Primary cutaneous CD 30+ anaplastic large cell lymphoma

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Abstract Cutaneous T cell lymphoma (CTCL) is a broad diagnostic group encompassing a spectrum of diseases with variable clinical presentations. Histologic and immunohistochemical confirmation is needed to establish a precise diagnosis. Primary cutaneous CD 30+ anaplastic large cell lymphoma, a definite variety of CTCL, presenting as a non-healing ulcer is presented here which, needed multimodality approach to heal.

Keywords Anaplastic large cell lymphoma · Non-Hodgkin's lymphoma

Introduction

Lymphomas are tumours of the lymph nodes and lymphatic system and extranodal lymphomas occur in organs and tissues outside the lymphatic system. When lymphomas occur in the skin with no evidence of disease anywhere else at the time of diagnosis, they are called 'primary' cutaneous lymphomas. They are broadly divided into two: Cutaneous T cell lymphomas – thymus derived (CTCL) and B cell lymphomas - B lymphocyte derived (CBCL), the majority being the former. Primary cutaneous lymphomas have variable presentations and need multimodality and multispecialty approach in the management.

Case report

A 45-year-old female presented with a painless, non-healing ulcer in the right gluteal region for over 4 months. She had no other systemic symptoms. On examination, there was no lymphadenopathy and hepatosplenomegaly. Local examination revealed an ovoid ulcer measuring 9 cm × 8 cm in the right gluteal region, which had thickened margins, necrosed edges at some places, thick slough covering the floor, indurated base with serous discharge (Fig. 1). This non-healing ulcer was excised with primary suturing by advancing a local flap. The histopathology was chronic non-specific ulcer. By the 10th postoperative day, the wound gaped and the ulcer reappeared (Fig. 2). Fungal culture was negative, and wedge biopsy of the recurrent ulcer was reported as cutaneous lymphoma T cell type. The immunohistochemistry showed positivity for CD30, CD4 and LCA. However it was negative for CD3, CD20, CD79a, CD43, CD8, CD56, CD15, AKL1 and MPO. The Ki-67 index was high. A final diagnosis of primary cutaneous CD 30+ anaplastic large cell lymphoma was made. CT of the neck, thorax and abdomen were done to

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Fig. 1 Primary presentation of ulcer

Fig. 2 Recurrent ulcer

establish the absence of systemic disease. Bone marrow aspiration and biopsy were done to establish the absence of its involvement. Confirming the primary localized disease of skin, external radiotherapy (50 Gy of 9 MeV electron) was given to the ulcer followed by excision with adequate margins, by closing the defect with a local rotation flap, and the wound healed well. Considering the young age, six cycles of multidrug chemotherapy (cyclophosphamide, doxorubicin, vincristine and prednisolone - CHOP) was also administered, with the advice for a regular follow-up.

Discussion

Anaplastic large cell lymphomas can be CD+ or CD- Primary cutaneous CD 30+ large cell lymphoma is classified as tumour with indolent behaviour and represents about

10% of all cases of CTCL [1]. They are defined according to the following criteria [2]:

- No evidence of lymphomatoid papulosis
- No previous or concurrent lymphomatoid papulosis, mycosis fungoides or other lymphoma
- No extracutaneous localisation at presentation
- Predominance (>75%) of large clusters of CD30+ blast cells in the initial skin biopsy.

They are common in males of 45 and 60 years and present as rashes, plaques, nodules and tumours with central ulceration and may undergo spontaneous regression, but episodic recurrence is common. Trunk and extremities are most commonly involved. They may be mistaken for other skin disorders like adult onset eczema, pyoderma gangrenosum, morphea, localised scleroderma and squamous cell carcinoma [3]. Systemic involvement is rare, and secondary regional lymphadenopathy is seen in only a quarter of cases, and is not a predictor of prognosis [4]. Imaging studies like CT scan of the neck, chest and abdomen are required to stage the disease and to differentiate the primary from the systemic form involving the skin. Bone marrow biopsy may be required to determine its involvement. Biopsy of the lesion is conclusive.

Histology shows diffuse, non-epidermotropic infiltrates with cohesive sheets of large CD30+ tumour cells, oval or irregularly shaped nuclei, prominent eosinophilic nucleoli and abundant cytoplasm with a high mitotic index. Immunohistochemistry is essential in determination of CTCL subtypes, and in the differentiation between primary and secondary disease.

Treatment options include photochemotherapy (psoralen), radiation (electron beam), topical therapy (topical nitrogen mustard), single agent or multiagent chemotherapy and combined therapies (most common being CHOP). For solitary and localised lesions, radiotherapy is the preferred mode of treatment. More generalised lesions require chemotherapy. Multiagent systemic chemotherapy, compared to single agent, does not result in a higher cure rate or prevent future recurrence.

Prognosis is generally excellent for CD30+ lymphomas. Partial or complete remission occurs in 42% of cases, but 40% relapse despite treatment [5]. In view of the risk for systemic progression, though albeit low, prolonged follow up is recommended.

Conclusion

When patients present with non-healing cutaneous ulcers to a surgeon, cutaneous lymphoma should be considered in differential diagnosis.

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