The Gulf Journal of Oncology

Indexed By PubMed and Medline Database

Issue 12, July 2012
ISSN No. 2078-2101

GFFCC Expanded International Cooperation

The Official Journal of the Gulf Federation For Cancer Control
# Table of Contents

## Original Studies
- **Penile Cancer in India: A Clinicoepidemiological Study**
  M. Pahwa, M. Girotra, A. Rautela, R. Abraham

- **Gastric Cancer: A Retrospective Analysis from AIIMS, New Delhi**
  R. Hadi, B.K. Mohanti, S. Pathy, G.K. Ralh, N.K. Shukla, S.V.S. Deo, A. Sharma, V. Raina

## Review Articles
- **Intensity Modulated Radiotherapy (IMRT) In Head and Neck Cancers – An Overview**
  C.M. Nutting

- **Adult T-Cell Leukemia/Lymphoma**
  K.I. Rasul, Z.A. Barwari

## Case Reports
- **Adrenocortical Tumors in Children: A Kuwait Experience**
  R. Mittal, D. G. Ramadan, N. M. Khalifa, S. O. Khalifa, Z. Mazidi, M. Zaki

- **Limb Sparing Surgery in Soft Tissue Sarcoma of Extremities: An Indian Perspective**
  R.V. Bhargavan, P. Kumar, K.C. Kothari

- **Mixed Germ Cell Tumor of Ovary and Clitoromegaly in Swyer’s Syndrome: A Case Report**
  S. Aminimoghaddam, B. Mokri, F. Mahmoodzadeh

- **Palmar Fasciitis and Arthritis Syndrome Associated With Metastatic Ovarian Cancer: A Paraneoplastic Syndrome**
  I.K. Nahar and M. S. Al-Rajhi

- **Trichilemmal Pilar Tumor of the Scalp: A Case Report**
  K. Al Saleh, H.S. Hooda, H. El-Wakiel, R. Safwat, A. Bedair, W. Eskaf

- **Carcinosarcoma of Renal Pelvis with Immunohistochemical Correlation**
  S.D. Deshmukh, V.L.Gaopande, D.P. Pande, G.S. Pathak, B.K. Kulkarni

- **5-Flourouracil Cardiotoxicity – An Elusive Cardiopathy: Case Report**
  G. M. Bhat, M. H. Mir, H. I. Showkat, B. Kasanna, F. Bagdadi, A. H. Sarmast, S. Quadri

- **An Unusual Variant of Prostatic Adenocarcinoma with Metastasis to Testis: A Case Report**
  K. R. Anita, T. Somananathan, A. Mathews, K. Jayasree

- **Mammary Fibromatosis in a Male Breast**
  N. Al-Saleh, T. Amir, I. N. Shaf

- **Primary Isolated Extramedullary Plasmacytoma of Mesentry: A Rare Case Report**
  R. Galhotra, K. Sagggar, K. Gupta, P. Singh

## Feature Article
- **Balsam Organization for rehabilitation and support for cancer patients and their families**

## Conference Highlights /Scientific Contribution
- **Conference Highlights – 1st Palliative Care Conference in Kuwait**

- **News Notes**

- **Advertisements**

- **Scientific events in the GCC and the Arab World for the 2nd Semester of 2012**
Primary Isolated Extramedullary Plasmacytoma of Mesentry: A Rare Case Report

R. Galhotra, K. Saggar, K. Gupta, P. Singh

Department of Radiodiagnosis, Dyanand Medical College & Hospital, Ludhiana, India

Abstract

Extramedullary plasmacytoma (EMP) is an uncommon entity that most commonly involves nasopharynx and upper repository tract. Involvement of GIT occurs in approximate 10% of cases. According to WHO plasma cell tumors have been classified into two main groups: Multiple myeloma and plasmacytoma. Plasmacytoma includes solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. EMP can be either primary without evidence of bone marrow involvement or may occur simultaneously with multiple myeloma representing extramedullary spread of the disease. It may occur in association with multiple myeloma and it may precede, accompany or follow the onset of multiple myeloma. Diagnosis of primary EMP requires the exclusion of associated multiple myeloma as shown by negative Bence Jones Proteins in urine, normal serum electrophoresis, normal bone marrow biopsy, normal skeletal survey and normal calcium levels. Here we present a case of 55-year male who came to Nephrology Department for urinary tract infection and pain abdomen. Patient was referred to Radiology for ultrasonography which revealed bilateral renal parenchymal disease with a well-defined mass in the mesentry which was further confirmed on computed tomography. Patient was surgically operated and diagnosis of primary EMP of mesentry was made on histopathological examination. Only three cases have been reported so far in the literature.

Keywords

Extramedullary plasmacytoma, mesentry, plasma cell dyscrasias

Introduction

B cell lymphocytes are derived from haematopoietic stem cells in the bone marrow. In response to foreign antigen B cells will mature and transform into plasma cells. When the transformation of stem cells to B cells is altered, it results to an abnormal or malignant plasma cells called plasmablast, developmentally early form of plasma cells. These cells can proliferate in bone marrow to form multiple myeloma or extramedullary in the soft tissue to form plasmacytoma (1).

