Eosinophilic Granuloma

Pages with reference to book, From 98 To 100

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Eosinophilic granuloma is a variant of Langerhan's cell histiocytosis which may be unifocal or multifocal in distribution[1-5]. The disorder specifically and predominantl1 involves bones, particularly the clavarium, ribs and femurs. Rarely other organs like lungs, stomach and liver are involved[1,2,5]. still more rare is the involvement of the skin as a unifocal disease[6,7]. Histologically the disease is characterized by an erosive accumulation of Langerhan's histiocytes, variably admixed with eosinophils, lymphocytes, plasma cells and neutrophils. The eosinophilic component ranges from scattered mature cells to sheet like masses of cells[8]. The prognosis of the disease is variable. The multifocal variety is a serious form of disease whereas the unifocal variety is generally considered relatively indolent with sometime spontaneous healing or cured after local excision or irradiation[8].

Case Reports

Case 1

A 9 years old mak child presented with the history of a swelling below the right eye since three months which subsequently fungated and ulcerated (Figure 1).

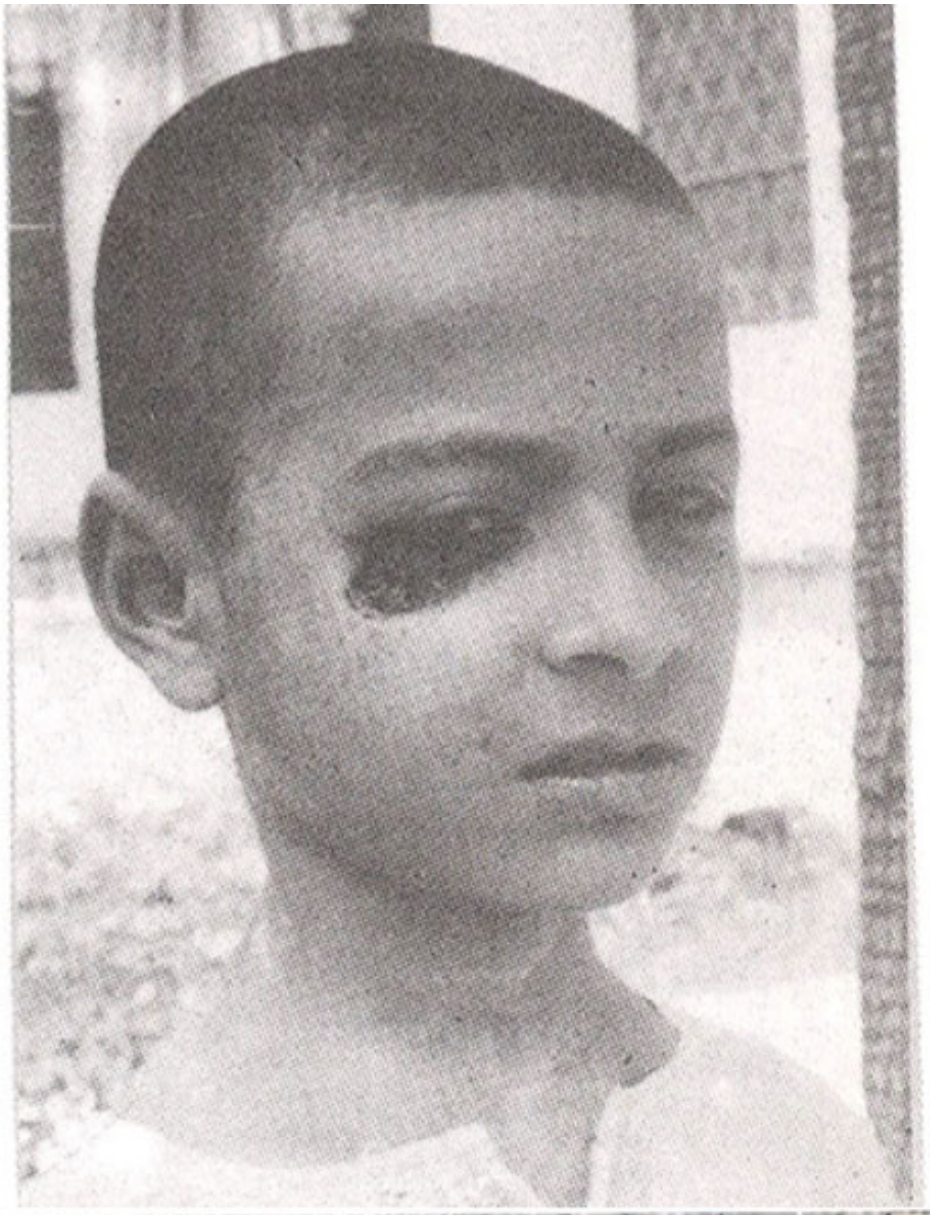

Figure 1. Photograph of the patient (case No. 1) showing a fungated, ulcerated lesion on the face below the right eye.

One month later, right cervical lymph node enlargement was found when he reported to the Paediatric Surgical OPD. His blood picture showed Hb 10,1 gm/dl, total leucocyte count 6500/cmm, (N.41%, L40%, M.06, EO. 13%), platelet count 250000/cmm,, ESR 10mm/first hour and the absolute eosinophilic count was 845/cmm. X-ray chest and facial bones did not reveal any significant changes.Urine analysis and stool examinations were normal, The ulcer and enlarged lymph node were biopsied and processed for histological examination, The morphology of the ulcer revealed well differentiated Langerhan's histiocytes admixed with numerous eosinophils alongwith dense acute and chronic inflammatory infiltrate and giant cells of the foreign body type (Figure 2).

