LIPOGRANULOMATOSIS SUBCUTANEA

(A report of two cases)

By

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(Lipogranulomatosis subcutanea connotes a specific type of panniculitis and has been described by Rothmann (1894) and Makai (1928). The disease entity is marked by spherical subcutaneous nodules of varying sizes. The distribution of these nodules is on extremities and the nodules are usually elastic or firm. The nodules are tender on deep pressure.

Characteristically, the nodules appear individually and not in crops as in the case of Weber-Christian disease. There is a remarkable paucity of systemic disturbances in this disease entity and the patient invariably presents with only multiple subcutaneous swellings on extremities and trunk. Lipogranulomatosis is known to occur at all ages, more often amongst younger age groups and is equally common in both sexes.

Pathological changes occurring in the fat are usually secondary to changes in the blood vessel wall supplying the fat; the blood vessels are thickened and edematous. The progression of these changes leads to focal degeneration of fat cells and subsequent necrosis. The changes following necrosis are usually progressive and in the initial stage consist of infiltration of polymorphonuclear leukocytes and histiocytes. These histiocytes imbibe the necrotic fat cells and there is formation of granulomatous tissue with giant cells and eosinophils. Fibrosis subsequently replaces these granulomatous tissue changes.

CASE REPORT No. 1

A young man, 25 years of age and a worker in a foundry reported to the hospital with a history of several small swellings on both upper extremities as well as the thighs. The swelling were of 3 months duration. He gave history of these swellings appearing individually and at no time he had any systemic disturbances, such as fever, pain or malaise. He gave no history suggestive of trauma and enquiries regarding similar swellings in the other members of his family were negative. Examination showed a healthy young man having a number of spherical firm nodules on the medial aspect of his right and left arm and both thighs. The nodules were not tender, the overlying skin was normal and not adherent to the swelling underneath. Only marked pressure elicited tenderness. (The patient was not very anxious to have any treatment given to him at all. However, he was persuaded to have a few investigations) An excision biopsy of the swelling in his right arm was done.

Investigations carried out were as follows:

HB: 12 gm per cent
WBC: 9000/ cmm

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Urine examination–N.A.D.
Examination for LE cells–Negative

Macroscopic examination:

A small bit of tissue; consistency firm.

Microscopic examination: (S–64/1484)

The subcutaneous fat showed necrosis and almost complete replacement by an inflammatory exudate composed of a few polymorphs, histiocytes and lymphocytes. An occasional eosinophil is also present. The exudate is quite dense. Blood Vessels do not show any particular lesion except swollen and proliferating endothelial cells in some capillaries. At places collection of histiocytes (epithelioid cells) are arranged in follicular fashion with giant cells of foreign body type. Few strands of fibrous tissue are also seen traversing the lesion (confirmed by Gomori – aldehyde fuchsin – Masson counterstain). Cystic spaces due to liquefied fat are also seen (Fig. 1 and 2).

The patient has been followed for a period of about 8 months and most of the swellings have become very small. Some of them have even disappeared.

CASE REPORT No. 2

Patient's name, J. L. aged 24 years old, sex male admitted on 6–7–65. On admission he gave history of a nodule in the hypogastric region probably of three months duration. A previous biopsy (S–65/1034) taken on 16–6–65 was available.

Local Examination:

A nodule left forearm situated subcutaneously; with increase in size he gets pain otherwise no particular complaint. During the last 15 days he has been getting urticaria. No lymphadenopathy.

In the extensor compartment of the fore–arm, a small oval hard nodule about 2.5 x 1.5 cm.; moves sideways and is deeper to the muscle, not tender; no other swelling detected. Clinical diagnosis: ? Neurofibroma; ? Fibroma.

7–7–1965.

No nodule detected (Minor surgery excision).

9–7–1965:

Urine: N. A. D.
X–Ray chest: N. A. D.
R. B. C.: 5.4/ cmm.
H. B.: 15.9/ gm.
W. B. C.: 7400/ cmm.
P. 50% L. 45%, E. 5%

Readmitted on 20–7–1965. Nodule left forearm; excised for biopsy, on 24–7–1965.

Macroscopic Examination: (Biopsy dated 16–6–1965)

A flat bit of irregularly oval tissue about 1 x $\frac{1}{2}$ x $\frac{1}{4}$ cms. Cut section: Appearance that of a whitish fibrotic tissue.

