Case reports

Cutaneous lymphoma – rare variant

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Sri Lanka Journal of Dermatology, 2004, 8, 23-25

Abstract

Two Sri Lankan males of 87 years and 48 years presented with rapidly enlarging skin nodules accompanied by constitutional symptoms. Histology and immunohistochemistry (Immune markers) confirmed the diagnosis of CD 30 negative cutaneous large T-cell lymphoma. This is a rare entity, which has not been reported in Sri Lanka prior to this.

Introduction

The term Cutaneous T-cell lymphoma (CTCL) describes a heterogeneous group of neoplasms of skin homing T-cells that show considerable variation in clinical presentation, histological appearance, immunophenotype and prognosis.

CTCL represents approximately 75-85% of all cutaneous lymphomas, where primary cutaneous B-Cell lymphoma (CBCL) accounts for approximately 20-25%[1]. One of the distinct subtypes of lymphoma is CD-30 negative primary CTCL. It could be large cell or plemorphic (small/medium) sized CTCL[2].

The characteristic presentation of this form of CTCL is the sudden appearance of solitary or multiple nodules or tumours on the skin with no preceding plaques or patches of typical mycosis fungoides (MF)[2,4].

Case 1

A 87 years old male was admitted to Dermatology Unit of National Hospital Sri Lanka with rapidly enlarging skin nodules of three weeks duration. In addition he had anorexia, loss of weight and generalized pruritus for three months duration.

Examination revealed three purplish nodules located on both arms, left forearm and left groin (Figure 1). Their diameters ranged from 10-15 cm. Left arm lesion was ulcerated.

On general examination there was no regional lymphadenopathy. Other systems were clinically normal.

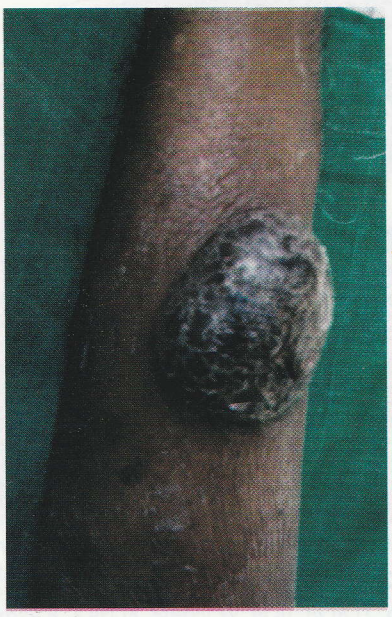

Figure 1. *Nodule on the L arm*

Investigations revealed the following.

ESR 08 mm, FBC Hb 10.8 gldl, WBC 5200/mm3, (N-30, L10, M58, EO2), Pit 180,000/mm3 and no atypical cells in blood picture. Biochemical investigations including liver and renal function tests and serum electrolytes were normal. Ultrasound examination of the abdomen did not reveal any organomegaly or Lymphadenopathy.

Bone marrow examination was found to be normal.

Aspiration from a nodule showed collection of lymphoid cells with predominantly large cells.

Histology showed unremarkable epidermis, dermal infiltration of atypical large lymphoid cells. (Figure 2). These features are compatible with infiltration of primary cutaneous non-hodgkins lymphoma. Leukocyte common antigen was positive in Immunohistochemical Study. We proceeded with flowcytometry and it showed positive T-cell makers (CD3, CD4) and negative CD 30.

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On the recommendation of oncologist, our patient was sent to Cancer Institute, Maharagama for radiotherapy.

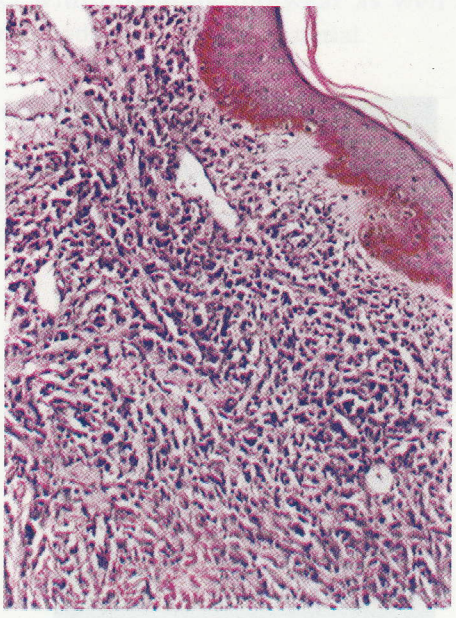

Figure 2. *Histological appearance from skin nodule*

Case 2

A 48-year-old unmarried salesman from Kotahena, presented to Dermatology Unit, National Hospital of Sri Lanka, in April 2004 with asymptomatic, rapidly progressing skin nodules on both upper and lower limbs for 3 months duration (Figure 3). He had associated constitutional symptoms of loss of appetite and loss of weight, but no fever.

