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Case Report

Primary Cutaneous Anaplastic Large Cell Lymphoma – A Usual Malignancy with Unusual Presentation at an Uncommon Site: A Case Report

Gunjesh Kumar Singh[1], Pragya Singh[2], Priya Baskaran Shanmuga[1], Karthik Periasamy[1], K T Bhowmik[1]

ABSTRACT

Malignant tumors of anal margin and perianal skin are rare, which accounts for approximately 2%-3% of all anorectal malignancies. Because of the variable presentation and pathologic similarity to benign anal lesions, the diagnosis and treatment is often delayed. We report the case of a 54-year-old Indian male who presented with non-healing anal fistula which progressed to a large chronic ulcer and was later diagnosed as anaplastic large cell lymphoma of perianal skin. Primary cutaneous anaplastic large cell lymphoma (PCALCL) of perianal area has been rarely reported, that too masquerading anal fistula is being reported for the first time.

Key words: Cutaneous lymphoma, Anal fistula, Ulcer.

Gunjesh Kumar Singh[1], Pragya Singh[2], Priya Baskaran Shanmuga[1], Karthik Periasamy[1], K.T Bhowmik[1]

[1]Department of Radiotherapy, VMMC & Safdarjung Hospital, New Delhi -110029, INDIA.
[2]Department of Pathology, VMMC & Safdarjung Hospital, New Delhi -110029, INDIA.

Correspondence

Dr. Gunjesh Kumar Singh
MD Department of Radiotherapy, VMMC and Safdarjung Hospital, New Delhi 110029, INDIA.

Phone: +91-8750546284
Email: gunjeshsingh00764@gmail.com

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INTRODUCTION

Malignancies involving the anal margin and perianal skin are relatively uncommon accounting for approximately 2% to 3% of all anorectal malignancies with squamous cell carcinoma being the commonest entity[1] The incidence of PCALCL among peripheral T-cell Non hodgkins lymphoma (NHL) is 1.7%.[2] Patients of cutaneous lymphoma mostly present with solitary or localized nodules, papules or plaques and ulcers. While rare presentations including perianal abscess, hemorrhoids and psoriasis have also been reported for perianal cutaneous lymphomas[3-6] [Table 1]. We hereby report a case of PCALCL involving a rare site i.e. the perianal region, the diagnosis of which got undue delayed due to unusual presentation as anal fistula.

Table 1: Perianal lymphomas and their atypical presentation.

Reference	Primary malignancy	Atypical presentation
Index case	PCALCL	Anal fistula
Hill *et al*[3]	PCALCL	Psoriasis
Dashkovsky *et al*[4]	ATCL	Peri-anal abscess
Jayasekera *et al*[6]	DLBCL	Peri-anal abscess
Gulcu *et al*[7]	MCL	Hemorrhoid

PCALCL: Primary cutaneous anaplastic large cell lymphoma; DLBCL: Diffuse large B-cell lymphoma; ATCL: Angiocentric T-cell lymphoma; MCL: Mantle cell lymphoma.

CASE HISTORY

A 54 year old male presented in general surgery department in June 2016 with chief complaints of swelling in perianal skin with history of intermittent pus and blood discharge for 3 months. On clinical examination the lesion was diagnosed as anal fistula, for which fistulotomy was advised. Patient didn't turn up for the treatment and took some ayurvedic medicine. After 6 months of initial presentation he presented to us with a large ulcer over the anal margin. On examination, there was a solitary ulcer measuring 8×5×3 cm involving 12 to 6 o'clock position in the perianal region with purulent discharge, everted margin and necrotic slough at base [Figure 1]. Per rectal examination could not be done due to the extensive involvement. Patient was advised a course of oral antibiotics and biopsy was done to rule out malignancy. Histopathological examination revealed anaplastic large cell lymphoma with tumour showing positivity for LCA, CD 3, EMA, CD 30, NSE and negativity for CD 19, CD 20, CD 99 and S 100 [Figure 2a, 2b, 2c, 2d, 2e, 2f]. There was no history of fever, night sweat and significant weight loss. Contrast enhanced computed tomography (CECT) neck, thorax, abdomen showed homogeneously enhancing mass involving anal canal, perianal skin and subcutaneous tissue [Figure 3]. Bone marrow examination was normal. Patient was discussed in multidisciplinary clinic and planned for 4 cycles of chemotherapy with cyclophosphamide, adriamycin, vincristine and prednisolone (CHOP) regimen followed by external beam radiotherapy to

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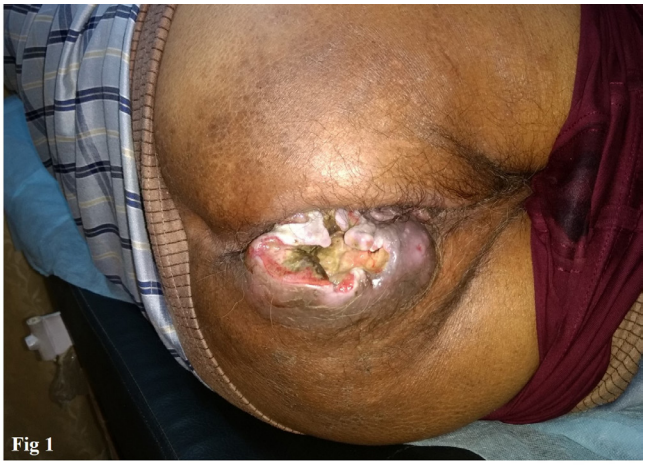

Figure 1: Ulceroproliferative growth measuring 8cmx5cmx3cm involving 12 to 6 o'clock perianal skin.

