

The Cytological Diagnosis of Extra-Oral Plasmablastic Lymphoma: A Rare Entity

KANTHILATHA PAI, LAKSHMI RAO

ABSTRACT

Non-Hodgkin's Lymphomas (NHLs) which are associated with the Acquired Immunodeficiency Syndrome (AIDS) are heterogeneous. Plasmablastic Lymphoma (PBL) was first recognized as an aggressive, invariably fatal subtype of Non-Hodgkin's Lymphoma which occurred mostly in patients with AIDS, with distinct histomorphologic and immunophenotypic findings, which affected the jaw and

the oral mucosa exclusively. Subsequently, there have been case reports which have described extra-oral plasmablastic lymphomas in the lung, jejunum, caecum, nasal mucosa, etc. We are reporting a case of this rare subtype of diffuse large B-cell lymphoma which presented as a soft tissue mass, which we believe is the first case to be diagnosed by FNAC.

Key Words: Extra-oral, Plasmablastic lymphoma

INTRODUCTION

PBL is a rare form of Non-Hodgkin's Lymphoma that is associated with AIDS, and it is commonly encountered in the oral cavity. There are few case reports on PBL which had occurred in extra-oral locations like the gastro-intestinal tract, the nasal mucosa and the soft tissues. We are reporting an unusual case presentation of PBL, which presented as a soft tissue mass clinically and masqueraded as a soft tissue sarcoma. To the best of our knowledge, this is the first case report on PBL which was diagnosed by FNAC.

CASE REPORT

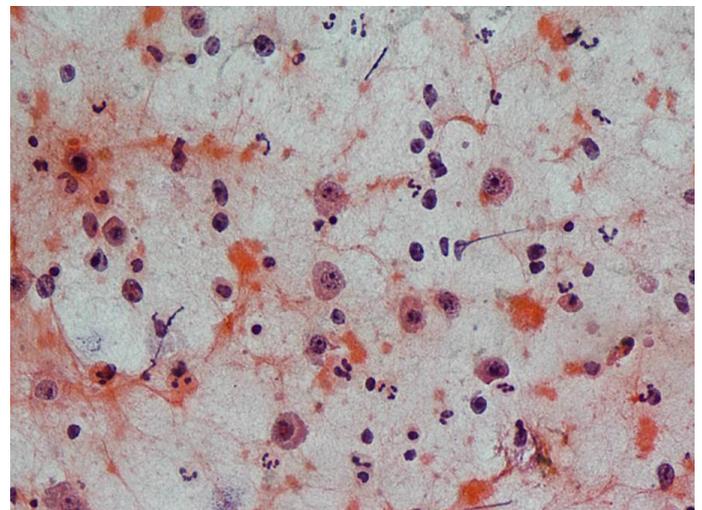
A 35 year old female, who was a known case of retropositive illness on HAART, presented with a swelling in the gluteal region, of 6 months duration, which had rapidly enlarged over the past 2-3 weeks. The patient had difficulty in walking by virtue of its size and also difficulty in lying supine. She gave a history of constipation of 2 weeks duration. She also complained of weight loss and intermittent fever.

On clinical examination, she was found to be poorly built and poorly nourished, with moderate pallor. There was no evidence of lymphadenopathy in the patient.

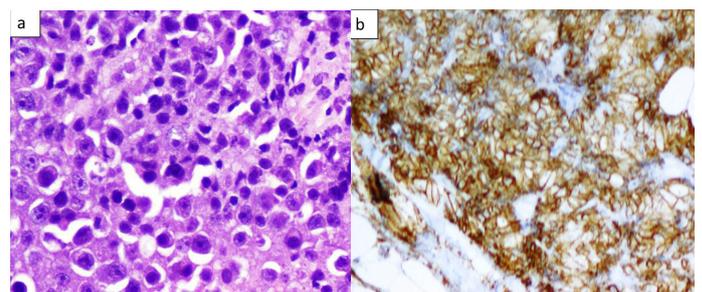
The local examination revealed a large smooth gluteal swelling which measured approximately 20.0 x 10.0 cm, which involved the sacral region and both glutei. There were dilated veins over the swelling. The swelling was hard in consistency. The systemic examination was unremarkable. A clinical diagnosis of a soft tissue sarcoma was made.