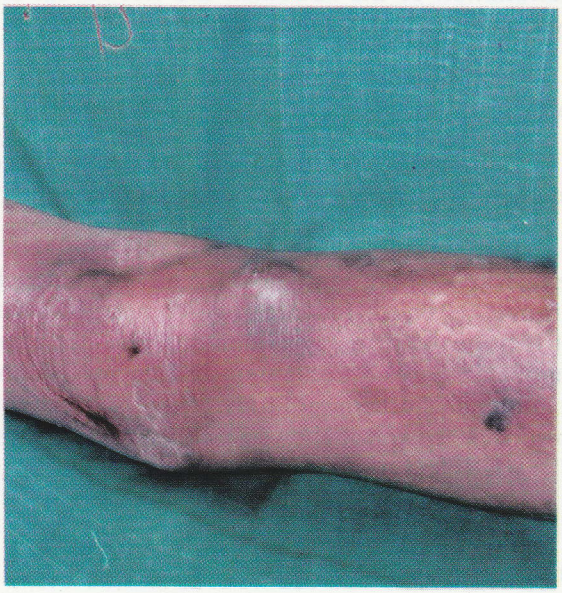

Figure 3. *Nordule from a leg*

These nodules were skin coloured and size varied from 6-8 cm and there were 7 such nodules. Nodules were not tender discrete and firm to hard on palpation. Overlying epidermis appeared normal except ulceration of few nodules on upper limbs. He was emaciated but not pale. There was no lymphadenopathy or hepatosplenomegaly. Examination of cardiovascular, respiratory, gastrointestinal and nervous system were clinically normal.

He was a chronic alcohol abuser and he developed alcohol withdrawal symptoms while in dermatology unit.

At this stage our differential diagnoses were primary cutaneous lymphoma, secondary deposits or subcutaneous fungal infection. Investigations revealed following. Hb 13.4gldl, WBC 6300/mm3, (N-76, L-15, E-5, M-3), platelets 329000/mm3, ESR 28 in 1st hour, SGOT-61 u/L, SGPT – 32 u/L, total serum protein 7.2 g/dl, albumin 3.1 gldl, globalin 4.1 gldl, alkaline phosphatase 148 u/l, and renal profile normal. Chest X-ray, ultra sound scan of abdomen was normal. Fungal smear and culture was negative.

Skin biopsy showed large undifferentiated malignant cells infiltrating entire dermis. No evidence of gland formation and there was no epidermotrophism. These features favour a non-Hodgkin lymphoma infiltration. Leukocyte common antigen was +ve in immuno-histochemical studies and we proceeded to do specific lymphocytic cell markers. It was positive for CD- 3 and Negative for CD-30 and CD-20.

Bone marrow studies were normal. Computed tomography of thorax and abdomen did not reveal any nodal enlargement, masses or organ involvement.

We referred him to Cancer Institute, Maharagama. He was seen by a consultant oncologist and his plan was to give him combined chemotherapy. During 2nd day in the oncology unit, he died due to pneumonia, before receiving the combination chemotherapy as planned.

Discussion

Rapidly enlarging asymptomatic nodules associated with anorexia, loss of weight (and generalized pruritus in case –1) in these patients prompted us to think of a malignant process involving the skin. Taking the monocyte predominant peripheral blood film initially in case – 1, we entertained the possible diagnosis of leukaemic deposits, but the bone marrow was repeatedly normal.

Tumour stage of the MF was the other differential diagnosis. Absence of previous or coexistent plaque

lesions and absence of typical histology excluded the possibility.

In case 2, basic investigations didn't give any positive evidence for our differential diagnoses. Skin biopsy findings favoured a non-Hodgkin's lymphoma infiltration in the dermis without epidermotrophism. At this stage our differential diagnoses were primary cutaneous lymphoma or secondary malignant lymphoid infiltration in the skin. As we couldn't find any primary focus of malignant lymphoma, we concluded that this must be a primary cutaneous lymphoma.

In both patients leukocyte common antigen was positive and it confirmed that these lesions originate from lymphoid tissue. Positive CD-3, CD-4 and negative CD-20 markers confirmed that these tumours arised from skin homing T-lymphocytes.

CD-30 negative CTCL is a distinct histological category. CD-30 negativity is found in tumour stage of mycosis fungoides[3,4]. (In these two patients there was no clinical or histological evidence of MF), CD-30 negative pleomorphic CTCL and pseudo T-cell lymphoma. However the cellular type in our patients was compatible with large cell CTCL.

This accounts approximately for 7% of all CTCL[1]. It is mostly found in adult males with one or several nodules without typical patches of MF. It is known to have a poor prognosis compared to CD-30 negative pleomorphic CTCL. Reported 5 year survival is 15%[1].

Patients should be treated with multiagent chemotherapy. Multiagent chemotheraphy is the accepted mode of treatment[2,3]. Radiotheraphy is useful as initial treatment in localized diseases only.

Acknowledgements

We wish to thank Drs. Ruchira Fernando (Consultant Pathologist) and Nayana Rathnayaka (Registrar Pathology) for helping us with histological studies.

References

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