NEVUS SEBACEOUS OF JADASSOHN

(A case report with review of literature)

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Summary

A clinical case report of a rare entity—Nevus Sebacecus of Jadassohn with review of literature is presented, specifying its unusual features—large size, bizzare configuration, and pinkish colour.

Clinically, it can easily be confused with Haemangioma (in pinkish lesions only) in addition to other differential diagnostic possibilities.

Results of treatment with full thickness surgical excision, even without the necessity of skin grafting is highly estimable.

The term nevus was previously used for the lesions containing nevus cells. Jadassohn's definition of organoid nevi includes stable localised malformation of the skin through excess or deficiency of one or more of the normal mature constituents, such as hair, glands, epidermis or connective tissue and it excludes adenomas and other less mature tumors. The so called nevus sebaceous also usually involves other skin constituents and sometimes most of them (Mehregan and Pinkus[15]).

The term organ-nevus (organoid nevus) dates back to 1895 when Jaddassohn[10] used it to distinguish localised and congenitally conditioned excess of glands and other constituents of the skin from pigmented (nevus cell) nevi. In the same paper, Jadassohn originated the designation Talgdrusen – nevus (sebaceous gland nevus) for a lesion, part of a systematised nevus of leg, which consisted entirely of large and well formed sebaceous lobules, but massed and out of proportion for the site in which they were found.

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Since its introduction into the American literature by Robinson[20] (1932), the term Nevus Sebaceous of Jadassohn has generally been used to connote a particular type of nevus which usually occurs on the scalp, face and neck and histopathologically characterised by excess of the sebaceous glands. It occurs as a solitary lesion, beginning at birth but sometimes developing in adult life (Montgommery[17]). It appears as a circumscribed, elevated, firm, yellowish plaque. Rarely the colour of the lesions may be pinkish (Conner & Bryan[3]). Usually, it measures several centimetres in diameter but larger ones are not rare and may involve the entire half of the face. Characteristically, it is asymptomatic (Biachine[1]). Other cutaneous findings include pigmented nevi and hirsuitism.

Mehregan and Pinkus[15] in 1955 published an extensive treatise based on their experience with 150 cases. The life history of this nevus (organoid nevi) includes three stages:—

(1) The first occurs in infancy and childhood and is characterised by an underdevelopment of hair and sebaceous glands ;

(2) During the second stage, initiated by puberty, massive development of sebaceous glands occur, with papillomatous epidermal hyperplasia and maturations of apocrine glands which are present in considerable number of cases and may be hyperplastic and cystic;

(3) The third stage results from the development of complications particularly benign or malignant or both the types of nevoid tumors developing within the original organoid nevus. The association with a variety of secondary nevoid tumors is an innate and not uncommon consequence in the life history of these lesions.

Amongst the tumors found and reported most prevalent ones are Basal Cell epithelioma - 15-20% as noted by several investigators. (Mehregan and Pinkus[15], Michalowski[16] Brannen & Fosuangh[2]). Syringocystadenoma papilloferum (Robinson[20], Parkin[18] and Mehregan & Pinkus[15]), sebaceous epithelioma (Mehregan & Pinkus[15]), and Michalowski[16]), Squamous Cell epithelioma (Parkin[18],) hydradenomas of various types (Mehregan & Pinkus[15], Pinkus[19]), Keratoacanthoma (Corbette[4]), and salivary gland adenocarcinoma.

Differential diagnosis should be made from Xanthoma, Syringo cystadenoma Papilliferum, Nevus Verrucosus (localised type), verruca vulgaris and other papillomatous and warty tumours.

Recently there have been reports of sebaceous nevi associated with multiple developmental abnormalities (Feuerstein and Mims[5] Marden and venters[14]). These reports have delineated a new Neurocutaneous syndrome, first described by Feuerstein & Mims[5] and further supported by Lantis et al[11], Bianchine[1] and Herbst et al[8]. The syndrome is constituted by the triad-linear sebaceous nevus, epilepsy and mental retardation. As quoted by Bianchine[1], the manifestations other than the triad have also been reported. These are skull abnormalities—like hydrocephalus, asymmetry, widening of the sella tursica and premature closure of the spheno-frontal suture; eye abnormalities including nystagmus, Lipo dermoids, scarring, and vascularisation of the cornea, choroidal coloboma and blindness and cardiovascular defects mainly coarctation of aorta. The bridge of the nose may be broadened. Orally, hypoplastic teeth, thickened mucosae secondary to nevi which may be present on the hard or soft palate. Genetically, thus far only sporadic cases have been reported with both males and females affected.

Like Neurofibromatosis, (Von-Recklinghausen), tuberous sclerosus (Bournville), angiomatosis retinae et cerebellae (Von-Hipple-Lindan) and encephalotrigemminal angiomatosis (Sturge-Weber), the nevus sebaceous of Jadassohn should be categorised as one of the phakomatosis (Gellis [7] et al).

Histopathologically, it is not a true adenoma but rather represents hyperplasia or the sebaceous glands. The glands appear to be mature. There is varying degree of hyperkeratosis and papillomatosis, (Montgommery[17]). In addition, the apocrine glands develop in a high percentage of cases in the lower part of the cutis, and they may appear hyperplastic and cystic (Lever[13]; Mehregan & Pinkus[15]).