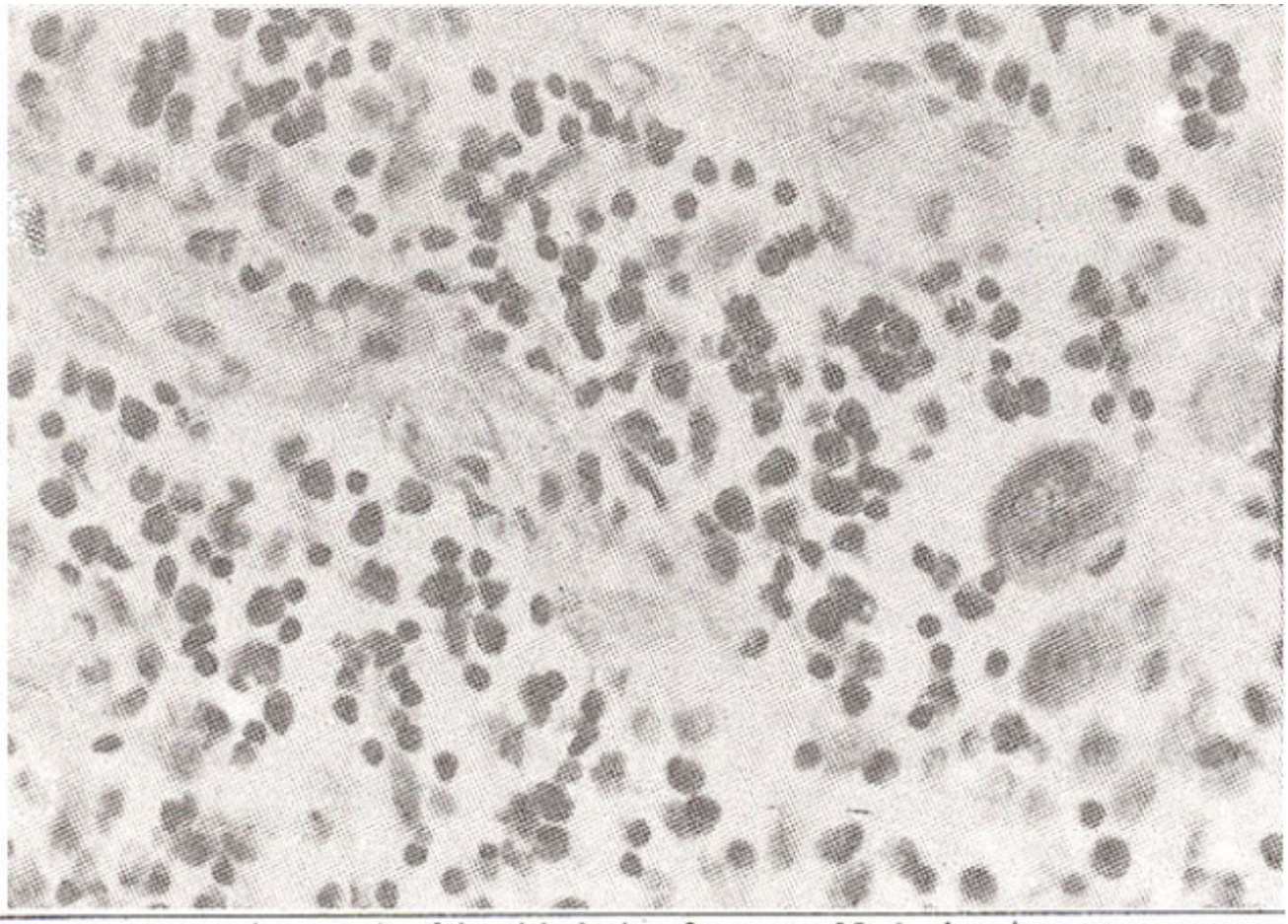

Figure 2. Photomicrograph of the skin lesion from case No.1, showing numerous eosinophils mixed with lymphocytes and a few giant cells. H&E x 380.

Areas of haemorrhages and necrosis were also present The lymph node showed loss of arehitecture and its replacementby sheets ofpolymorphous cellular infiltrate with an identical morphological appearance as that in the skin lesion. Therefore, the diagnosis of eosinophilic granuloma of skin was made with involvement of the regional lymph node (Figure 3).