Microscopic examination: (S–65/1034)

The section (H & E. Stain) shows areas of fat necrosis where fibrin and necrotic debris can be seen. Such areas are surrounded by inflammatory exudate amongst which a few polymorphonuclears are also seen (Fig. 3). At the periphery of the nodule, tubercle like follicles with epithelioid cells and lymphocytes surrounded by young collagenous tissue are also seen (Fig. 4), There is no evidence of arteriolitis but at places perivascular cuffing is seen. Small spherical or oval blue structures surrounded by eosinophilic material are seen in the necrotic debris and its vicinity. These structures in periodic acid Schiff's– Weigert counterstain appear black surrounded by intensely, PAS positive material. One of the capillaries shows evidence of thrombosis and endothelial cells swelling and proliferation. Gomori aldehyde fuchsin–Masson counterstain shows varying amount of collagen separating the necrotic and the tubercle like lesions. Sections stained with Ziehl–Neelsen stain did not reveal acid fast organisms.

Macroscopic examination: (biopsy dated 24–7–65)

A small oval nodule about 3x2 mm. in size.

Microscopic examination: (S–65/1280 A & B)

The lesion consists of well formed fibrous tissue in which a few blood vessels are seen. There is perivascular cuffing with lymphocytes, histiocytes and an occasional polymorph. At places the cellular exudate is quite dense. There are also two small islands or adipose tissue surrounded by fibrous strands with chronic inflammatory cells. S–65/1280 B shows an area of fat necrosis surrounded by lymphocytes, histiocytes and an occasional polymorph. The lesion appears to suggest a late stage in the evolution of the disease process–healing with fibrosis and chronic inflammatory reaction.

24–7–65–A Nodule has appeared yesterday. Over anterior abdominal wall, right lumbar region. Electrophoretic pattern–Normal.

26–7–65–Patient discharged.

DISCUSSION

Baumgartner and Riva (1945) suggested the following classification of panniculitis:

1. Secondary to local inflammations and tumours.
2. Caused by external factors.

A. Subcutaneous injection of oil, paraffin, or insulin.

B. Mechanical trauma (adiponecrosis subcutanea Neonatorum)

3. " Spontaneous " circumscribed panniculitis.

A. Weber–Christian disease or syndrome.

B. " Lipogranulomatosis subcutanea (Rothmann–Makai), Kooij (1950)" suggested two more forms:

B1 Caused by medications (bromides and iodides)

B2 Due to systemic infections (tuberculosis, syphilis, sporotrichosis, Leprosy etc.)

In view of this patient's occupation in the foundry, initially, a diagnosis of fat necrosis following trauma was entertained. However, considering the multiplicity of these nodules and the unusual sites trauma was not considered a factor in the causation

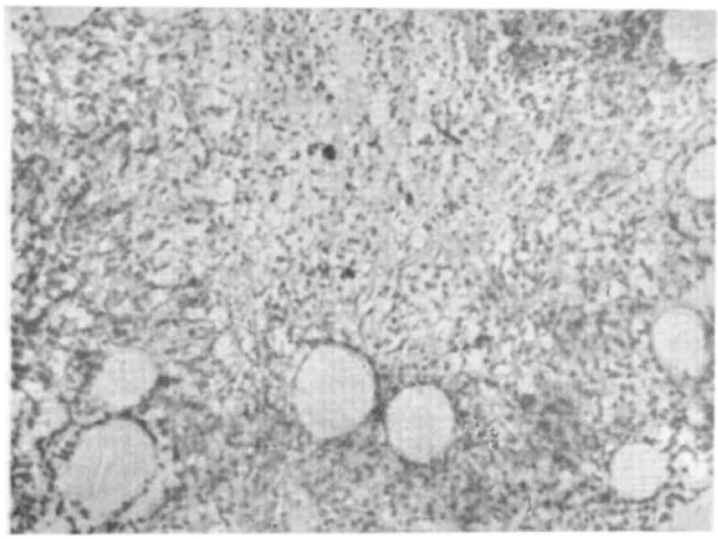

Fig. 1 : Photomicrograph showing an area of fat necrosis and inflammatory cells; small fat cysts are also seen (H. & E. 420)

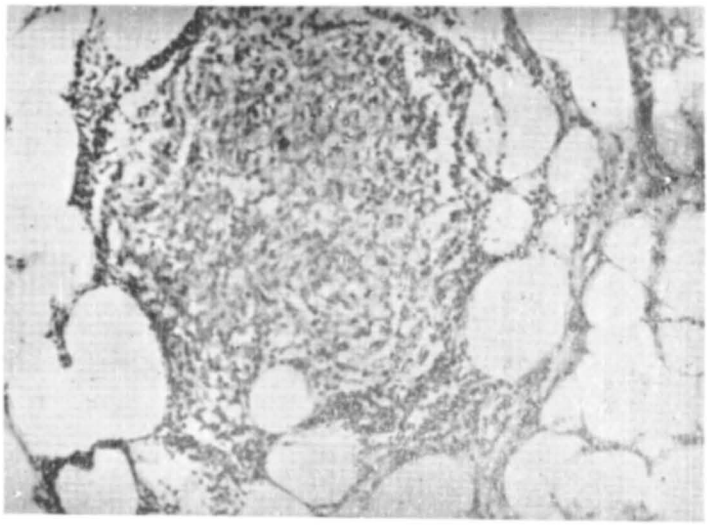

Fig. 2 : Photomicrograph showing a collection of epithelioid cells and an occasional giant cell. (H. & E. X 420).

