Plasmablastic Lymphoma- Rare tumour in an Uncommon Site: A Case report

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ABSTRACT

Plasmablastic lymphoma is a rare aggressive malignancy of B cell origin. Commonly the nasal and Paranasal regions are involved in HIV positive individuals. We report a retro positive individual presenting with a swelling over the right leg. Clinical and radiological investigations suggested the swelling arising from the extensor digitorum longus (EDL). Differential diagnosis of Rhadomyosarcoma and schwanomma considered after clinico-radiological evaluation. Plasmablastic lymphoma was diagnosed after biopsy and IHC marker analysis.

Plasmablastic lymphoma though common in retro-positive patients, arising from the muscle is uncommon. An aggressive tumour of poor prognosis, delay in its diagnosis and treatment because of its uncommon location can cause rapid spread of tumour and death of patient. In our report we want to create awareness to the treating clinician and pathologist that swellings arising from uncommon site in HIV positive individual should be evaluated to rule out plasmablastic lymphoma.

Key Words: HIV positive, Muscle, Plasmablastic Lymphoma

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Conflict of interest: NO

Case report is Original: Yes

Case reports publish any other journal: No

INTRODUCTION

Plasmablastic lymphoma (PBL) is a rare aggressive lymphoma of B cell origin frequently involved in retro positive patients. HIV related lymphomas are frequently associated with Ebstein barr virus (EBV) and human herpes virus 8 (HHV8). Oral cavity (nasal and paranasal cavities) is commonly involved, rarely testes, skin, CNS and GIT are involved[1,2]. We report a case involving the extensor digitorum longus (EHL) of right leg. To our best knowledge it has not been reported before.

CASE REPORT

A 42 year old male presented with swelling and pain over the right leg for 2 years. The patient was apparently normal about two years back. Swelling and pain insidious onset and progressive in nature, associated with tingling and numbness with no constitutional symptoms.

On examination 16 x 5 cm swelling present on the anterolateral aspect of the right leg. Swelling tender, firm, ill defined with smooth surface and prominent on contraction of underlying muscle. Movements are normal at knee, ankle and subtalar joints. Pulses at distal region are normal. Blood parameters ESR-40 mm/hr, Renal and liver function, total leucocyte and platelet count normal. Patient was diagnosed HIV positive three years back and on irregular treatment. Serology confirmed retropositrive status with absolute CD4 count of 470 cells/µL and CD4/CD8 ratio – 0.28.

Radiological investigation revealed enhanced soft tissue shadow in the anterolateral aspect of leg [FIGURE:1].

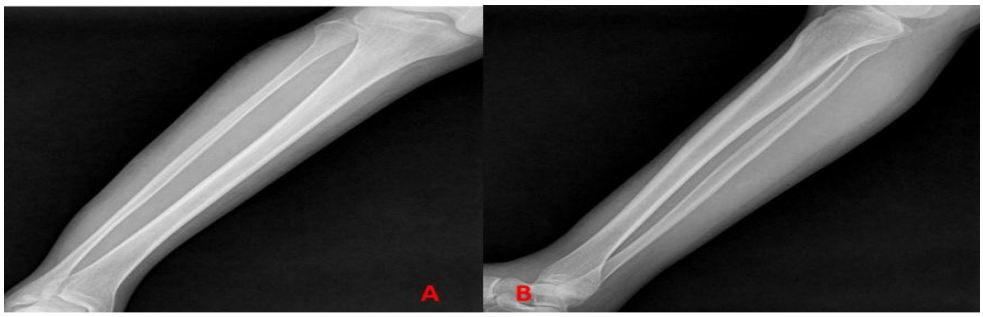

Figure 1: X-ray of right leg anterior-posterior view[A] and lateral[B] view showing enhanced soft tissue density on the antero-lateral aspect of leg.

Colour Doppler showed multiple intramuscular hypoechoic lesions with internal vascularity in anterior aspect of leg and ankle. Chest x-ray showed bilateral normal lung fields. MRI showed multiple lobulated septated lesion in the intra-muscular and inter muscular component of Extensor digitorum longus extending from proximal anterior aspect of leg to the proximal part of foot [FIGURE:2].

