Plasmacytic Neoplasm: An Unusual Presentation as a Shoulder Mass in Chronic Immunosuppression

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Introduction

Post-transplant lymphoproliferative disorder (PTLD) is a rare but well-established entity following solid organ transplant as a consequence of long-term immunosuppression. The most common malignancy associated with PTLD is diffuse large B-cell lymphoma (DLBCL). However, other lymphoproliferative disorders have rarely been described, including Burkitt lymphoma and peripheral T-cell lymphoma. Perhaps, the most rarely described presentation has been PTLD manifesting as a plasmacytic neoplasm. We present a case of plasmablastic lymphoma of the shoulder joint in a patient on long-term immunosuppression as a rare presentation of PTLD.

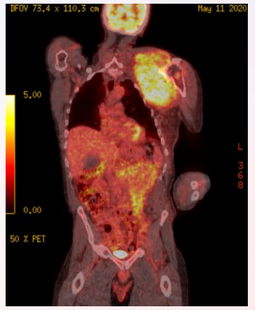
Clinical Presentation

- A 71-year-old male with a past medical history of rheumatoid arthritis treated with methotrexate, etanercept, and adalimumab over the course of 30 years presented with left shoulder pain and edema.
- Physical exam revealed a limited range of motion and diffuse soft tissue edema of the left shoulder region, extending inferiorly along the lateral chest wall.
- After conservative management was unsuccessful, he underwent CT of the shoulder, which revealed severe left glenohumeral joint primary osteoarthritis, full-thickness rotator cuff tear, severe acromioclavicular joint arthropathy, and a poorly defined soft tissue mass associated with the left scapula.
- Further workup with MRI demonstrated diffuse muscular hypertrophy and a likely infiltrative process.
- To determine the etiology of these findings, he underwent ultrasound-guided biopsy of the left infraspinatus muscle.
- Pathology revealed a lambda light chain restricted plasmacytic neoplasm.
- The differential diagnoses included immunodeficiency related lymphoproliferative disorder with plasmacytic features (monomorphic PTLD-like), plasmablastic lymphoma, plasmablastic myeloma, and high-grade B-cell neoplasm.
- Flow cytometry demonstrated atypical cells with CD19, cCD22, CD33, CD38, cCD79a, and CD138 positivity with 74.2% lambda clonal plasma cells.
- Additionally, the neoplasm was positive for Mum1 and negative for CD20, CD30, PAX5, HHV-8, Alk1, EBV, Ki-67 and c-Myc by immunostaining. MYD 88 mutation was negative by PCR.
- FISH myeloma panel demonstrated 17p (TP53) deletion, 1p (CDKN2C) deletion, and additional copies of chromosome 9.
- FISH was negative for BCL2, BCL6, and Myc re-arrangement or amplification.
- Based on the overall findings, the neoplasm was best classified as immunodeficiency-related plasmablastic lymphoma.
- PET scan revealed a large lobulated hypermetabolic soft tissue mass in the left lower neck, axilla, and shoulder compatible with lymphoma (Figure 1).
- Bone marrow biopsy was negative for clonal plasma cells or lymphoproliferative disease.
- He was also found to have new-onset visual changes and headache from hyper-viscosity syndrome with an IgM level 8,010 and viscosity of 5.46.
- Chemotherapy with bendamustine and rituximab was initiated.
- Plasmapheresis was performed for hyperviscosity syndrome.
- The patient responded exceptionally well to treatment with a complete clinical response at three months.

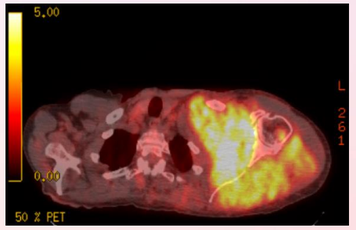
Discussion

This case adds to the scant literature on immunosuppression related plasmablastic lymphoma with a very unusual presentation as a solitary left shoulder mass. A high index of suspicion for an infectious or neoplastic cause should be maintained in patients on long-term immunosuppression. The patient's diagnosis was confounded by his osteoarthritic changes and history of rheumatoid arthritis. Prompt biopsy of the soft tissue mass led to a conclusive diagnosis and successful treatment.

Figure 1. PET

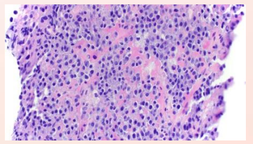


A. Coronal View of Plasmablastic Lymphoma

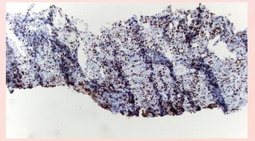


B. Axial View of Plasmablastic Lymphoma

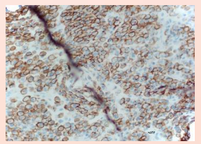
Figure 2. Immunohistochemical Staining



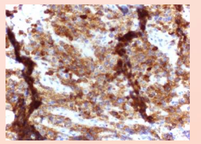
A. Left Shoulder Mass Histology



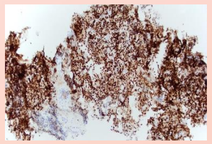
B. Ki-67



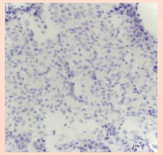
C. CD138



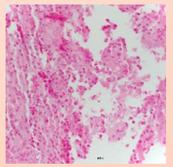
D. CD79a



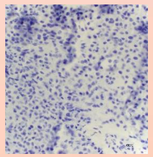
E. MUM1



F. CD20



G. EBV



H. HHV-8

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