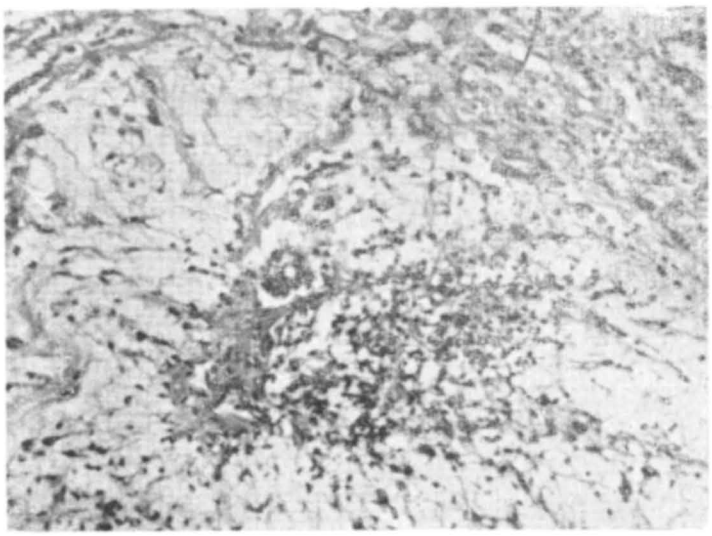

Fig. 3 : Photomicrograph showing an area of necrosis and inflammatory exudate consisting of fibrin and a few polymorphonuclear cells (H. & E X 420).

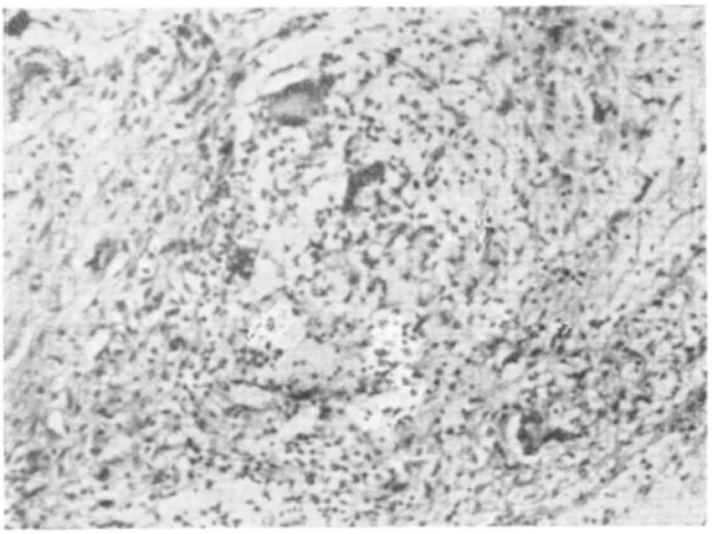

Fig. 4 : Photomicrograph showing epithelioid cells, giant cells and a few inflammatory cells (H. & E. X 420).

of this lesion. There was no evidence of any irritating factor either physical or chemical and in view of total lack of systemic disturbances as well as spontaneous regression of such nodules, the diagnosis of Weber–Christian disease was considered untenable.

These cases are unquestionably those of lipogranulomatosis. The appearance of these nodules singly without associated fever rules out the possibility of Weber–Christian disease. The absence of acid fast bacilli may be considered a point against the diagnosis of Erythema induratum in which similar histology is seen. Further caseation is conspicuous by its absence. The possibility of Erythema nodosum is excluded because in this condition necrosis and abscess formation does not occur and usually there is associated phlebitis. In cases presented here necrosis is present and there is no evidence of phlebitis. Both these patients on follow up did not show any ulceration over such lesions.

Could these be cases of Rothmann–Makai type of lipogranulomatosis? The appearance of solitary nodules at intervals, the histology and the history of diminution in size are points in favour of such a diagnosis. The histology of the biopsy of the first case and that of the first biopsy of the second case is compatible with the first stage of Rothmann–Makai syndrome. The availability of two biopsies in case No. 2 at an interval of 5 weeks shows definite evidence of resolution of the lesion with fibrosis; and healing by fibrosis is known to occur in the third stage of this syndrome.

Treatment:– Corticosteroids, Antibiotics and Sulphanamides have been tried but have been found to be ineffective. There is no satisfactory medical treatment for this entity and excision of persistantly painful nodules is the only effective answer to this problem.

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