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Case Report

Dentistry Section

A Rare Entity of Giant Epidermal Exophytic Naevus of Hemifacial Region

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ABSTRACT

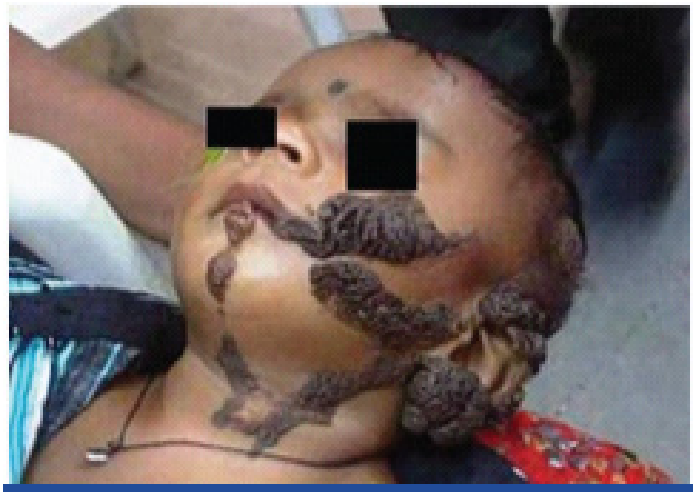
The term nevi constitute a hamartomatous growth of either skin or mucosa that is generally found at birth or shortly after birth. An epidermal nevus consisting chiefly of sebaceous glands is called as nevus sebaceous. Typically this entity is seen as a solitary bald patch over the scalp at birth. We report a case of exophytic lesion following blaschko's lines at birth. We find this case as peculiar because of the multiplicity of lesions, their pattern of distribution, as well as their atypical morphology.

Keywords: Ectodermal, Epithelial, Hamartoma, Sebaceous glands

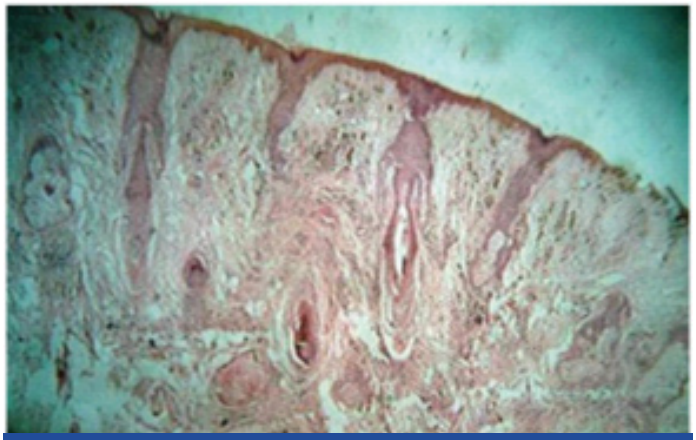
CASE REPORT

A three-month-old male baby reported to the Department of Oral and Maxillofacial Surgery, S.B Patil Dental College and Hospital, Bidar, Karnataka, with chief complaint of an ugly scar on the left side of the face since birth. History revealed no pre-natal complication and no history of consanguineous marriage.

Initial physical examination revealed large hyper pigmented multiple linear bands on the left side of the face extending from infra orbital region to corner of the mouth, tragus of the ear to cheek, involving pinna of the ear, ear lobe, sub mental region, lower border of the mandible and left temporal region (scalp). The lesions were elevated with velvety surface, black in colour [Table/Fig-1].



[Table/Fig-1]: A 3-month-old baby with nevi



[Table/Fig-2]: Photomicrograph showing thinning of epithelium and nevus cells (5X)

