

Retro-peritoneal plasmacytoma: A case report and review of literature

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Abstract

Solitary Extramedullary Plasmacytoma (EMP) is an uncommon neoplasm. When diagnosed, head and neck region is its most likely location. Rarely, it may occur in the retro-peritoneum. We report a 44-year old man with solitary extramedullary plasmacytoma in the retro peritoneum (RPEMP). The patient did not show response to three cycles of VAD chemotherapy. Thereafter Surgical excision of the mass was performed successfully. This is probably the first case report from Indian subcontinent.

Key Words: Extramedullary, Plasmacytoma, Retro peritoneum

Solitary Extramedullary Plasmacytoma (EMP) is an uncommon neoplasm. When diagnosed, head and neck region is its most likely location. Rarely, it may occur in the retro-peritoneum. We report a 44-year old man with solitary extramedullary plasmacytoma in the retro peritoneum (RPEMP).

Case Report

A 44-year old man was referred to our hospital with history of effort intolerance, abdominal distension and pain for four months. He did not have history of fever, weight loss, bladder or bowel dysfunction. Physical examination revealed an irregular, firm, non-tender mass occupying almost whole of the abdomen. There was no pallor; icterus, lymphadenopathy and both testes were normal. Abdominal CT scan revealed a 17 cm x 13 cm lobulated mass extending from the level of third part of duodenum to hypo gastric region displacing and partially encasing the mesenteric vessels with inferior vena caval compression. The mass showed areas of calcification and necrosis. There was no retroperitoneal adenopathy. Fine needle aspiration cytology of the mass was suggestive of a plasma cell neoplasm.

Investigations revealed Hemoglobin 7.0 gms/dl,

Erythrocyte sedimentation rate 65-mm 1st hour, Total Leukocyte Count and Platelet Count were normal. Serum total protein, albumin, globulin were 8.5, 1.7 and 6.8 gm/dl respectively and Serum β_2 micro globulin was 4.8 mg /L. Renal function tests, hepatic enzymes, serum electrolytes, serum calcium and phosphorus were within normal limits. The Serum-Immunoelectrophoresis (SIEP) revealed M band whose concentration was 4.7 gms/dl. Serum IgG level was 9360 mgs/ dl (normal range 700 - 1500). Serum IgA and IgM levels were within normal range. Urinary Bence Jones proteins were negative. Skeletal survey was normal. Bone-marrow aspiration and biopsy were normal. A final diagnosis of solitary retroperitoneal extramedullary plasmacytoma (RPEMP) was made. Patient was started on chemotherapy with VAD (Vincristine 0.4 mg/day, Adriamycin 9.0 mg/m² /day continuous infusion x 4 days through a central catheter along with oral dexamethasone 40 mg/day x 4 days). At the end of 2 chemotherapy cycles, clinically abdominal mass was same while the CT scan showed a marginal reduction in size. Repeat SIEP showed increase in the M- band concentration to 7.0 gms/dl. One more cycle of VAD chemotherapy was given without any response. Thereafter Surgical excision of the mesenteric mass (with resection of small bowel, right hemi-colectomy

and ileo-colic anastomosis) was done. Postoperative recovery was uneventful.

Histopathology revealed an 18x 16 x 10 cms encapsulated tumor with adherent bowel loops. Microscopic examination confirmed plasmacytoma while the bowel mucosa was normal. Three weeks after surgery, SIEP showed reduction of M-band to 2.9-gms/dl and Ultrasonography of the abdomen showed no residual tumor. On follow up, M-band became undetectable. The patient is under observation 16 months post surgery and is asymptomatic.

Discussion

Extramedullary plasmacytoma (EMP) constitutes 4% of plasma cell tumors. It is defined as a solitary tumor composed of monoclonal proliferation of cells with plasmacytic differentiation in an extramedullary site.¹ 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 heads and neck region. Other documented sites include gastrointestinal tract, CNS, urinary tract, thyroid, breast, testis, parotid gland and lymph nodes. Solitary EMP in the retro peritoneum is very rare. Marks² reported bilateral renal vein occlusion with renal failure and fatal hemorrhage due to tumor erosion and vena caval perforation in a case of RPEMP. Kobayashi et al³ has reported tumor thrombus within the renal vein in a 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.⁴ Immunohistochemistry using CD 45 and CD 20 negative stains is specific for plasma cells.⁵

There are no clear guidelines for treatment of RPEMP due to its variable presentation and rarity. All 3 modalities chemotherapy, radiotherapy and surgery have been tried with variable results. Radiotherapy has been effective in achieving long-term local control.⁶ However it is associated with high morbidity particularly when

used for large retroperitoneal tumors. Tanaka et al⁷ have tried chemotherapy before and after surgical resection (which was incomplete). Their patient progressed later and died 33 months after initial treatment. Chen et al⁸ 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 with chemotherapy to which he did not respond well. Radiotherapy was not feasible because of the large bulky abdominal tumor. However complete surgical resection was done successfully. It also resulted in complete disappearance of M protein and long-term complete remission.

In summary, we have presented a rare case of primary EMP in the retro peritoneum. EMP should be kept in mind as differential diagnosis of abdominal hemolymphoid malignancy. Surgical resection should also be an important treatment option in such patients.⁹

References

1. Weichhold W, Labouyrie E, Merlio JP, Masson B, DeMascarel A: Primary Extramedullary plasmacytoma of the liver. *Am J Surg Pathol* 1995;19:1197-202.
2. Marks ES, Lee KM. "Acute renal failure secondary to vascular occlusion by a retro peritoneal plasmacytoma" *Cancer* 1984;53:1228-9.
3. Kobayashi H, Itoh, T, Murata R, Tanabe M. Primary retro peritoneal plasmacytoma with tumor thrombus within the renal vein. *J Urol* 1992;147:452-4.
4. Henry K, Farrer- Brown G: Primary lymphoma of the gastrointestinal tract. 1. Plasma cell tumors. *Histopathology* 1997;1:53-76.
5. Strickler JG, Audeh MW, Copenhaver CM, and Warnke RA: Immunophenotypic differences between Plasmacytoma/ Multiple Myeloma and immunoblastic lymphoma. *Cancer* 1988;61:1782-6.
6. Shih IY, Dunn P, I eung WM, Chen WI, Wang PN: Localized Plasmacytoma in Taiwan: Comparison between extramedullary plasmacytoma and solitary plasmacytoma of bone. *Br J Cancer* 1995;71:128-33.
7. Tanaka Y, Samma S, Hayashi Y, Fujimoto K, Nagayoshi J, Yoneda T, et al. "A case of retroperitoneal extramedullary plasmacytoma" [Article in Japanese] *Hinyokika Kiyo* 1993;39:639-43.
8. Chen TC, Wu JH, Ng KF, Lien JM, Hung CF, "Solitary extramedullary plasmacytoma in the retroperitoneum" *Am J Hematol* 1998 Jul;58:235-8.
9. Molina Boix M, Ortega Gonzalez G, Perez Gracia A, Garcia Perez B, Munoz L, Bermudo J. "Thoracic and retroperitoneal dissemination of multiple myeloma 2 cases." *An Med Interna* 1989;6:315-7.