair follicles are not properly developed and also sebaceous glands are underdeveloped. Apocrine glands are absent usually .In postpubertal stage, sebaceous glands become highly developed, numerous, hyperplastic. The hair follicles are small and primordial and hair roots are malformed (buds of basaloid cells) or disappear altogether. Epidermal hyperplasia, large sebaceous glands and ectopic apocrine glands are important findings .Eccrine gland hyperplasia can also be seen in some cases [6] .The neoplastic stage has characteristic benign and malignant tumors .The differential diagnosis includes wart, xanthogranuloma and mastocytoma .Management is mainly for cosmetic purpose and prophylactic excision can be done in small lesions in childhood owing to its potential for malignant transformation. Various treatment modalities includes excision, laser resurfacing, dermabrasion and photodynamic therapy [7].Patients should be educated regarding the associated benign and malignant tumors and if any change in texture color, size and occurrence of new neoplasms.

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