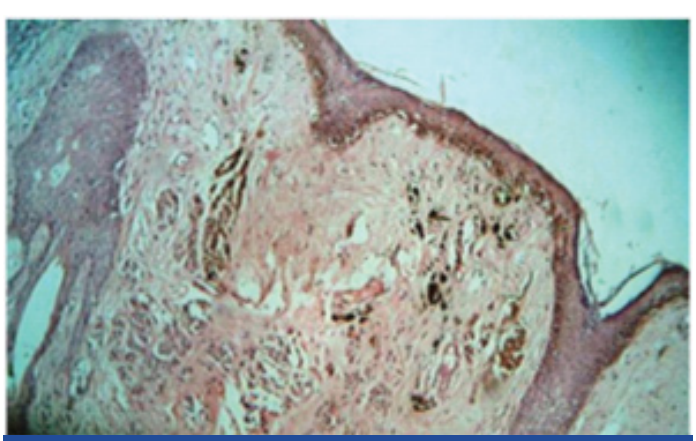
The baby showed normal growth parameters and appeared well nourished. General systemic evaluation by a pediatrician revealed no systemic involvement. The case was diagnosed as Linear Exophytic Epidermal Nevi and Linear Lichen Planus, Epidermodysplasia Verruciformis, Epidermolytic Hyperkeratosis (Bullous Congenital Ichthyosiform Erythroderma), Keratosis Follicularis (Darier Disease) and Nevus Comedonicus were included in differential diagnosis [1-3].

Investigations

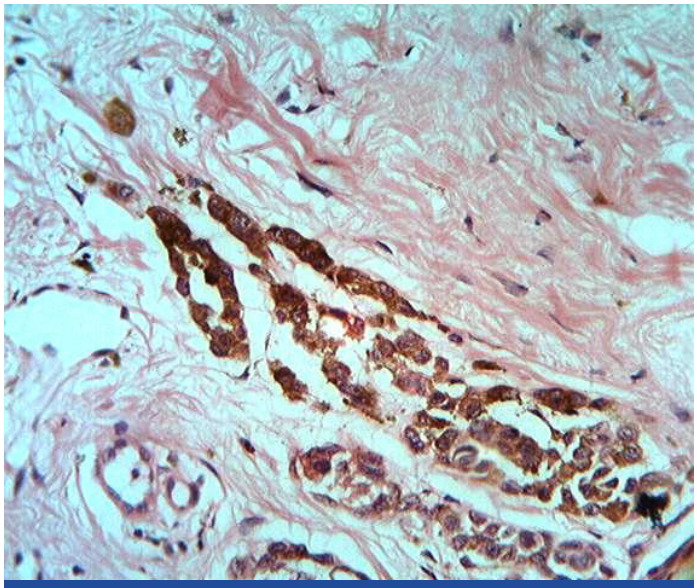
Biopsy of the lesion was done and histopathological examination was carried out. Epidermis showed focal thinning with mild hyperkeratosis. Upper dermis showed theques of nevus cells separated by hyalinized collagen bundles. Individual nevus cells are polygonal to ovoid with moderate amount of pale eosinophilic cytoplasm and central to eccentric semilunar nuclei with bland nuclear chromatin [Table/Fig-2-4]. The given histological picture was characteristic of nevus sebaceous.

Treatment

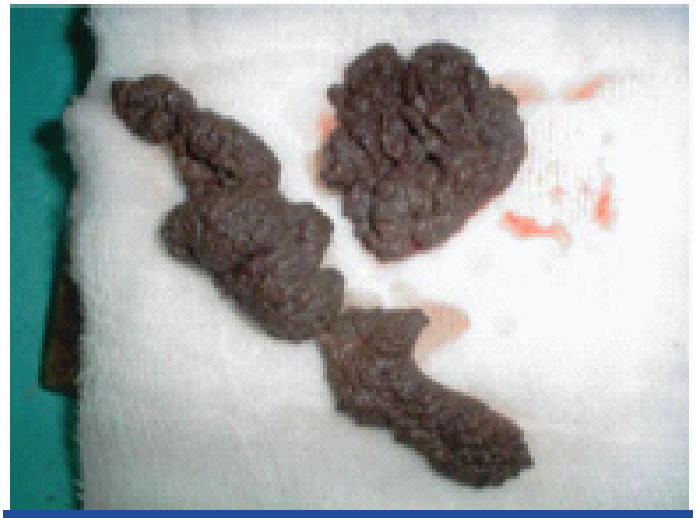
Different treatment options like surgical excision, laser treatment, topical vitamin D (calcipotriol), 5-fluorouracil, combination of Tacrolimus and fluocinonide, shave excision followed by phenol peeling were discussed with the parents and surgical excision was the choice. Surgery was done at 6 months of age, with excision of lesions on the neck, and chin till ear lobule. Excision and split thickness skin graft, with reconstruction of posterior of ear lobule with split thickness skin graft was done [Table/Fig-5].