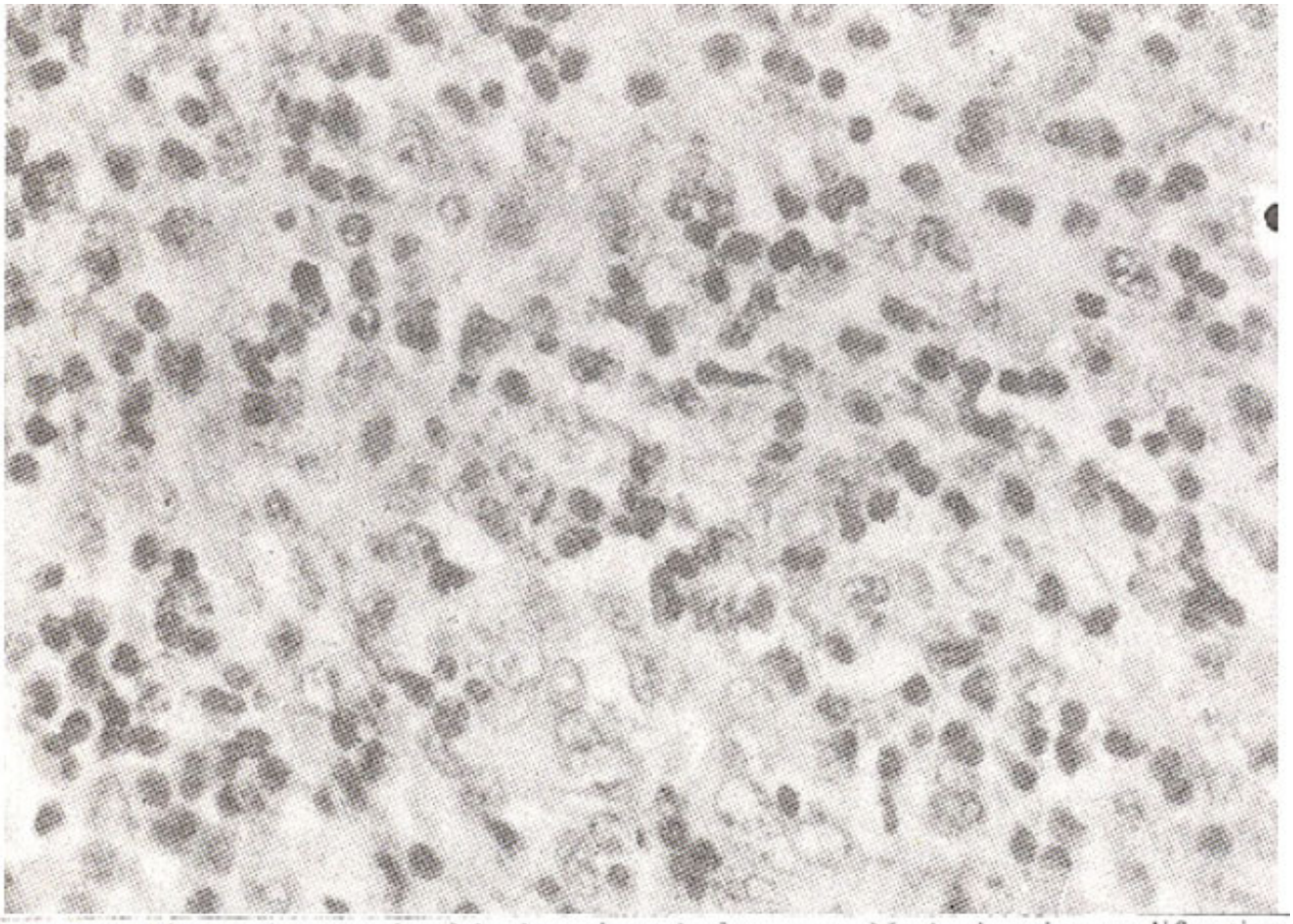

Figure 3. Photomicrograph of the lymph node from case No.1, showing proliferation of histiocytes with lymphocytes, plasma cells and eosinophils. H&E x 460.

Case2

A boy of 12 years presented with a swelling in lower right side of the neck since one year. It increased in size gradually to its present size followed by ulceration. The patient was subjected to routine investigations. His blood picture showed total leucocyte count 7800/cmm, DLC(N.56, L.20, M.3 and E.21) the absolute eosinophilic count 1638/cmm platelet count . was 250000/cmin and ESR 14mm/first hour. The chest, facial and cervical bones on x-rays, urine and stool examinations were nonnal. The lesion was excised and processed for microscopic examination. The morphology revealed predominantly ulcerated skin lesion composed of abundant histiocytes infiltratingthe skin and the subcutaneous tissue accompanied by dense inflammatory infiltrate of lymphocytes, plasma cells, macrophages, scattered neulrophils. Small foci of necrosis and haemorrhages were also evident. These features were consistent with the diagnosis of eosinophilic granuloma of the skin (Figures 4 and 5).

