Retroperitoneal Plasmacytoma: A Case Report

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Summary:

Solitary Extramedullary Primary Plasmacytoma (SEMPP) is a rare neoplasm. When diagnosed, head and neck region is its most likely location. Rarely, it may occur in the retroperitoneum. We report a case of an elderly male who was admitted in the department of surgery, Dhaka Medical College Hospital (DMCH) with a Solitary Extramedullary Retroperitoneal Primary Plasmacytoma (SEMRPP). Subtotal excision of the mass was done. The patient was referred to the department of Oncology DMC for radiotherapy. The first report of an extramedullary plasmacytoma was in 1905 in Taiwan. To the best of our knowledge, there was no report of a Solitary Extramedullary Retroperitoneal Primary Plasmacytoma (SEMRPP) from Bangladesh.

Keywords: Extramedullary, Plasmacytoma, Retroperitoneum.

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Introduction:

Plasmacytoma, a neoplastic proliferation of plasma cells, may be primary or secondary to disseminated multiple myeloma and may arise from osseous (medullary) or non-osseous (extramedullary) sites. Approximately 80-90% of extramedullary Plasmacytomas (EMPs) involve the Mucosa Associated Lymphoid Tissue (MALT) of the upper airways and 75% of these involve the nasal and paranasal regions.[1] Isolated plasmacytomas are rare, comprising only 4% of all plasma cell malignancies. EMP is an uncommon low grade malignant neoplasm with relatively good prognosis. Rarely, EMP may occur in the retroperitoneum. The first report of an EMP was in 1905 in Taiwan[2]. We report a 60 years elderly man with a Solitary Retroperitoneal Extramedullary Plasmacytoma (SREMP). This is probably the first case reported in Bangladesh.

Case Report:

A 60-year old man was referred to our hospital in March 2008 with the history of a swelling in the left side of upper abdomen for last 1 year, which was not increasing in size, and mild pain in the swelling for last 5 - 6 months. He did not have history of fever, anorexia, weight loss, vomiting and bowel or bladder dysfunction. He had two episodes of sudden sweating, palpitation, with restlessness and anxiety within last one year. Physical examination revealed a non-pulsatile, oval, firm to hard, rough surfaced, slightly tender, intra-abdominal lump (measuring about 15x9cm) occupying the left hypochondriac region without local rise of temperature. There was no pallor; icterus, lymphadenopathy and both testes were normal. Ultrasound report revealed a large solid mixed echogenic retrogenous mass (15.2X11cm) seen in the left upper quadrant of abdomen separated from liver, left kidney, left suprarenal gland and aorta. Body and tail of pancreas were pushed forward by the

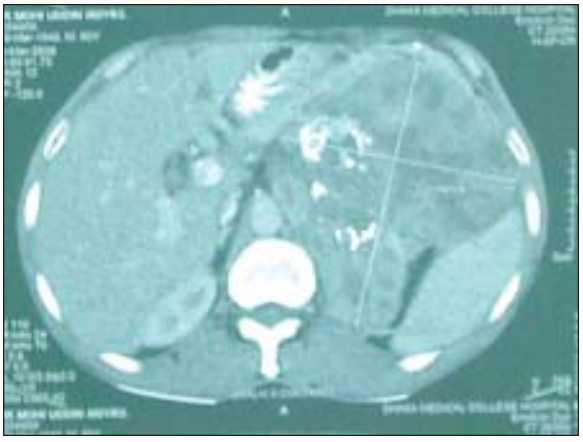

Fig.-1: *Computed tomography revealing a soft tissue between spleen and stomach and anterior to left kidney Distal body and tail of the pancreas are pushed anteriorly by the mass*

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mass. CT scan of abdomen (Figure: 1) revealed a mixed density mass with amorphous calcification measuring about (15X14X12) cm in the left side of abdomen in between spleen and stomach and anterior to left kidney. Distal of body and tail of the pancreas was pushed anteriorly by the mass; showing left sided large retroperitoneal mass. US guided Fine Needle Aspiration Cytology showed the mass compatible with adreno-cortical adenoma / carcinoma.

Investigations revealed Haemoglobin 10.5 gms/dl, Erythrocyte Sedimentation Rate (ESR) 120 mm in 1st hour, Total Leukocyte Count (TLC) was normal. Renal function tests, hepatic enzyme levels, serum electrolytes, serum calcium were within normal limits. Plasma protein electrophoresis revealed polyclonal gammopathy. Skeletal survey was normal. Bone marrow aspiration and biopsy revealed myeloid hyperplasia. Subtotal excision of the retroperitoneal mass (Figure 2 & 3) was done, leaving small upper portion of the mass that was firmly adhered with great vessels, spleen and left dome of the diaphragm. Post operative recovery was uneventful.

