Case of HIV positive multiple solitary recurrent extra medullary plasmacytoma

Dr. Koushik*, Dr. Shailaja, P.Jadhav*, Dr. Lata bhoir**

* Department of General surgery, B.J. Government Medical College and Sassoon general hospital, Pune 411001, India
**Assistant Professor Department of Surgery BJ Medical College and Sassoon Hospital Pune India

***Abstract*- Objective –** HIV is known to increase the chances of occurrence of B-cell neoplasms. There have been citations of cases of primary solitary plasmacytomas at various sites in HIV positive patients. We present a case of HIV positive initially diagnosed a solitary extra medullary plasmacytoma post radiotherapy recurring as multiple solitary extra medullary plasmacytomas involving de novo soft tissue sites which is a rare presentation

Design – case report

Result—63yr old HIV positive male patient on HAART presented with solitary extra medullary plasmacytoma of left maxillary sinus in the year 2010 and was treated by radiotherapy alone. He presented in the year 2012 with recurrent multiple extra medullary plasmacytoma involving the recto sigmoid, chest wall skin and left axilla. This patient had absent M band on electrophoresis. Patient was operated with Hartmann's procedure and wide local excision for chest and axillary swellings.

Conclusion – extra medullary plasmacytoma can recur at multiple soft tissue sites. HIV infection is known to cause increase in severity of B-cell neoplasms. Literature search reveals cases of plasmacytoma associated with HIV. Case of multiple solitary recurrent plasmacytoma in a case of HIV is rare and has not been cited.

***Index Terms*-** extramedullary plasmacytoma, recurrent EMP, HIV with EMP.

I. INTRODUCTION

Plasmacytoma is an immune proliferative, monoclonal disease of the B-cell line and is classified as non-Hodgkin lymphoma. It originates as a clone of malignant transformed plasma cells. Extra medullary plasmacytomas are unusual plasma cell tumors arising outside the bone marrow. Isolated EMPs are rare tumors and comprise 4% of all plasma cellular diseases. Preferred site of EMP is the Upper aero digestive tract in 82.2%. Human immunodeficiency virus (HIV) is likely to play a role in the onset of plasma cell tumors (PCT). In fact, HIV could be involved in plasmacytomagenesis in several ways most importantly chronic viremia and antigen presentation to B cells and dysfunctioning T-cells in a background of altered cytokines and interleukins.

We present a case which had a combination of uncommon presentations in the literature on EMP, in terms of being a recurrent case with multifocal involvement and involving the less common sites like colon and skin and being associated with HIV positivity.

II. CASE REPORT

A 63 yr old PLHIV male patient presented with the chief complaints of enlarging perianal, chest wall and axillary masses since 6 months. Patient was diagnosed of left maxillary sinus plasmacytoma in the year 2010 when he had received 25 cycles of radiotherapy totaling to 4000cGy. The then investigations included complete haemogram, chest roentgenogram which were normal. Serum electrophoresis revealed absent M band and bone marrow biopsy was normal. In the current presentation patient had a large 10*7 cms mass in the perianal region situated to the right of anal orifice. On per rectal examination the mass was extending along the anorectal wall occluding the lumen. The proximal extent of the mass was not identified.

There was a 4*4 cms mass on the anterior chest wall just right of midline non tender firm with excoriated overlying skin, non fluctuant reddish in color.

One more mass of 3*4 cms present in the left axilla close to the medial wall firm to hard in consistency, mild tenderness present. Not freely mobile. Overlying skin being normal. No associated neurovasculopathy in left upper limb

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Figure 1 perianal mass

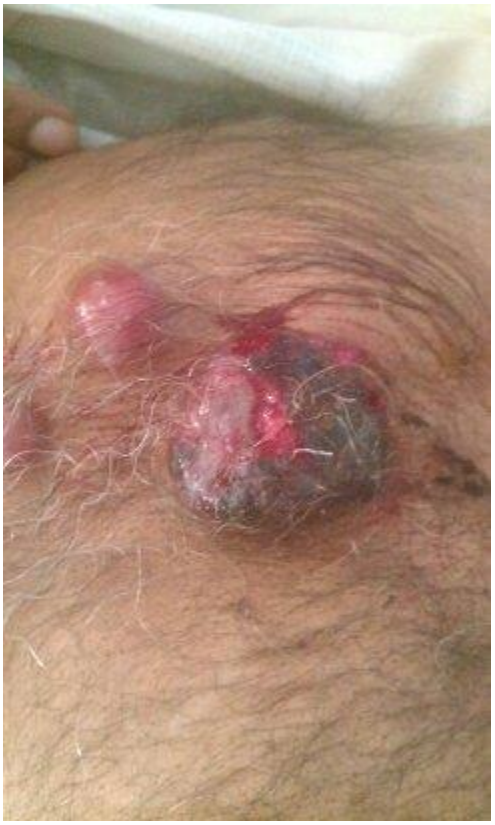

Figure 2 chest wall mass

Patient was moderately built and nourished. With no associated comorbidities in form of hypertension, diabetes, cardiovascular diseases. He had no significant past medical or surgical history.

Treatment history includes HAART for HIV with a CD4 count being 327

No abnormality found in systemic examination.

Complete haemogram revealed normal result. Renal and liver function tests were normal. Total serum proteins being 5.6 with maintained albumin and globulin ratio. Chest x-ray revealed calcified granuloma in the upper and mid lobes of right lung with bilateral bulk hila. No bony lesions were noted.

Patient did not consent for bone marrow examination in this admission. FNAC report from both chest wall and perianal lesions were suggestive of neoplastic lesions with plasma cells seen in clusters and containing large eccentrically placed nuclei suggestive of plasmacytoma.

