B cell lymphoma with unusual clinical cutaneous presentation

ABSTRACT
Cutaneous B cell lymphoma (CBCL) is a lymphoproliferative disorder of neoplastic B cell of the skin. These are rarer than T cell lymphomas. Commonly, the presenting features of CBCL are plaques or nodules. Here is a case report of CBCL with a very unusual presentation of cutaneous horns. The patient was managed by a combination chemotherapy and local radiotherapy only. He is disease-free for about the last five years. The case has been presented because of its rarity and its management protocol.

KEY WORDS: B cell lymphoma, local radiotherapy, non-Hodgkin’s lymphoma, skin horns

INTRODUCTION
Cutaneous B cell lymphoma is a lymphoproliferative disorder of epidermotropic, neoplastic B cells, with a wide range of clinical manifestations. Incidence of T cell lymphoma is about 85% and the remaining cases are of the B cell origin. The following case is reported because of its rarity, its unusual cutaneous manifestation and some interesting observations regarding the clinical course and management.

CASE REPORT
A 65-year-old male, noted within a short period, successive appearance of two nodular swellings above the right elbow on medial side in December 1999. There was no pain or discharge. Both swellings were growing linearly outwards. The proximal one was growing more rapidly. Both of these ultimately looked like horns. After about a month, he noted painless neck node swellings on the left side. He attended a hospital at Mumbai on June 27, 2000, where the disorder was diagnosed as non-Hodgkin’s Lymphoma (NHL) on the basis of following investigations.

Fine needle aspiration cytology (FNAC) on June 27, 2000, from right arm swellings and neck node was inconclusive as there was only lymphoid aspirate showing floridly reactive lymphoid cells and few large atypical cells. Gun biopsy of the swellings on July 5, 2000, yielded only necrotic material, but wedge biopsy of neck node on July 5, 2000, showed features of non-Hodgkin’s Lymphoma - diffuse large cell type. Immunohistochemistry showed atypical cells reactive to CD 20 (B cell marker), reactive lymphocytes were marking with CD 3 (T cell marker) and chloroacetate esterase was negative. X-ray chest was normal. USG of abdomen on July 18, 2000, revealed no abnormality except mild amount of peritoneal fluid. Bone marrow biopsy on July 31, 2000, was negative.

The patient attended the Radiotherapy Department of our institute on August 3, 2000, for treatment. The clinical features noted at that time were two horn-like projecting masses on medial aspect of right arm at about 5 cm and 8 cm respectively above the medial condyle of humerus [Figure 1]. The proximal one was about 6 cm long and the distal one was about 2 cm long. The skin overlying the lesions was featureless except at the tip of the longer proximal one where there was a small ulcerated area of about 1 cm diameter. Neither of these cutaneous swellings was fixed to deeper structure and there was no induration at the base. Both the masses appeared to arise from the skin.

Two posterior deep cervical lymph nodes were noted on the left side. These were firm, mobile and about 2 and 3 cm in diameter respectively. The biopsy incision mark was visible at the site of the lymph nodes [Figure 2]. There was no other lymphadenopathy, no hepatosplenomegaly and no mass palpable in the abdomen. At Medical College, Kolkata, on the basis of diagnosis of NHL from history and investigations, at first patient was given chemotherapy (CT) in combination with doxorubicin 50 mg/m², cyclophosphamide 750 mg/m², vincristine 1.4 mg/m² - all I.V. day 1; and prednisolone 100 mg orally, day one to five. Six cycles were given at three-weekly intervals from August 28, 2000 to December 27, 2000. The swellings...
became much reduced in size. CT was followed by radiotherapy (RT) over both the swellings from January 30, 2001 to March 8, 2001. The dose of RT was 5,000 cGy in 25 fractions. During radiotherapy, γ ray was used since our center does not have facility of electron beam. There was further reduction in size of both the swellings. The larger one underwent autoamputation during the last phase of RT and smaller one almost disappeared [Figure 3]. Patient is being followed-up regularly. He is disease-free for about the last five years.

DISCUSSION

A lymphoma is considered extra-nodal when the principal expression or the main bulk is at sites other than the lymph nodes.[2] About 40% of lymphomas arise at extra-nodal sites.[3] Usually extra-nodal lymphomas are of non-Hodgkin’s type (NHL). Skin is one of the common sites for extra-nodal lymphomas and B cell type is less common than T cell type. Cutaneous B cell lymphomas (CBCL) are rare and usually secondary to systemic nodal lymphomas. Only recently, the existence of B cell lymphomas presenting clinically in the skin without evidence of extra-cutaneous involvement has been accepted as primary CBCL.[4]

The present case is a B cell lymphoma with some associated cutaneous manifestation. As revealed on biopsy, this is a diffuse large B cell type NHL. Clinically, these lymphomas usually present as papule, nodule or plaque. But the unusual feature in this case is that neither of these is of usual categories of presentation; these were rather horn-like, though initially nodular.

The management plan as for NHL was followed because FNAC of both swellings and cervical lymph nodes showed similar cytological features with a lymphoid background and lymph node biopsy proved non-Hodgkin’s lymphoma - diffuse large B cell type.

The management of CBCL is currently more focused on local or skin-directed therapies, biological response modifiers and extracorporeal photopheresis. Though the current measures could not be applied, the present case showed satisfactory response with chemotherapy followed by local RT using γ rays. The response was excellent.

The unusual clinical cutaneous manifestations in the case report presented here were accepted as NHL because of the following facts: The swellings appeared first and at the onset, these were nodular, subsequently taking the shape of horns. Within a short period, left cervical lymph node swellings were noted; cytological features of both the lymph nodes and the swellings were similar with a lymphoid background. Biopsy of lymph nodes showed features of NHL, the swellings disappeared after treatment and there is no recurrence following the regime of CT and RT.

REFERENCES


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