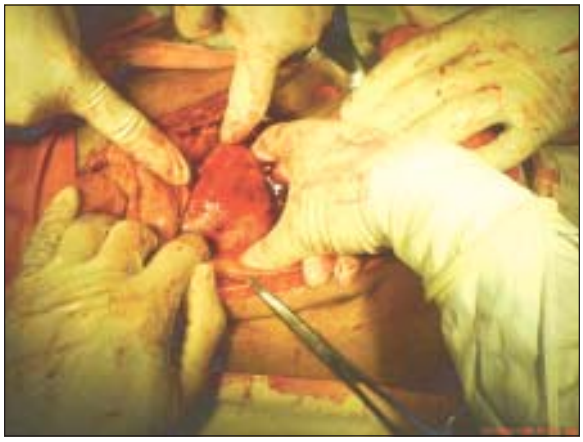

Fig.-2 & 3: *Figure showing huge retroperitoneal mass with piecemeal resection.*

Histopathology revealed sheets of mature and immature plasma cells (Figure: 4) suggestive of plasmacytoma. A final diagnosis of solitary retroperitoneal extramedullary plasmacytoma was made.

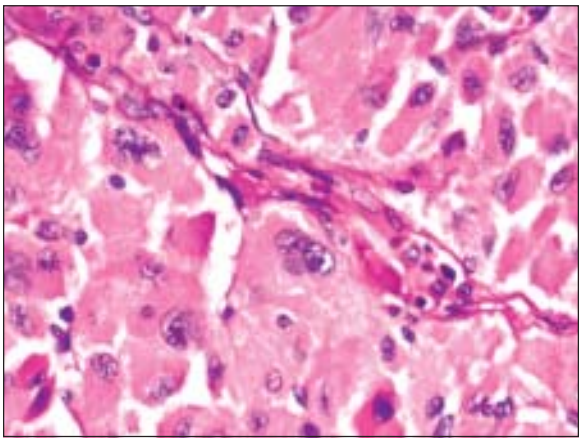

Fig.-4: *Showing picture of the retroperitoneal plasmacytoma under microscope.*

Discussion:

Extramedullary Plasmacytoma (EMP) constitutes 4% of plasma cell tumours. It is defined as a solitary tumour composed of monoclonal proliferation of cells with plasmacytic differentiation in an extramedullary site.[3] It is classified as either primary EMP (when there is absence of coexisting multiple myeloma) or secondary EMP (when it is associated with multiple myeloma). EMP most commonly occurs (>90%) in head and neck region. Other documented sites include gastrointestinal tract, CNS, urinary tract, thyroid, breast, testis, parotid glands and lymph nodes.Solitary EMP in the retroperitoneum is very rare. Marks [4] reported bilateral renal vein occlusion with renal failure and fatal haemorrhage due to tumour erosion and vena caval perforation in case of a RPEMP. Kobayashi et al [5] has reported tumour thrombus within the renal vein in case of RPEMP involving kidney.

The differential diagnosis of RPEMP includes lymphoplasmacytic lymphoma and immunoblastic lymphoma. Many cases of gastrointestinal plasmacytoma were misdiagnosed as low grade B-cell lymphoma with plasma cell differentiation.[6] Immunohistochemistry using CD45 and CD20 negative stains is specific for plasma cells. [7]

There are no clear guidelines for treatment of RPEMP due to its variable presentation and rarity. All 3 modalities, surgery, radiotherapy and chemotherapy have been tried with the variable results. Radiotherapy has been effective in achieving long term local control.[8] However it is associated with high morbidity particularly when used for large retroperitoneal tumours. Tanaka et al [9] have tried chemotherapy before and after surgical resection (which was incomplete). Their patient progressed later and died 33 months after initial treatment. Chen et al [10] has reported in a case of retroperitoneal extramedullary plasmacytoma with obstructive jaundice who showed complete response to treatment with sequential radiotherapy and chemotherapy.

We first treated our patient by doing subtotal excision of the mass (as complete excision was not possible as it was adherent with great vessels). Then we referred the patient to the department of Oncology in DMCH for radiotherapy.

In summary, we have presented a rare case of primary extramedullary plasmacytoma in the retroperitoneum. Extramedullary Plasmacytoma should be kept in mind as a differential diagnosis of abdominal haematolymphoid malignancy. Surgical resection is also an important treatment option in such patients .[11]

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