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Necrobiosis Lipoidica in Non-Diabetic Individual - A Case Report.

Prakashiny*, and Hemalatha Ganapathy.

Department Of Pathology, Sree Balaji Medical College Hospital, Chennai, Tamil Nadu, India.

ABSTRACT

Necrobiosis lipoidica is a non-infectious granulomatous disease of unknown etiology and pathogenesis. It is more commonly seen in young adults and early middle aged individual. It is often associated with diabetics and typified by indurated plaques of the shin. Herein, we report a case of necrobiosis lipoidica in a 62 year old non-diabetic female who presented with the complaints of pain and swelling over right knee joint for 3 weeks duration. On examination there was diffuse edema of right lower limb and pigmentation with papule in the right knee joint. Skin biopsy taken from the papule and sent for histopathological examination. Histopathology showed a granulomatous dermatitis, pointing towards necrobiosis lipoidica. Special stain showed Acid Fast Bacilli negative. Histopathology plays a major role in the diagnosis of this rare disorder, besides clinical history.

Keywords: necrobiosis lipoidica, granuloma, acid fast bacilli, histopathology.

**Corresponding author*

INTRODUCTION

Necrobiosis lipoidica (NL), is a chronic spreading inflammatory skin disorder of the shin and has a special tendency to occur in diabetics [3].

In 1966, Muller and Winkelman reported that two thirds of patients with necrobiosis lipoidica had overt diabetes at the time of diagnosis. Of the rest, only 10% developed diabetes within 5 years. Necrobiosis lipoidica commonly develops in young adults and in early middle age [1].

Necrobiosis lipoidica may resemble granuloma annulare clinically [3] giving a diagnostic dilemma.

Here we report a case of necrobiosis lipoidica in a 62 year old, non-diabetic individual.

Case Report

A 62 year old female presented to the dermatology out-patient department, Sree Balaji medical college hospital, Chennai, with the complaints of swelling with pain over right lower limb for 3 weeks duration. There was no history of Diabetes mellitus. Her blood glucose were within normal limits. Her Glucose Tolerance Test (GTT) also normal.

On examination, there was a diffuse edema of right lower limb, with pigmentation and papules over the knee joint.

Punch biopsy taken from the papule and sent in formalin to the pathology department for histopathological examination.

Grossly, it was a skin with a soft tissue measuring less than 0.5cm.

Microscopically, Haematoxylin and eosin sections showed skin overlying a fibrocollagenous dermis enclosing a discrete granulomata composed of multinucleated giant cells, epitheloid cells, histiocytes and lymphocytes [2]. Mononuclear collection composed of plasma cells and lymphocytes were seen in the superficial as well as deep dermis. [fig1]

Special staining with fite faraco showed Acid Fast bacilli (AFB) negative.

Impression was given as granulomatous dermatitis- necrobiosis lipoidica.

DISCUSSION

Necrobiosis lipoidica was first coined by Urbach, as necrobiosis lipoidica diabetorum in 1932.

Necrobiosis lipoidica is an idiopathic disorder typified by indurated plaques of the shin [3]. It is commonly associated with diabetes mellitus. It occurs at any age group, but common in adult age group with female preponderance. It is usually bilateral.

The common sites of necrobiosis lipoidica are lower extremities including ankles, calves, thighs, popliteal area and feet [3]

It is a granulomatous disease of unknown etiology and pathogenesis.

According to Muller and Winkelman, Necrobiosis lipoidica can be classified histopathologically into two groups:

- Pallisading granulomatous type &
- A tuberculoid or sarcoidal type, which is characterized by presence of many epitheloid cells and giant cells, frequently grouped in pseudotubercles [2,6]. This type is named as necrobiosis lipoidica granulomatosis and was believed to be identical to Miescher's granuloma described early in 1948. [5]

Microscopically, the epidermis may be normal, atrophic or hyperkeratotic. Dermis may show granulomatous component, histiocytes arranged in palisades, inflammation, degeneration of collagen and sclerosis. Occasionally, there may be epithelioid cells and giant cells. Giant cell may be of langhan's or foreign body type.

