

# NEVUS LIPOMATOSUS CUTANEUS SUPERFICIALIS

(Case Report with Review of Literature)

By

H. J. SHROFF\*, J. M. MOSES\*\*, R. K. GADGIL\*\*, & J. C. SHROFF\*

In 1921, Hoffmann and Zurhelle first described a clinical condition called 'Nevus Lipomatous Cutaneus Superficialis'. The second case was published by Robinson and Ellis (1937). In more recent years cases have been presented by Nikolowski (1950), Kutz (1951), Thone (1951), Holtz (1955), Blumenthal (1955), Hering (1956), Lunch and Goltz (1958), Abel and Dougherty (1962) and Stollmann et al (1969). To our knowledge, this is the first case report from India.

Nevus Lipmatosus is a rare naevoid anomaly of the skin and is, clinically and histologically, a distinct entity. Usually, at least a part of the nevus is present at birth, though there is extention of the lesion during childhood. In one patient, however, the nevus appeared at 18 years of age (Holiz, H., 1955). Sexual preponderance has not been observed. The nevus has a predilection for the region of the pelvic girdle. Common sites of occurrence are the gluteal region, lower back, sacral and coccygeal region, and various surfaces of the upper thighs. Kuta (1951) reported a case where the lesion was on the right side of the chest near the angel of the ribs.

Clinically, one finds an asymptomatic non-tender plaque measuring upto 8x15 cm and consisting of groups of papules or nodules, often arranged in linear or systematized distribution (Lunch & Goltz, 1958). These may even follow the lines of the body folds. The individual papule may be as small as a pinhead, elevated only a millimeter. Confluent nodules may reach 2x0.5 cm. above the surrounding skin surface. The colour is like that of the nearby normal skin, pale yellow or waxy. Usually, the surface is smooth but as time progresses the lesion may assume a papillomatous or verrucous tendency, or become cerebelliform (Nevus cerebelliformis) (Andrews, 1963). The consistency is invariably soft.

Histology of the nevus reveals mature fat cells present in the middle and deeper portions of the corium, often extending as high up as the sub-papillary layer (Lever, 1967). For this reason, it is also known as 'Nevus Lipomatous cutaneus sub-epidermalis (Robinson & Ellis, 1937). The aggregated fat cells are embedded among the collagen bundles of the dermis. Serial sections show that some of these accumulations have connections with the underlying sub-cutaneous fat while others have none. There is often grouping of these fat lobules around appendages and, particularly, blood vessels. Epidermal changes are secondary and resemble those of epidermal nevi.

*Histogenesis*: Formerly, it was assumed that fat cells in nevus lipomatous formed as the result of a fatty degeneration of collagen (Hoffman & Zurhelle, 1921; Nikolowski, 1950; Abel & Dougherty, 1962). However, it is more likely that nevus

---

\* Department of Dermatology, J. J. Group of Hospitals

\*\* Department of Pathology, Grant Medical College Byculla, Bombay 8.

lipomatosus represents a naevoid anomaly in which ectopic fat cells form in the dermis (Robinson & Ellis, 1937; Holtz, H., 1955; Lynch & Goltz, 1958). Holtz, H., (1955), demonstrated that in addition to large mature fat cells, less mature forms comparable to those found in embryonic panniculus adiposus also occurs. These apparently arise from the walls of the venous capillaries around which the abnormality centers and grow into the surrounding connective tissue. The connective tissue persists as very fine collagen, elastic and reticulin fibres reaching from the unaffected connective tissue to the wall of the central vessel. He concluded that the primary change is a growing out of primitive 'pre-adipose' tissue from the blood vessel walls. Edema and invasion of the surrounding connective tissue result in its destruction. The young adipose cells then progressively take on fat and at last come to be identical with mature cells of the sub-cutaneous layer. This process corresponds to that described by Wasserman (1926) in the embryonic development of normal fatty tissues and in lipomas.

*Differential diagnosis:* Focal dermal hypoplasia has been reported as nevus lipomatodes superficialis (Medzinski, 1963). This condition is characterised by irregular linear streaks of telangiectasia, atrophy and pigmentation on the trunk and limbs with widely distributed soft nodules. Nail anomaly, ocular defects, skeletal malformations and facial asymetry are sometimes associated. Microscopically, although fat cells are more pronounced as compared to nevus lipomatosus, their clustering around the appendages and blood vessels is not commonly found. Of late, striated nevus lipomatosus has been reported in association with Incontinentia pigmenti (Stollmann, 1969).

*Treatment:* Like other cutaneous naevi, lipomatosus is left alone except for cosmetic purposes when excision with a skin graft, if necessary, may be performed.