[Table/Fig-3]: Photomicrograph showing theques of nevus cells (10X)



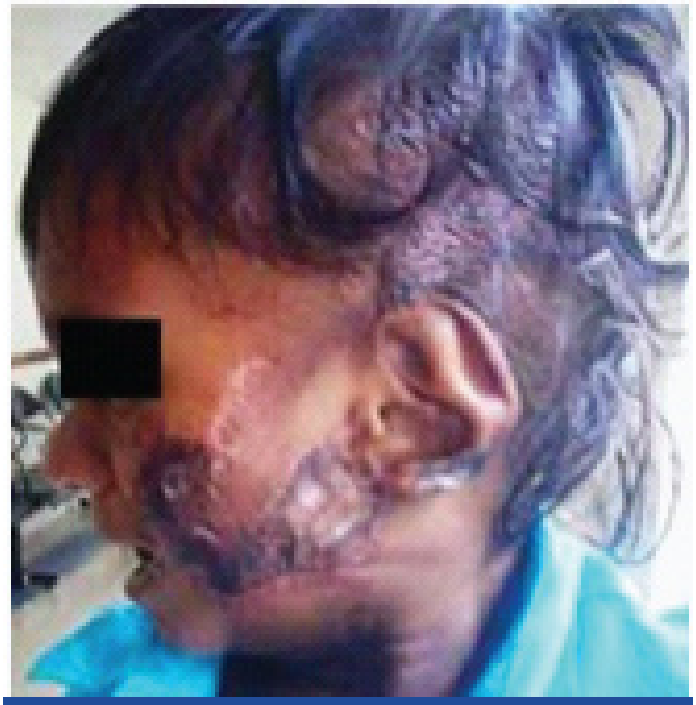
[Table/Fig-4]: Photomicrograph showing theques of nevus cells (40X)



[Table/Fig-5]: Gross specimen showing multiple bits

Outcome and Follow Up

One year follow up was done and no recurrence was seen postoperatively. Except for facial scars no other facial deformity was seen [Table/Fig-6].



[Table/Fig-6]: Postoperative picture after one year

DISCUSSION

Nevi or birth mark or mole are generally seen at birth and are defined as hamartomas, involving any part of skin or mucosa. The term giant is usually used if nevi involve larger part of the body. Nevi that originate from ectoderm and made of proliferated epithelium at birth and characteristically show linear configuration along the Blaschko's lines are termed as epidermal nevi [2,3].

Epidermal nevi are plaques, patches or nodules, generally arranged in a linear fashion along lines of Blaschko. On the basis of their principal component, these nevi are categorized sebaceous, eccrine, apocrine, keratinocytic or follicular nevi. Various studies showed incidence of epidermal nevi to be approximately 2 per 1000 live births and they show no gender predilection [4]. In 2/3^{rds} of cases these nevi manifest without involving other organs and in 1/3^{rds} of cases in syndromic form as epidermal nevi syndrome involving systemic organs like nerves, eyes, skeletal system and rarely renal and cardiac system [5].

Epidermal nevi syndrome has many variants with sebaceous nevi and verrucous being most frequent. Sebaceous variant is most commonly seen in head and neck region and verrucous variant is seen in other areas. Jadassohn in 1895 first used the term Nevi sebacei and described them as congenital cutaneous lesions arranged in a linear fashion. They are found in approximately 0.3% of all neonates [6] and presents as slightly raised, well circumscribed, yellow, orange, or tan plaques, having surface that is smooth or velvety. These lesions are usually asymptomatic. Most common sites of these lesions being scalp but may also be seen on the face, neck, and forehead. They show an abrupt midline demarcation. Even though they may run in families, majority of cases are sporadic in nature [7,8]. Conventional epidermal nevi present as a single bald patch over the scalp area at the time of birth and usually remain unchanged till puberty [7].