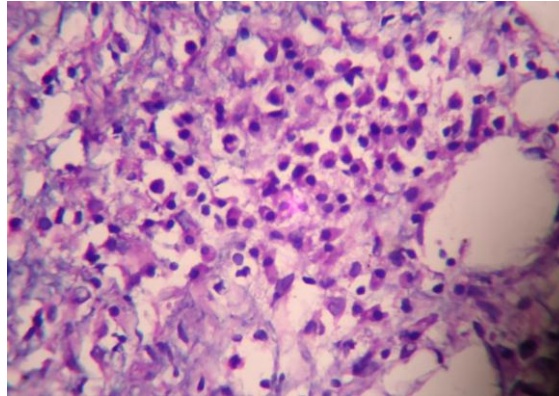


FIGURE 1: HIGH POWER VIEW-FAT CELLS SURROUNDED BY PLASMA CELLS AND LYMPHOCYTES.

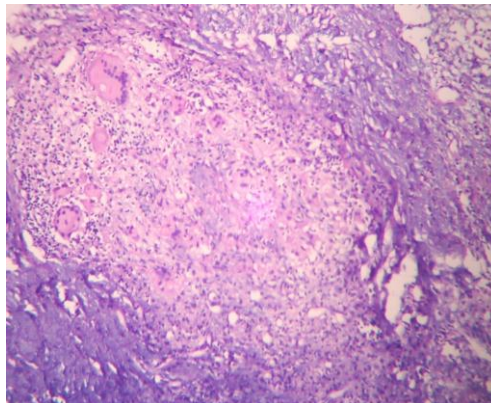


FIGURE 2: LOW POWER VIEW -DISCRETE GRANULOMA SHOWING LANGHANS GIANT CELL, EPITHELOID CELLS AND LYMPHOCYTES.

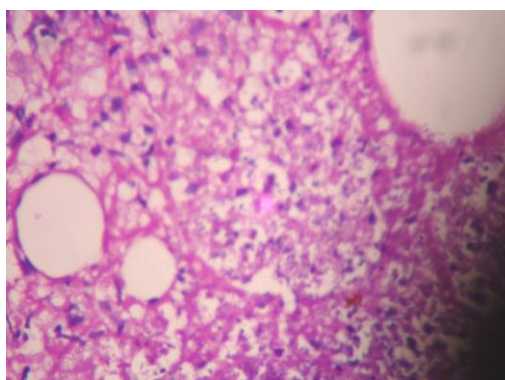


FIGURE 3A: HIGH POWER VIEW-FAT CELLS SURROUNDED BY AREA OF NECROSIS.

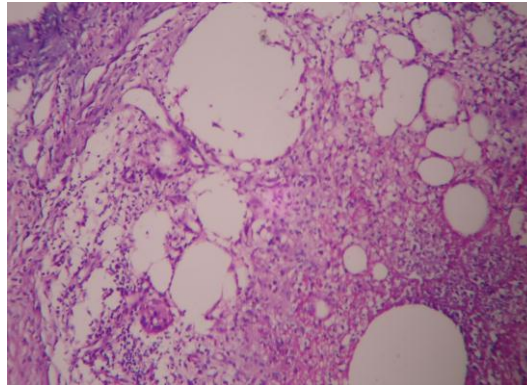


FIGURE 3B: HIGH POWER VIEW-FAT CELLS WITH NECROSIS

Lipid may be seen in the upper dermis. Plasma cells at dermal subcutaneous junction are typical of necrobiosis lipoidica as seen in our case.

Electron microscopy shows degenerative changes in collagen and elastin with loss of cross-stratifications in collagen fibrils [3].

Direct immunofluorescence shows deposits of immunoglobulin and C3 in the vessel wall. [10]

Differential diagnosis

- Granuloma annulare,
- Sarcoidosis,
- Necrobiosis xanthogranuloma.

In granuloma annulare, histiocyte palisading with prominent mucin is seen [8]. Lipid deposition is not seen. Sarcoidosis is a noncaseating granuloma with intracytoplasmic asteroid bodies. In necrobiosis xanthogranuloma, numerous cholesterol clefts may be seen [9].

CONCLUSION

Our case is a rare example of Necrobiosis lipoidica in a non-diabetic patient. Besides clinical, histopathology is essential for the diagnosis of this rare disorder and to rule out the other differential diagnoses.

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