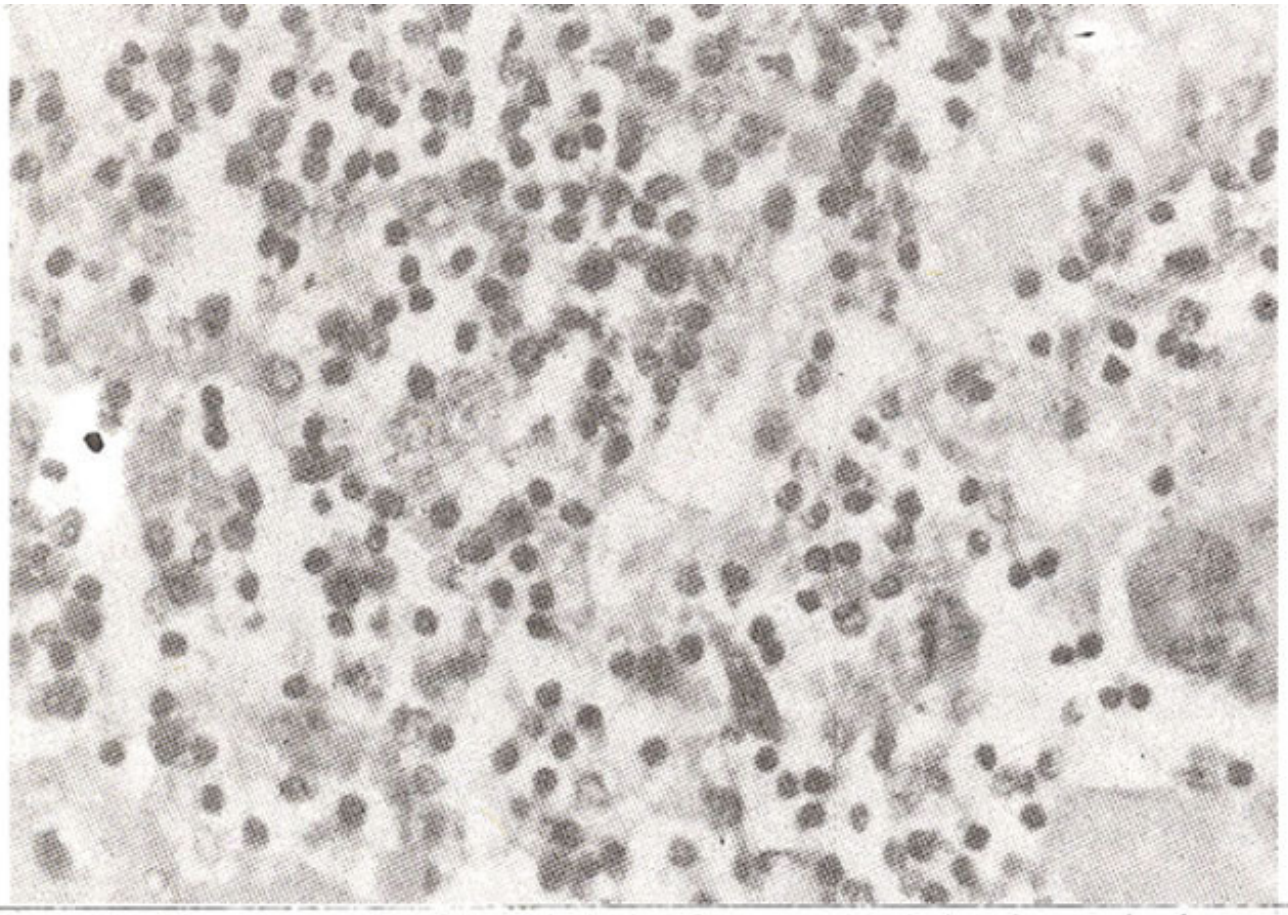

Figure 4. Photomicrograph of the skin lesion from case No.2 showing a mixed population of cells including histiocytes, lymphocytes, plasma cells with many eosinophils H & E x 380.