Fine needle aspiration was performed, which revealed a haemorrhagic smear with large plasmacytoid cells which had central to eccentrically placed large nuclei, coarse chromatin and prominent nucleoli with abundant densely eosinophilic cytoplasm which exhibited nuclear pleomorphism, occasional binucleate forms and atypical mitotic figures. Also present were smaller atypical lymphoid cells and mature appearing plasma cells, scattered neutrophils, eosinophils and apoptotic bodies [Table/Fig-1]. Serum and urine

protein electrophoresis were requested, to rule out a plasmablastic myeloma, which revealed the absence of paraprotein or free light chains. A diagnosis of an extra-oral PBL was made. A subsequent biopsy showed a tumour which was composed of sheets of plasmablasts, amidst many mature plasma cells and atypical lymphoid cells [Table/Fig-2a]. The immunohistochemical staining with CD₂₀



[Table/Fig-1]: FNAC smear showing plasmacytoid cells with prominent nuclei in a background of atypical lymphocytes, PAP smear, 200x



[Table/Fig-2a & 2b]: Photomicrograph showing sheets of plasma blasts, mature plasma cells and atypical lymphocytes H & E 200X. b) CD 138 diffuse membranous positivity among the cells

was negative, while CD₁₃₈ revealed a strong positivity among these cells [Table/Fig-2b] Based on the clinical presentation, morphology and immunophenotype, a diagnosis of an extra-oral plasmablastic lymphoma was confirmed.

DISCUSSION

PBL of the oral cavity was first described in 1997 [1]; it has been recognized as a distinct entity, a subtype of a diffuse large B-cell lymphoma, by the WHO classification of lymphoproliferative disorders [2]. Since it was first described, more than 150 patients with this disease have been reported [3]. PBLs have been strongly associated with immunodeficiency, most particularly in patients with the HIV infection [3]. PBL accounts for 2.6% of all the HIV-related non-Hodgkin's lymphomas [4].

Recently, the primary sites have been more frequently reported to be outside of the oral cavity or to be concurrent with an oral involvement [5]. Over one-third of all the cases with PBL were first noted at extra-oral locations, and the gastrointestinal tract has been observed to be the most common extra-oral site (10.6%) [3].

The morphologic spectrum ranges from large immature tumour cells (immunoblasts, plasmablasts) to cells with a mature plasmacytic differentiation, which present in a variable and somewhat overlapping maturity range. PBLs usually have a characteristic immunophenotype; they are negative for the typical B-cell antigens (e.g., CD₂₀ and CD₄₅) and positive for the plasma cell markers such as MUM1, EMA, CD₃₈, and CD₁₃₈ [1,3]. PBLs characteristically display a high rate of mitotic activity by the Ki-67 proliferation index [1].

Although it is not specific, the combination of these histopathological features is characteristic of PBL; therefore, immunophenotypic studies must be performed to confirm the diagnosis. PBL should be differentiated from neoplastic plasma cells and other varieties of NHL.

The main differential diagnoses are extramedullary plasmablastic myeloma and diffuse large B cell lymphoma. It is clinically important and critical to differentiate plasmablastic myelomas and PBLs, as the treatment for these two diseases is significantly different

[6]. The distinction between PBLs and plasmablastic myelomas frequently depends on their clinical presentations. The detection of paraproteinaemia in blood and/or excess light chains (Bence-Jones proteins) in urine, lytic bone lesions, and hypercalcaemia or anaemia favours the diagnosis of a plasma cell myeloma over PBL. Immunosuppression, especially HIV-related, is much more frequently associated with PBL. Unlike PBL, a plasmacytoma typically consists of mature plasma cells without a high rate of mitotic activity.

The B-cell lymphomas nearly always express CD₂₀, CD₄₅-RA and CD_{79a}; which is not typical for PBL [1,3].

CONCLUSION

The clinical features of PBL, which include its association with HIV, the male gender and predilection for the oral cavity, may help in the differential diagnosis, however, an extra-oral location in female patients makes it more difficult to suspect it clinically. This case highlights an unusual presentation of PBL which presented as a soft tissue mass. Hence, PBL may be included in the differential diagnosis of soft tissue masses in HIV positive patients.

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