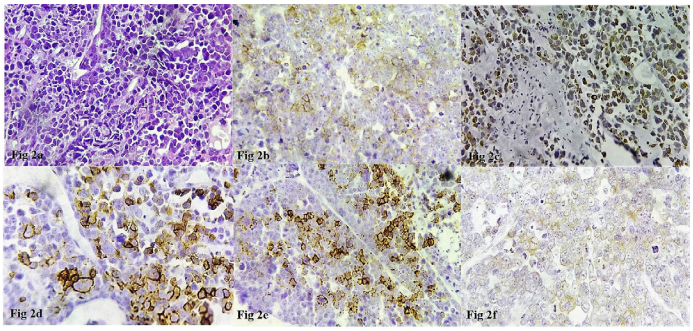

Figure 2a: (40 X) Tumour is composed of medium to large size round cells, arranged in diffuse sheets. The tumour cells have round to oval vesicular nuclei with prominent nucleoli. There is brisk mitotic activity.

Figure 2b, 2c, 2d, 2e, 2f: Tumour cells are positive for LCA, CD3, CD30, EMA, NSE.

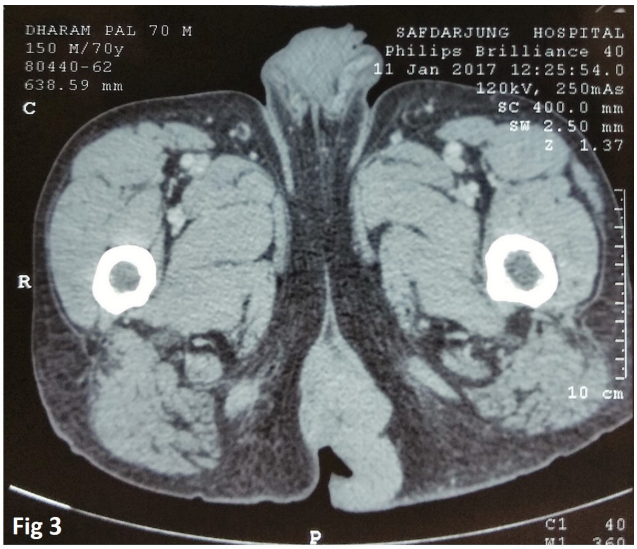

Figure 3: CECT abdomen - Homogeneously enhancing mass involving anal canal, perianal skin and subcutaneous tissue.

pelvis. Patient is currently receiving chemotherapy and is tolerating it well.

DISCUSSION

Anaplastic large cell lymphoma (ALCL) is a subtype of T-cell lymphoma which is biologically and clinically heterogenous. Clinically, ALCL may present either as a localized (primary) cutaneous disease or widespread systemic disease. Skin can be secondarily involved in the systemic disease. The PCALCL mainly affects older patients in the sixth decade with a median age of 61 years.[7]

Clinical features – Presentation may be with solitary or multiple nodules, papules or plaques and sometimes ulcer.[2] Trunk, face and extremities are the commonest sites of involvement.[8] Extracutaneous dissemination occurs sometimes, especially to regional lymph nodes. Involvement of unusual sites with unusual presentations (mimicking anal abscess, haemorrhoids etc) may delay diagnosis and treatment.

Histology - The lesions show diffuse infiltration of skin with large sized T lymphocytes showing positivity for CD4, CD30, CLA, negativity for CD15, ALK and -/+ for EMA and TIA1.[2,8] Despite these, diagnosis of primary cutaneous lymphomas is difficult with lymphomatoid papulosis [smaller <3 cm, diffuse and self-limting] and systemic ALCL with cutaneous involvement [young, lymphadenopathy, B symptoms and a short and progressive course, t (2.5), ALK+] being the commonest differential diagnoses.[9]

Prognosis – It carries a good prognosis with five-year survival rate of 76-96%.[10]

Treatment - Localized radiation therapy or surgical excision is the treatment of choice for localized PCALCL. A radiation dose of 30–36 Gy is recommended which is associated with response rates of greater than 90%.[11] Systemic therapy [e.g - methotrexate, CHOP or CHOP like, interferon-α, retinoids and thalidomide] is reserved for patients refractory to local therapy, with multifocal disease, and/or extracutaneous spread of disease.[12]

The case is being presented here because of the unusual presentation and site of involvement of a usual lymphoma leading to erroneous diagnosis causing undue delay in the treatment and right management thereby contributing significantly to the morbidity.

CONCLUSION

PCALCL in general has indolent nature and carries good prognosis. The rare involvement of perianal skin may mimic benign etiologies; however, unusual diseases such as primary C-ALCL should always be kept in the list of differential diagnoses. A high index of suspicion in non-resolving perianal conditions can prevent delay in diagnosis and treatment. Indexed case will be closely followed up to know the response and behaviour of primary.

CONFLICT OF INTEREST

We authors declare no conflict of interest.

ABBREVIATIONS USED

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