Mycosis Fungoides Granulomatous Variant Revisited Case Report:

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Abstract:

Mycosis fungoides is a peripheral non-Hodgkin's T-cell neoplastic process, representing the most common type of primary cutaneous malignant lymphoma. Histopathologically, though a granulomatous component can occur in all the stages of mycosis fungoides, it accounts for only 4% cases of Mycosis fungoides.

A 63-year-old male presented with a recent history of erythroderma along with generalized tender lymphadenopathy. Peripheral smear of blood showed leukocytosis with atypical lymphocytes. Biopsies were taken from cutaneous plaques and cervical lymph nodes. Immunolabelling study was done to ascertain the origin of atypical lymphocytes in lymph nodes and a diagnosis of granulomatous variant of mycosis fungoides was confirmed. Unfortunately no specific therapy could be instituted as patient died of septicemia within a few days of confirming the diagnosis.

In the present case report, besides stressing on a clinico-pathological correlation, histopathological differential diagnosis of granulomatous mycosis fungoides has been discussed.

Keywords: *Mycosis Fungoides, Erythroderma, Lutzner cells.*

Case Report:

A 63-year-old male presented with a recent history of erythroderma of two months duration. He had vitiligo vulgaris for 26-years.

Examination revealed generalized erythroderma associated with multiple, erythematous, reddish-brown indurated plaques of different shapes and sizes. Face showed diffuse infiltration.

Generalized tender lymphadenopathy was present. Systemic examination was insignificant. Peripheral smear of blood showed leukocytosis (15,800/mm^3). Abnormal lymphocytes were seen (4/100 normal WBC). These atypical lymphoid cells (Fig. 1) had deep clefts, asocial cerebriform nucleus, high nuclear/cytoplasmic ratio and eosinophilic cytoplasm with vacuolations. X-Ray Chest was normal. A provisional diagnosis of erythroderma secondary to lymphoma was made.

Skin biopsy (Figure2) from the plaque showed epidermis of varying thickness with focal atrophy and spongiosis. Occasional Paurtrier's microabscesses were seen. The basel cell layer showed vacuolations. At the dermo-epidermal junction (Figure 3) and in the dermis, single to focal clusters of Lutzner cells were seen. These cells of medium sized and had irregularly folded hyperchromatic nuclei. Granulomas with marked lymphocytic aggregates and giant cells (Figure 4) were present in the upper dermis.

Biopsy from cervical lymph node showed thinned out capsule with obliterated subcapsular sinus. Diffuse infiltration of pleomorphic atypical lymphocytes was seen. The lymphocytes were uninucleate, occasionally multinucleate, with indentations and atypical mitosis.

Immunolabelling study of cervical lymph node (Figure 5) showed presence of CD3$^+$ve atypical lymphocytes, which is indicative of peripheral T-cell lymphoma.

Diagnosis of granulomatous variant of mycosis fungoides was confirmed. Unfortunately, no specific therapy could be instituted as patient died of septicemia within a few days of confirming the diagnosis.

Discussion:

With an annual incidence of 0.29-0.36/1,00,000[1], mycosis fungoides accounts for 40% of all the primary cutaneous lymphomas.[2] Alibert coined the term "Mycosis Fungoides, in 1806.[3]

Granulomatous Mycosis fungodies is one of the several clinico-pathological variants of Mycosis fungoides. Though Pautrier was the first one to report an association of cutaneous T-cell lymphoma and granulomatous inflammations, Ackerman and Flaxman gave the original description of a granulomatous reaction in Mycosis Fungoides.[4] Since then only 28 cases have been reported in literature.[5]

Patients of granulomatous Mycosis Fungoides are usually adult females who present classically with numerous widely disseminated reddish brown patches and plaques on trunk and extremities.[6] Pruritic exfoliation or infiltrated erythroderma may also occur, and 15-