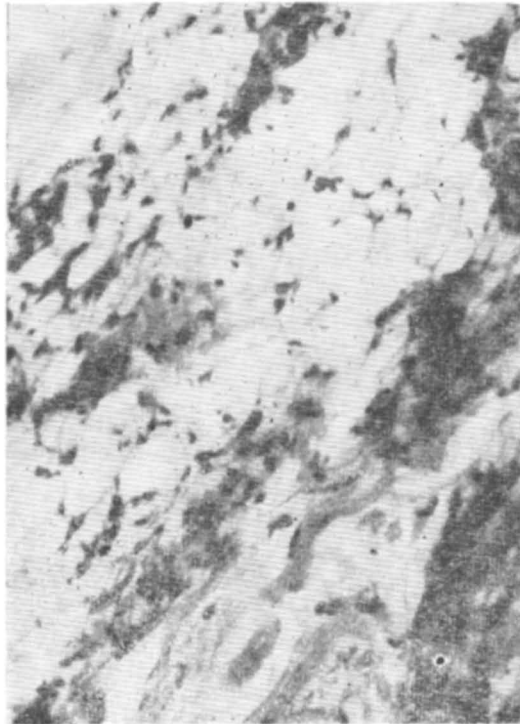


#### CASE REPORT

A school boy, 13 years of age, was brought to the skin department of J. J. Group of Hospitals seeking advice for his skin tumor (fig. 1.) The history revealed

that at the age of 4 years, he developed a small skin lesion on the lower back on the left hand side. Six months later a nodule formed in the adjacent area. The lesion gradually enlarged over a period of a years to the present size. It remained asymptomatic throughout the course. No treatment was received. The patient's general health was good. There was no evidence of any other naevoid disorder. Family history was non-contributory.

On local examination, a horizontal plaque measuring 15 cm x 5 cm, pale yellow in colour and consisting of papules and nodules was seen on the lower back mainly on the left side, but also crossing the mid-line over to the right side for 1/2". The consistency was soft. The tumour was non-tender and was situated entirely in the skin, being free from the deeper structures. There was no lymphadenopathy. Vertebral column was normal. Systemic examination did not reveal any significant finding.



Histology of the lesion showed groups and islands of fat cells in the middle and lower dermis (fig. 3) embedded among the collagen bundles (fig. 4.) and situated particularly around the blood vessels. The individual fat cells were all of the mature type. A mild and patchy non-specific inflammatory-infiltrate was also seen in the dermis. The epidermis showed a moderate acanthosis with irregular downward proliferation of the rete ridges.

*Summary:* A rare case of Nevus Lipomatosus Cutaneus Superficialis is presented. To our knowledge, not more than twelve cases have been reported in the world's literature, upto date, this being the first case reported from India. Clinical and histological findings are described. The literature is reviewed in detail.

*Acknowledgement:* We are thankful to Dr. G. K. Karandikar, Dean, Grant Medical College and J. J. Group of Hospitals, Bombay for allowing us to publish this case.



## REFERENCES

- Abel, R. & Dougherty, J. W. (1962) 'Nevus Lipomatosus Cutaneus Superficialis' Arch. Derm. 85:524-526.
- Andrews George C. (1963) 'Diseases of the skin' published by W. B. Saunders Co., Philadelphia & London, p. 579.
- Blumenthal, F. (1955) 'Nevus Lipomatodes', A. M. A., Arch. Derm. 72:279.
- Goltz R. W. et al (1962) 'Focal Dermal Hypoplasia' Arch. Derm. 86:708.
- Hering, H. (1956) 'Kasuistischer Beitrag zum Nevus Lipomatodes Cutaneus Superficialis' Ztschr Haut u Geschlkr. 21:123-125.
- Hoffmann, E. & Zurhelle, E. (1921) 'Über einen Nevus Lipomatosus Cutaneus Superficialis' der lincker Glutaalgegend, Arch. Dermat u Syph. 138:327-333.
- Holtz, H. (1955) 'Zur Histologie des Nevus Lipomatodes Cutaneus Superficialis (Hoffmann-Zurhelle) als Beitrag zur Entwicklung des Fettgewebes, Arch. Dermat. u Syph. 200:526.
- Holtz, K. H. (1955) 'Beitrag zur Histologie Nevus Lipomatodes Cutaneus Superficialis' Dermatologica. 103:42-47.
- Kuta A. (1951) 'Nevus Lipomatodes Cutaneus Superficialis s Subepidermalis' Dermatologica. 103:42-47.
- Lever, Walter F. (1967) 'Histopathology of the skin' Fourth Ed. published by Pitman Medical Co. & J. B. Lippincott Co. P. 669.
- Lynch, W. F. & Goltz P. W. (1958) 'Nevus Lipomatosus Cutaneus Superficialis' A. M. A. Arch. Dermat 78:479.
- Miedzinski, F. (1963) 'Nevus Lipomatodes Cutaneus superficialis (Hoffmann Zurhelle), Dermat Wchnschr 122:735-742.
- Nikolowski, W. (1950) 'Nevus Lipomatodes Cutaneus superficialis (Hoffmann Zurhelle), Dermat Wchnschr 122:735-742.
- Robinson H. M. & Ellis F. A. (1937) 'Nevus Lipomatosus Subepidermalis seu Superficialis Cutis' Arch. Dermat & Syph 35:485-488.
- Stellmann, K. et al (1969) 'Striated Nevus Lipomatosus Cutaneus Superficialis' in Incontinentia Pigmenti' Geschlechtskr 44:555-556.
- Thone, A. W. (1951) 'Ein Fall von Naevus Lipomatodes' Hautarzt 2:512-513.
- Wassermann, F. (1926) 'Ztschr. Zellforsch. 3:235-328.