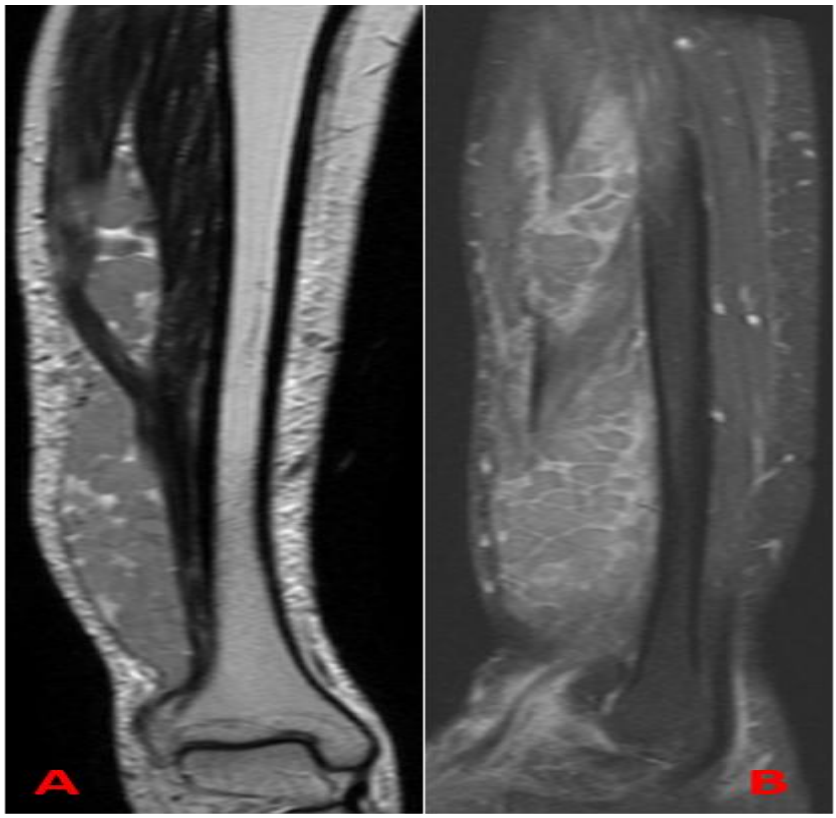

Figure 2: MRI section sagittal [A] and coronal [B] showing multiple lobulated, septated soft tissue lesions within the intramuscular and inter muscular compartment of EDL extending from proximal part of leg to proximal foot.

Intra-operative mass of 15x4 cm was excised with irregular nodular surface and was well vascularised. Mass present in the intermuscular plane abutting EDL with no bony or vascular extension [FIGURE: 3].

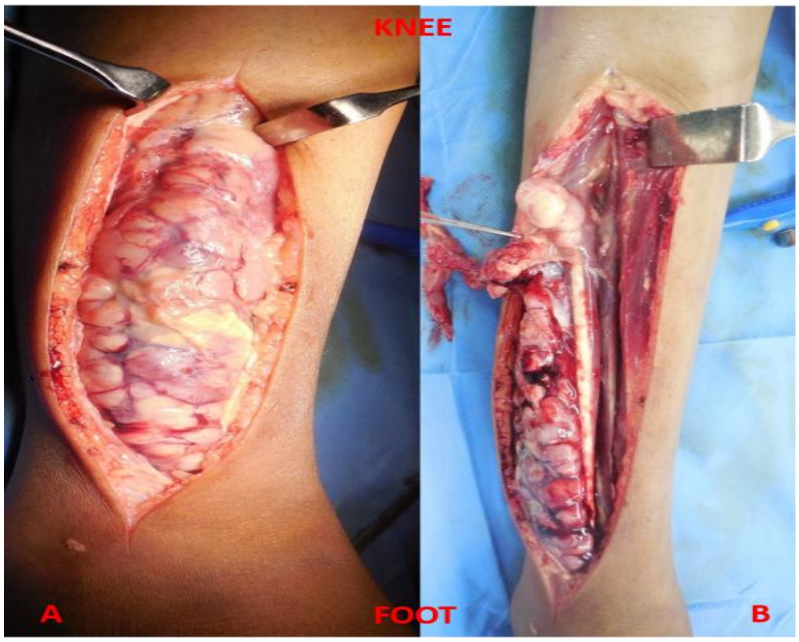

Figure 3: Intra-operative [A] and[B] showing greywhite mass of 15x4x3 cm with irregular nodular surface. Well vascularised tissue in the intermuscular plane abutting EDL with no underlying bony or vascular extension.