The clinical appearance of epidermal nevi depends on many factors such as the age of patient, site of the lesion, the predominant type of cell involved and its degree of differentiation. As in case of sebaceous nevi, sebaceous glands are predominantly involved and affected site is head and neck region. Epidermal Nevus Syndrome with underlying systemic abnormalities is also known by various terms such as nevus sebaceous syndrome, Jadassohn syndrome, Solomon syndrome and Schimmelpenning-Feuerstein-Mims syndrome, showing its heterogeneity [4,5]. Epidermal Nevus Syndrome show four major types: linear sebaceous nevus with predominant sebaceous glands, linear epidermal nevus, linear nevus comedonicus with predominant pilosebaceous units and inflammatory linear verrucous epidermal nevus in which keratinocytes are predominant [5]. This syndrome represents a continuum, as many other syndromes under its blanket are included by many authors. Since nearly 30 % of patients with epidermal nevi show organ abnormalities, their presence should alert the clinicians [9].

Linear epidermal nevus is a hamartoma and is often seen at birth as in our case, but may also occur in early childhood [9]. Reports have shown that there are no significant gender variations with male to female ration of 7:6, our case was seen in a baby boy. Generally nevi are asymptomatic, where as inflammatory linear verrucous epidermal nevus is symptomatic. Sebaceous nevus shows abnormal sebaceous, follicular, and apocrine glands. In early life, these lesions due to maternal hormonal expression appear to be well developed, but as the child grows they are under developed and reduced in number and size [7-9].

Clinically nevus sebaceous presents as a single, linear, hairless, slightly raised, yellow or brown plaque, with a smooth or velvety surface seen either at birth or in early childhood, the most common site being scalp. Other sites being face and neck regions. Our case was unusual as multiple lesions instead of solitary patch were seen [Table/Fig-1]. Later in adolescence, they appear as verrucous and nodular, linear, oval or round in shape; with varied size ranging from

few millimeters to centimeters in length. Studies have shown that tumors like syringocystadenoma papilliferum and trichoblastoma have been found to be arising with nevus sebaceous [10,11]. Rodins K and Baillie L have reported a rare case of hybrid follicular cyst with matrical differentiation consisting of basaloid matrical-type cells with luminal shadow cell keratinization [12]. The malignant transformation potential of sebaceous nevi has been found to be less than 1% [13].

Histopathologically epidermal nevus shows papillomatous hyperplasia of the epidermis. In the underlying dermis many mature sebaceous glands are found. Beneath these sebaceous glands few ectopic apocrine glands are often seen. Study of literature has revealed more than ten histological patterns, with more than one pattern in a single lesion [14,15]. The characteristic appearance in majority of the cases present with epidermal hyperplasia, papillomatosis, hyperkeratosis, variable parakeratosis and acanthosis.

Genetic studies have shown that the activation of an autosomal dominant lethal mutation that survives by mosaicism is the basis of occurrence of these nevi [16]. They suggested that time at which mutation occurs play a vital role, if it occurs early in embryonic life, they follow the Blaschko lines, whereas, if mutation occurs late, then they tend to be more fixed. In recent times to explain the patterns in nevoid skin disease, Gilmore proposed a hypothesis [17]. He suggested that these nevi are caused by effect of a few functionally abnormal clones. He categorized nevi in to two patterns, anatomical in which abnormal clone of cells are limited to the pathologic skin and functional, in which the two are spatially unrelated and hence may not follow Blaschko lines [12,13].

These patients are brought to doctors for aesthetic concerns. Various treatment plans have been proposed like topical treatment with a combination drug therapy of retinoic acid and 5-fluorouracil dithranol, topical steroids, chemical peels and podophyllin, which have shown to improve the keratotic lesional surface. But recurrence was seen in most of such treated cases. In these cases aggressive treatment was done with procedures like cryosurgery, electrocautery and dermabrasion and laser with limited success and recurrence. Complications like hypopigmentation were also seen to occur [15-18]. We advised regular follow-up of the patient, but they didn't turned up even after repeated reminder calls.

CONCLUSION

Epidermal nevi not only pose aesthetic problems but also should alert us of systemic complications as about 30% of such patients are found to have some or other underlying systemic defects.

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