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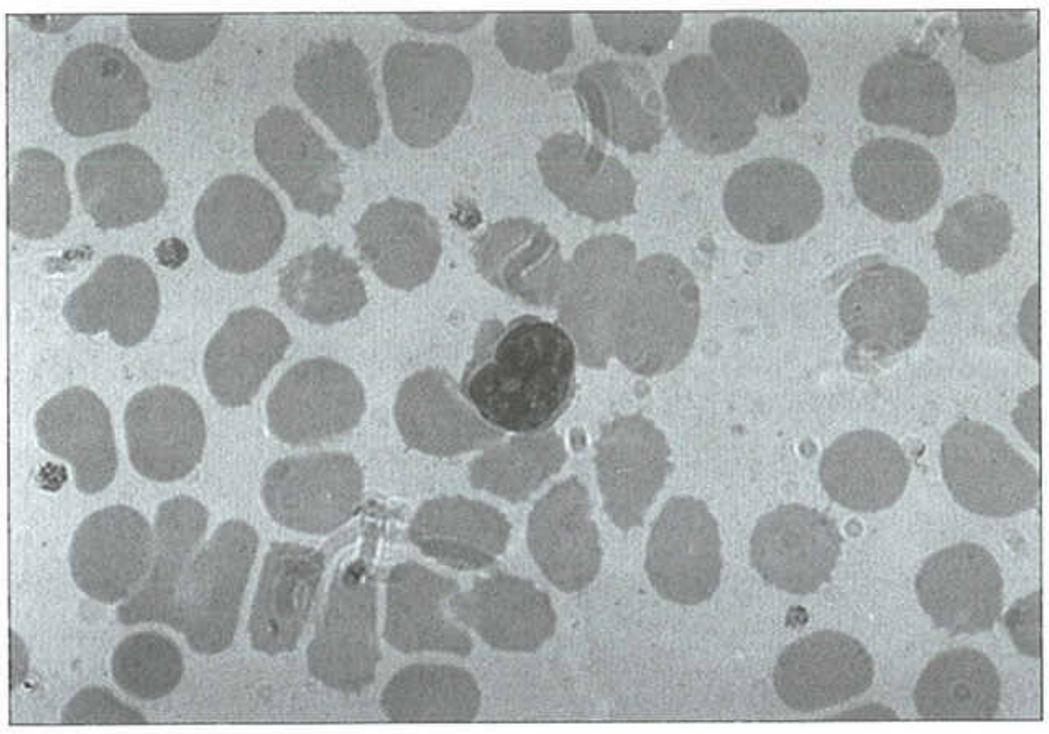

Fig. 1: Peripheral Smear of blood showing an atypical lymphocyte with cleft in nucleus and high nuclear-cytoplasmic ratio.

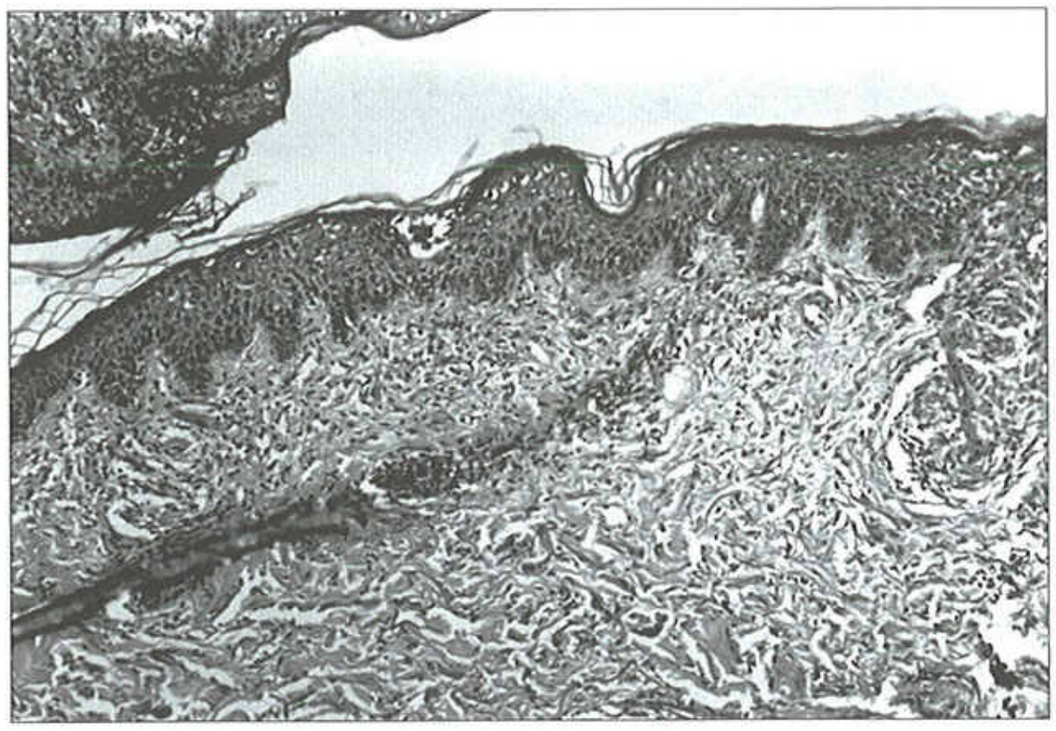

Fig. 2: Skin biopsy showing Pautrier's microabscess in epidermis.