Due to (1) wanting of the case reports available in Indian literature, (2) large area of involvement, (3) bizarre configuration of the lesions, (4) pinkish colour of the lesion which is unusually described and (5) successful treatment by full thickness excision, we seized an opportunity of reporting this uncommon entity. This communication described an adult male, with a congenital facial nevus of the type described by Jadassohn[10] in 1895.

Case Report

A male patient, aged 20 years, presented with a raised, pinkish lesion of the right cheek since birth. The lesion has been gradually increasing in size to acquire the present size of 6.1 cm x 3 cm x 1 cm. There is no history of similar lesion in his parents, siblings or other remote members of the family. No history of epileptic fits. The patient is quite intelligent. No history of any sort of trauma at the site.

These is no abnormality on systemic examinations. Clinically, there is no evidence of any opthalmological or neurological abnormality. Local examinations revealed a circumscribed, pinkish, papillomatous plaque with a velvet like surface and on the surface were present patulous hair follicular openings. The plaque was composed of closely packed rounded elevations. Size of the lesion was 6.1 cm x 3 cm x 1 cm. The lesion was involving, the right cheek, extending medially towards the right angle of the mouth and upper lip and right side of the nose; and it was irregular in shape (fig 1).

Fig. 1

Showing a cirumscribed pinkish Papillomatous plaque; involving right cheek extending medially towards right angle of mouth upper lip and right side of the nose

Clinical diagnosis suspected by us as well as surgeons was haemangioma, or some epithelial nevus, though histopathologically, it turned out to be Nevus Sebaceous of Jadassohn.

Laboratory Investigations

Haemoglobin	13.2 gms %
Total Leucocyte count	10600/cm.
Differential Leucocyte Count	Polymorphs - 76% Lymphocytes - 20% Monocytes - 2% Eosinophils - 2%
Erythrocyte Sedimentation Rate	- 8 mm/1st hour Westergren
Bleeding time	1' - 18"
Clotting time	3' - 52"
X-Ray Chest	N. A. D.
X-Ray Skull	No evidence of intracranial calcification.
Electrocardiogram	N. A. D.
Electroencephalogram	Could not be done
Opthalmological examination including fundoscopy	N. A. D.
Skin Biopsy	Showed extensive hyperplasia of sebaceous glands and lobules, the glands appear to be mature (fig. 2). Varying degree of hyperkeratosis and papillomatosis. Presence of mature and cystic apocrine glands in the lower part of the cutis (fig. 3).

Discussions

Nevertheless, ample case reports are available in world literature [1, 4, 7, 8, 11, 12, 15, 20], but hardly there is any case report traceable in Indian literature, which has prompted us to publish this case report.

Examples of extensive and multiple lesions are sparse in the literature (Lentz [12] et al,). The case reported in this series too, had huge size of 6.1cm., though it was a solitary lesion as

Fig. 2

Shows hyperplasia of the sebaceous glands and lobules. The glands appear to be mature.

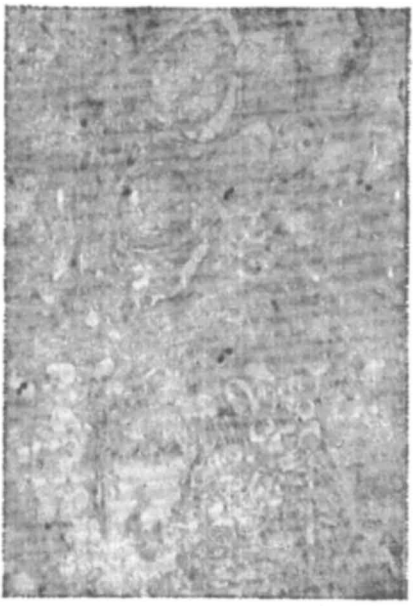

Fig. 3

Shows mature apocrine glandes in the lower part of cutis and hyperplastic sebaceous glands in the upper cuts

compared to Lentz [12]'s case who had multiplicity and enormously large size of the lesions as well.

Almost exclusively, the colour of lesions reported thus far has been yellowish. However, Conner & Bryan [3] did report pink colour of lesion in a few of their cases, which corresponds to the pinkish plaque presented by our patient.

Linear distribution has also been reported by many presenters (Gellis[7] et al; Lentz[12] et al and Feuerstein and Mims[5]), though the case in the present communication had a bizarre configuration which is not a usual presentation (Mehregan and Pinkus[15]).

The most commonly involved site i.e. scalp was uninvolved, rather face the second commonest site (Mehregan and Pinkus[15]) was the area of involvement in our case.

Other components of the neurocutaneous syndrome-epilepsy plus mental retardation[1,7,8,11], skull deformities, including hydrocephalus (quoted by Bianchine[1],) and eye changes (Lentz[12] et al) which has been reported by the workers quoted above, were absent in our case.

There was no associated benign epithelial tumor of appendage origin or sebaceous malignancy in our case as has been reported by other authors [3,4,15,18,19].

The patient in the current series underwent full thickness excision in several stages because of the histological evidence of apocrine glands present in the lower cutis as has also been suggested by Mehregan and Pinkus[15]. The cosmetic results were good even without necessity of skin graft. Moreover, superficial destruction of the lesion may actually provoke a papilliferum syringadenoma (Pinkus[19]), as minor acci dental trauma. Even more serious is high incidence of Basal cell epithelioma in early life. Some authors have considered these nevi as precancerous lesions (Michaloseki[16]).

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