Patient was operated with all universal precautions taken. Abdominoperineal resection of the mass was done by infra umbilical midline laparotomy. End sigmoid colostomy was done. Resection was done with a proximal 5 cms clear margin. Intra operatively there was evidence of 4*4 cms mass in the recto sigmoid region with the mass extending towards right involving the right levator ani muscle. And there was also involvement of anal sphincters. There was also another 3*3cms parietal peritoneal mass in lower abdomen. Laparotomy wound closed with tension wire suturing. Wide local excisions of Chest wall and axillary masses were done.

Histopathological report of all three masses was consistent with plasmacytoma.

Patient had an uneventful recovery with tension abdominal sutures removed on 21st post-operative day.

The limitations In this case were inability to perform a repeat bone marrow study in this admission due to lack of patient's consent and lack of immunohistochemistry testing at our facility to further investigate the patient. But in view of having a normal total protein and globulin levels, normal skeletal imaging, no anemia and renal failure, i.e. absence of major and minor criteria to diagnose multiple myeloma, the possibility of patient having progressed to multiple myeloma post radiotherapy in the past appears less likely.

Patient had been followed up for a period of 6 months and later lost for follow up.

III. Discussion

Plasmacytoma, or plasmoma, was mentioned by Unna in I89I and first described by Schridde in I905. It is a tumor composed almost exclusively of plasma cells arranged in clusters or sheets with a scant, delicate, supportive, connective tissue stroma.

Plasma cell diseases originate from pathologic plasmablasts that dedifferentiate during the maturation process from primary and secondary B blasts to plasmablasts into malignantly transformed plasmablasts situated in the bone marrow. They migrate and then return to establish themselves in the bone marrow. In rare instances, with the assistance of adhesion molecules, they also may settle in soft tissue or in an extracellular connective tissue area. This is the origin for monoclonal plasma cell foci located outside the bone marrow, called extra medullary plasmacytoma (EMP)[1]

The different types of plasma cell tumors are: 1.) MGUS 2.) Related organ or tissue injury (end organ damage). 3.) asymptomatic myeloma (smouldering myeloma) 4.) Symptomatic multiple myeloma. 5.) Solitary bone plasmacytoma. 6.)extra medullary plasmacytoma. 7.) Multiple solitary plasmacytomas (+/- recurrent) 8.) plasma cell leukemia[2]. Isolated EMPs are rare tumors and comprise 4% of all plasma cellular diseases

In a review of more than 400 published articles, 82.2% of extra medullary plasmacytomas were found in the upper aero digestive tract with 17.8% arising in the gastrointestinal tract, urogenital tract, skin, lung, and breast in that order. Although liver, spleen, and lymph nodes are common extra medullary manifestations of multiple myeloma, primary extra medullary plasmacytomas of these organs—including the pancreas and adrenal gland—are extremely rare[1]

Human immunodeficiency virus (HIV) is likely to play a role in the onset of plasma cell tumors (PCT). In fact, HIV could be involved in plasmacytomagenesis in several ways: it has the ability to lessen the immunosurveillance to such a degree as to impair the immune response against tumor cell growth[3].

HIV-induced immune-cell activation is one of the few widely accepted hallmarks of HIV pathogenesis and disease progression. The hyper activation of B cells by HIV is characterized by several features: hypergammaglobulinaemia; increased polyclonal B-cell activation; increased cell turnover; increased expression of activation markers, including CD70, CD71 (also known as TFRC), CD80 and CD86; an increase in the differentiation of B cells to plasmablasts as measured by

phenotypical, functional and morphological measures; increased production of autoantibodies; and an increase in the frequency of B-cell malignancies[4].

In general, there are no international guidelines for the treatment of EMP. However, based on the well-known radiation sensitivity of the plasma cell tumor, radiotherapy is accepted as the treatment of choice for EMP[5].

Surgery alone or combined surgery and RT or chemotherapy has also been used in the treatment of EMP

The main findings in our case are

i) Recurrent case of multifocal plasmacytoma involving the anorectal region with extension as a perianal mass, chest wall and axilla.
ii) No anemia, no impaired renal functions, no hypercalcemia
iii) Though serum electrophoresis was not done, in view of normal serum protein levels and normal albumin to globulin ratio and a normal urine routine and microscopy reports, M band is unlikely to have been present in this patient.
iv) Patient is HIV positive on long term HAART.

These findings suggest this to be a case of multiple solitary plasmacytoma- recurrent type --- as per international working group on myeloma classification 2003.

In conclusion HIV positive patients are as such known to be increasingly susceptible for plasma cell neoplasms due to HIV induced B cell dysfunction. Plasmacytomas can occur at multiple sites and can be recurrent. Further studies are hence required to know if HIV positive status has a role in recurrence of plasmacytomas. Patients who are diagnosed with plasmacytoma should be screened for HIV status as this can predict a poorer prognosis/recurrence[6, 7].

Further studies are required for establishing the role of PET CT and immunohistochemistry in routine work up of a diagnosed case and in establishing a protocol for follow up of patients with plasmacytomas.

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AUTHORS

First Author – Dr. Koushik – resident at department of General surgery in B.J. government medical college and Sassoon general hospital PUNE INDIA 411001
Second Author – Dr. Shailaja .p. jadhav- professor and head of the unit department of general surgery – B.J. government medical college and Sassoon general hospital PUNE INDIA 411001
Third Author - Dr. Lata Bhoir, assistant professor department of surgery BJ medical college and Sassoon hospital Pune India

Correspondence Author – Dr. koushik. Email address- koushik.herle@gmail.com contact no - +91 8407987044