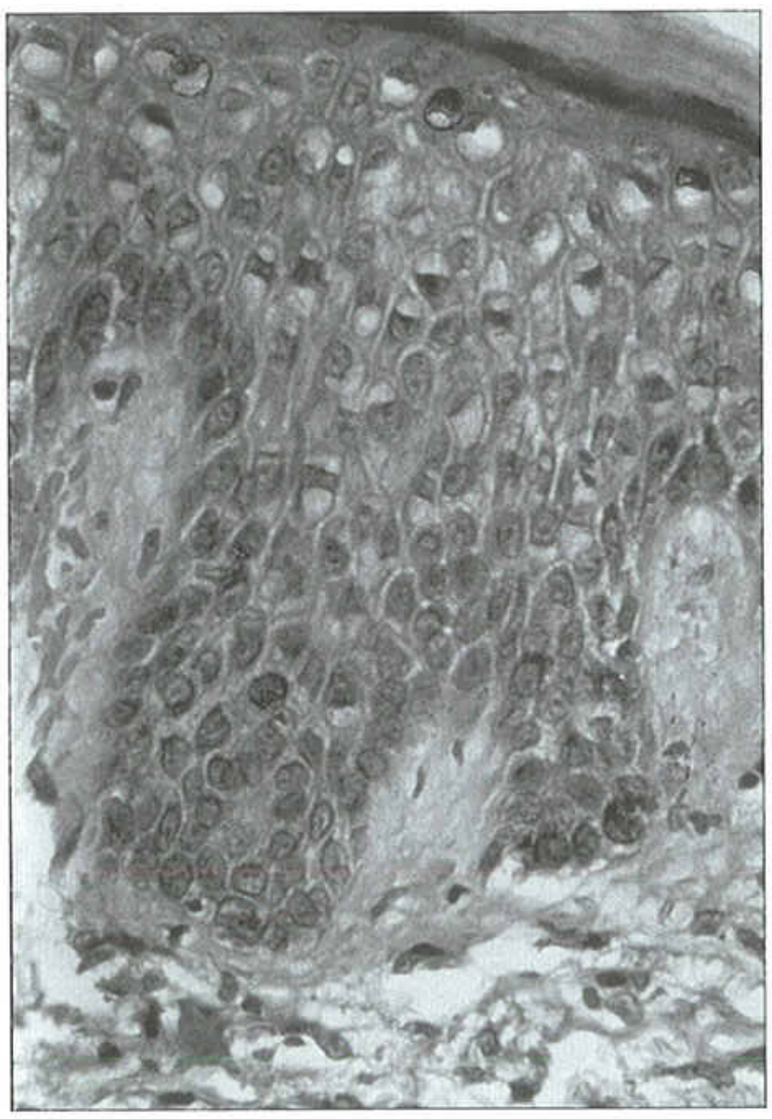

Fig. 3: Skin biopsy showing single to focal clusters of Lutzner cells at dermo epidermal junction and upper part of dermis.