Bone marrow biopsy showed no evidence of infiltration. Intra-operative culture was negative. Differential diagnosis of neurogenic tumor (schwannoma) and rhabdomyosarcoma was considered. Histopathological examination of the gross specimen consists of two irregular grey white to grey brown nodular tissue masses weighing 80 gm, largest mass measured 13x5x2.5 cm. External surface appeared multinodular. Cut section shows multinodular grey white tumor measuring 11.5x4 cm [FIGURE:4].

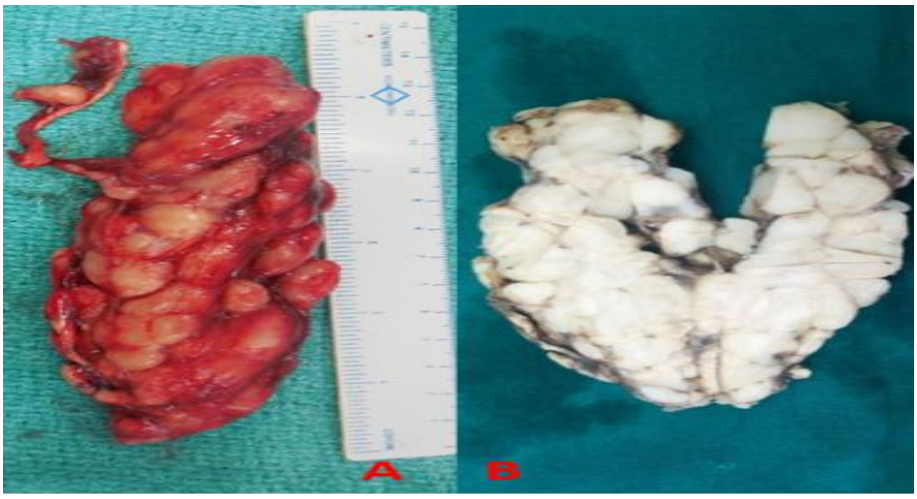

Figure 4: Excised mass gross specimen [A] and cut section[B]. Mass irregular, grey white to grey brown nodular tissue weighing 80 gram. Largest mass[A] measuring 13x5x2.5 cm. Externally surface appears multinodular. Cut section [B] showing multinodular grey white tumour measuring 11.5x4 cm.

Microscopic examination showed ill defined nodules and islands of neoplastic lymphoid cells with hyperchromatic cell and small nucleoli. Cells with moderate amount of eosinophilic cytoplasm suggesting plasmacytoid differentiation with numerous tingible body macrophages [FIGURE 5]. Pheriphery showing foamy macrophages with normal skeletal muscle fibres. Immuno histochemistry showed CD 138 positive [FIGURE:6] and CD 20 negative.

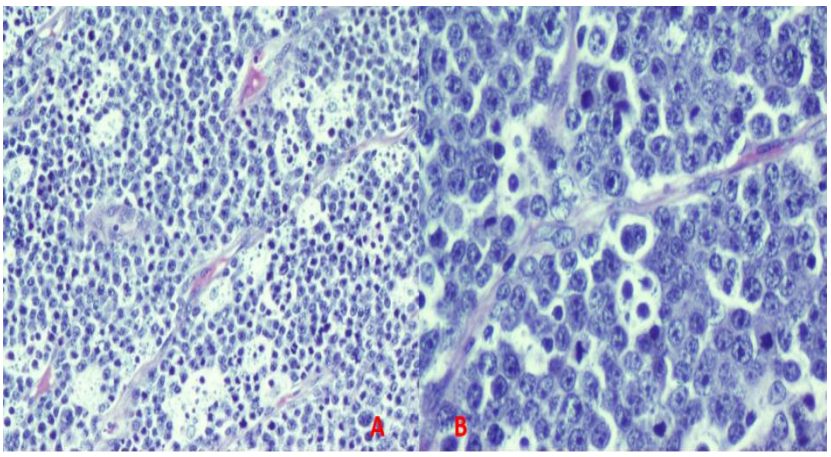

Figure 5: Microscopic section of low power 10X[A] and high power 40X[B] showing ill defined nodules and neoplastic cells. Neoplastic cells being large to medium sized with hyperchromatic nuclei. Cells showing moderate amount of eosinophilic cytoplasm with interspersed binucleate and bizarre forms with numerous apoptotic bodies, mitotic figures and tangible body macrophages.