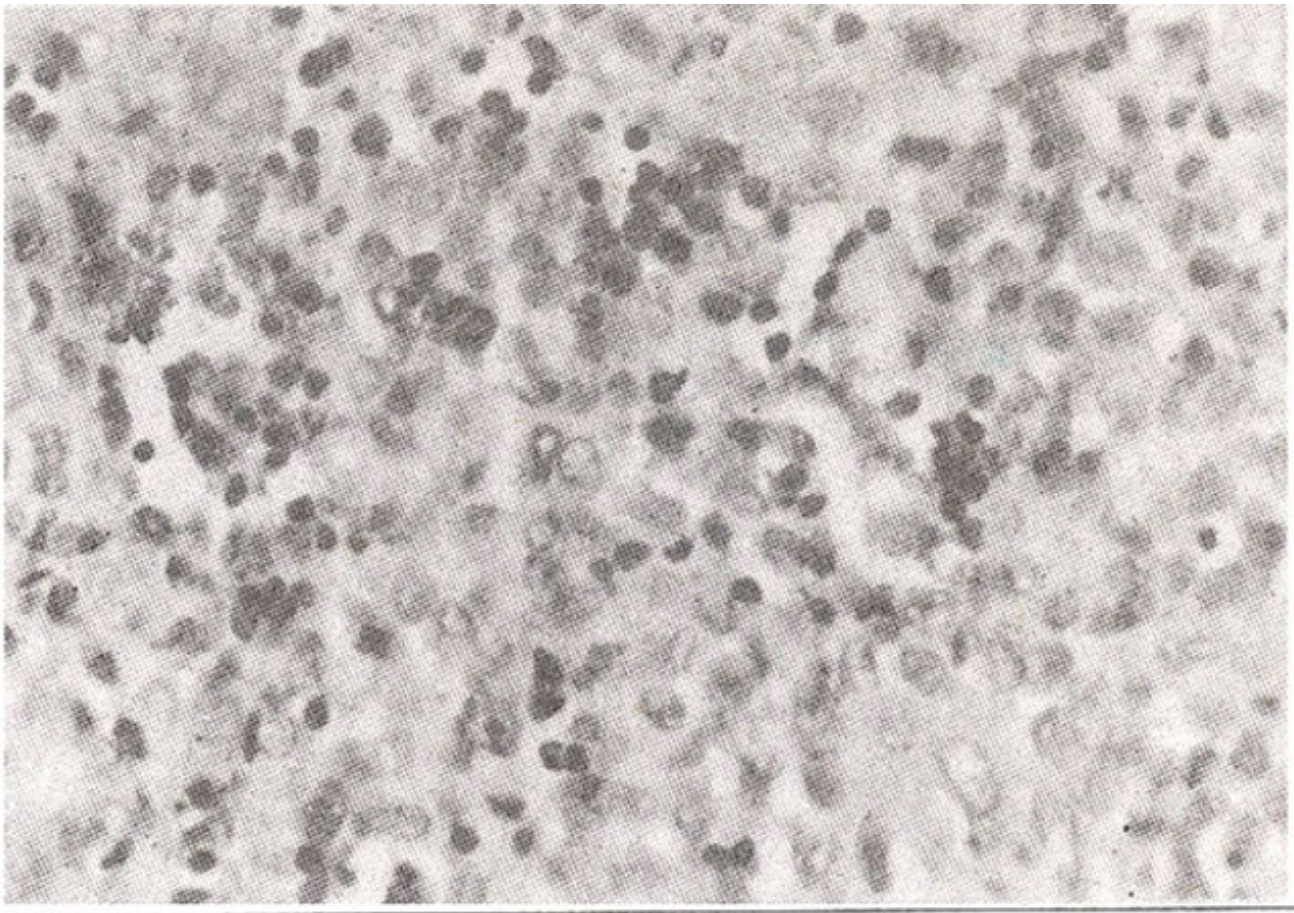

Figure 5. Photomicrograph of the skin lesion from case No.2 showing a diffuse proliferation of histiocytes mixed with lymphocytes, plasma cells and eosinophils. H&E x 460.

Discussion

Eosinophilic granuloma is avanant of Langerhan's cell histiocytosis which have different sites of involvement and clinical presentations[1,2,4,5]. In majority of cases it involves bones and is characterized by the presence of erosive and destructive bony lesions[6]. This disorderis much more frequent in children as compared to adults[1,2,6,7] and multifocal involvement is commoner and a serious form of the disease'. In contrast, unifocal disease, although rare is relatively indolent and potentially curable problem. Sometimes spontaneous healing has been seen, but recurrence is also noted5. Extra-skeletal involvement ineosinophilic granuloma have occasionally been reported in the lungs, liver and Stomach[1,2,5,7]. However, lesions together in the skin and the regional lymph node have not yet been described in the literature, although occasional report is available where either the skin or the lymph node were found involved separately[5,7]. One case presented here showed skin lesion on the face which was considered as primaiy site from where the disease has extendedto aregionallymphnode inthe neck. Inthe othercase only the skin was involved. The exact nature of eosinophilic granulomais still undetermined. The general consensus seems that it is not a neoplastic disease, rather a reactive disorder in which proliferation of Langerhan's cells results from distuthances in immuno-regulation8. The proliferating cell is proved to be the Langerhan's cell of marrow region which is normally found in the epidermis and is believed to be a part of mononuclear phagocytic system[8].

Acknowledgement

We are grateful to Mr. Niamat Ali for typing this manuscript.

References

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