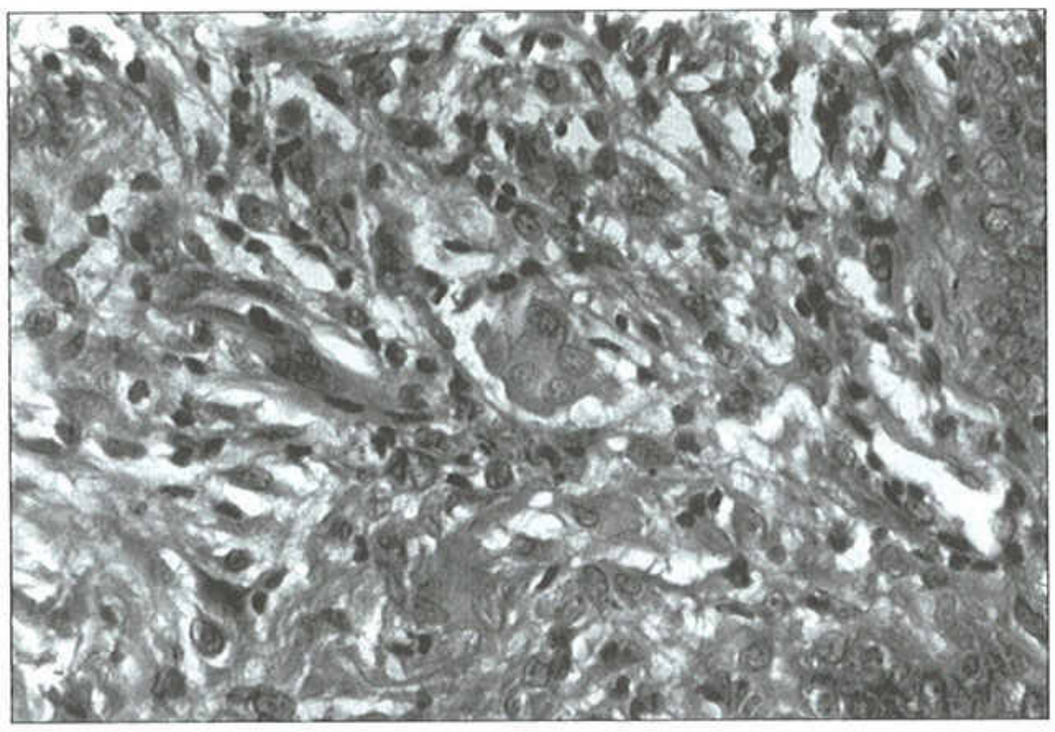

Fig. 4: Giant cell seen in the upper dermis.

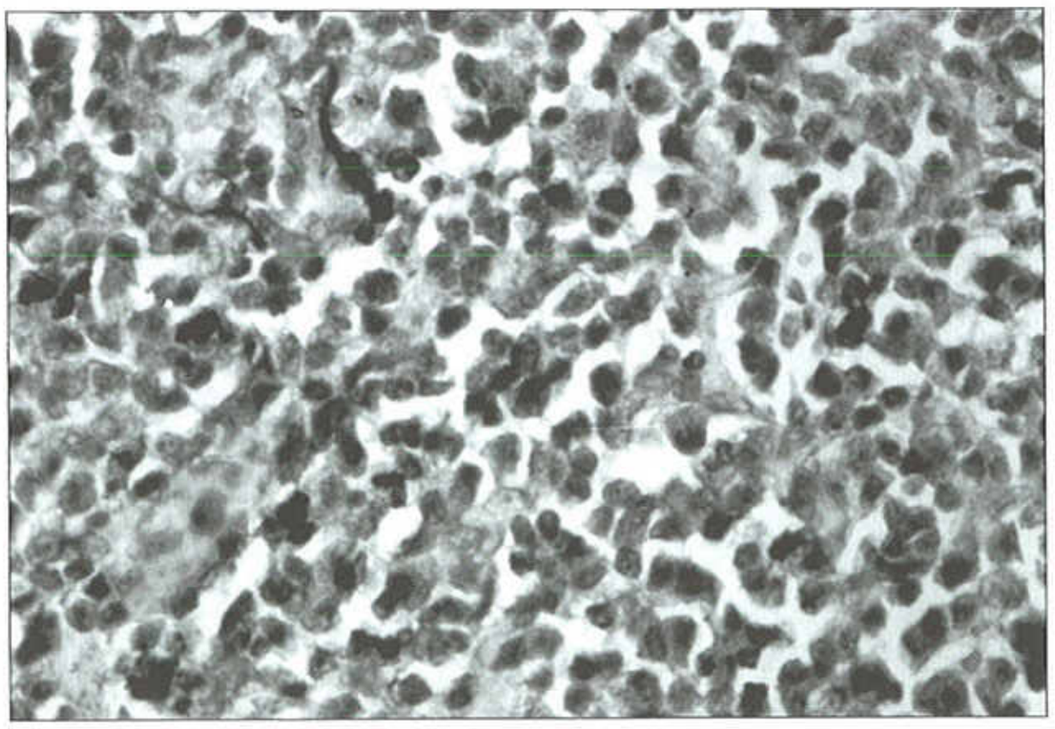

Fig. 5: Immunolabelling study of cervical lymph node showing CD3⁺ atypical –lymphocytes.