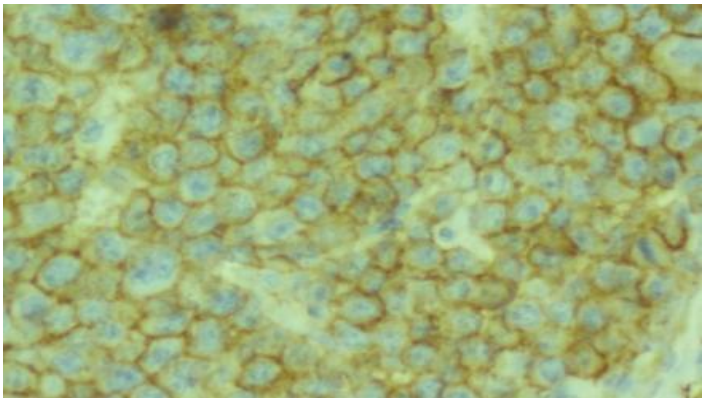

Figure 6: CD 138 diffusely positive immune-histochemical marker.

Biopsy report confirmed Non hodgkins lymphoma- Plasmablastic variety. PET scan for staging showed multiple enlarged metabolically active cervical, axillary, mediastinal, periportal, portocaval, left gastric, para-aortic, mesenteric and pelvic nodes. PET scan also revealed active tonsillar and adenoid hyperplasia with intramuscular soft tissue lesion in the right anterior compartment of leg. Staging of non-hodgkins lymphoma was done and categorised into stage four.

Chemotheraphy was started for plasmablastic lymphoma with cyclophosphamide, adriamycin, vincristine and prednisolone. Totally four cycles of treatment with three weeks interval. Patient now on remission at 12th month of follow up. No clinical and radiological evidence of recurrence. Patient was also instituted on anti-retroviral treatment in view of his retropositive status.

DISCUSSION

Plasmablastic lymphoma is an aggressive B cell tumour. It is a variety of diffuse large B cell lymphoma and a terminally differentiated lymphoma[3]. Plasmablastic lymphoma remains a diagnostic challenge because of its rarity.

Neoplastic cells resemble B cell immunoblasts with plasma cell immunotype which mimics various B cell and plasma cell tumours. It commonly affects males with mean age of 40 yrs. Epidemiological review of literature from 1997-2014, 590 cases of plasmablastic lymphoma with 369 cases HIV positive, 164 cases HIV negative,37 cases post transplant and 20 cases were transformed into PBL[1,3,4]. Incidence of PBL in oral cavity (44%)[5].

Only one case of PBL was reported arising from muscle in a HIV negative individual. To our best knowledge PBL of the muscle (EDL) in a retropositive individual has not been reported before. Pathogenesis involves a complex interaction between immune system, chronic antigenic stimulation, molecular genetics and oncogenic virus like EBV[6]. Excisional bioipsy is considered gold standard.

Clinical and radiological features led to ambiguity in diagnosis and management because of its uncommon location. Differential pathological diagnosis for PBL include immunoblastic diffuse large B cell lymphoma which is CD 20 + and CD 138 –ve. Extra medullary plasma cell myeloma is characterised with anaemia, hypercalcemia and paraproteinemia with or without lytic bone lesion, Plasmablastic lymphoma (PBL) and Plasma cell myeloma(PCM) mimic each other in clinical,radiological and histopathlogy, hence it becomes very important to differentiate this pathology to prevent treatment delay. Extracavitary primary effusion lymphoma with IHC markers CD 30 and CD 38 positive and CD 138 negative, arising commonly from neck region associated with HHV-8 is to be also considered as a differential in retropositive individual[7].

Literature review of 300 patients shows poor prognosis with a median survival of 8 months. Overall response rate to chemotheraphy is 77% with 46% complete response and 31% partial response[8]. Our patient is on remission with no recurrence till now.

CONCLUSION

Plasmablastic lymphoma of muscle in a retro positive individual leads to diagnostic dilemma because of clinical, radiological and histopathology similarities with other haematological malignancies. Awareness of this

aggressive tumour and its possibility to arise from uncommon location is needed to prevent in delay in diagnosis and prompt treatment.

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