Extramedullary plasmacytoma constitute 4% of plasma cells tumors. It is defined as a solitary tumor composed of monoclonal proliferation of cells with plasmocytic differentiation in an extramedullary site. It is classified either as primary EMP (when there is absence of coexisting multiple myeloma) or secondary EMP (when it is associated with multiple myeloma). EMP occurs most commonly in the head and neck region (>90%). Other documented sites are GIT, CNS, urinary tract, thyroid, breast, testis, parotid gland and lymph nodes (2).

Primary EMP is a rare form of disease accounting for 20% of cases with 7% only manifesting in GIT (1). Like local bone disease, EMP most commonly affects males in their early sixties. Clinical presentation and radiological appearance may vary according to the involved organ (3).

Here we present a case of isolated primary EMP of mesentry with patient presenting to the OPD with pain abdomen from last 4-6 months. On ultrasound and CT scan well defined mass lesion was seen in the mesentry. Patient was operated for the lesion and final diagnosis of mesenteric EMP was made on histopathological examination.
Case report

55-year old made came to the Nephrology Department for dysuria, frequency of micturition and pain abdomen from last 4-6 months. On general physical examination patient was afebrile, hypertensive from last 2 years. Chest and CVS was normal. Patient was referred to Radiology Department for ultrasonography.

Ultrasound was done using 3.5 Hz curved probe in supine position and patient was reported to have bilateral renal parenchymal disease and prostatomegaly grade I. Along with a well-defined hypoechoic mass approximately 11 x 9.6 cm was noted in the mesentry in paraumblical region which was reported as ? lymphnodal mass and patient was referred for CT scan.

CECT was done on Siemens 128 slice CT scanner. CT revealed a well-defined hypodense mildly enhancing mass lesion in the mesentry which measures 11.9 x 9.2 x 5.9 cm. No necrosis or calcification was seen. Fat plains with surrounding structure were maintained. The rest of the major organs were normal. Diagnosis was lymphnodal ? GIST. Patient was operated and histopathological examination was done.

Histopathology report - Sections from the mesenteric mass show sheets of plasma cells having exocentric nuclei and moderate amount of eosinophilic cytoplasm. Interspersed is seen large amount of eosinophilic material – amyloid which is surrounded by foreign body giant cells. Areas of haemorrhage with numerous vascular channels identified. Lymphoid follicles are seen at the periphery.

Impression - histology consistent with plasma cell dyscrasia–plasmacytoma of mesentry. Patient was further investigated for systemic disease. His calcium levels were 8 mg% (normal). Urine was negative for Bence Jones Proteins. Skeletal survey was normal. Serum electrophoresis was normal. Bone marrow biopsy showed normal maturity and morphology of all haemopoietic elements with a patchy prominence of plasma cells. So the final diagnosis was isolated primary EMP of mesentry.

![Fig. 1.a](image1a.png)
![Fig. 1.b](image1b.png)
![Fig. 1.c](image1c.png)
![Fig. 1.d](image1d.png)

Figures 1 a, b, c & d: CECT (axial, sagital, coronal sections and bone window) showing well defined mildly enhancing hypodense mass lesion in the mesentry with well maintained surrounding fat plains measuring 11.9 x 9.2 x 5.9 cm
EMPs are rare tumors. They may be solitary or may occur with multiple myeloma. In the later cases, it may precede, accompany or follow the onset of multiple myeloma. Diagnosis of EMP requires the exclusion of associated multiple myeloma as shown by negative Bence Jones Proteins in urine, normal electrophoresis and normal bone marrow biopsy (4). GIT involvement by EMP is rare and is seen in less than 5% of all EMPs (5). Mesentry is an unusual site of EMP (6) with only 3 cases reported so far in the literature. Histologically these lesions are characterized by monoclonal proliferation of plasma cells. Monoclonal proliferation can be confirmed by demonstrating single type of paraprotein in tumor cells by immunoperoxidase staining. The demonstration of single type of paraprotein in tumor cells is considered to be the confirmed evidence for making the diagnosis of plasmacytoma (4).

These tumors are highly radiosensitive however, if the neoplasm can be completely surgically resected, the long term results have been reported to be equal to those involving radiation alone. At least 70% of these patients remain disease free after 10 years and fewer than 30% show signs of recurrence or develop a further failure in form of multiple myeloma (3).

The prognosis of solitary plasmacytoma is dependent on the presence of disseminated disease. Multiple myeloma is a systemic disease having no potential for complete cure. The prognosis of plasmacytoma with multiple myeloma is very poor (10). Long term follow

Patient was discharged after 7 days and was kept for the follow up after every six months.

Discussion

According to WHO plasma cell tumors are of two types: multiple myeloma and plasmacytoma. Plasmacytoma includes solitary plasmacytoma of bone or solitary EMP (4). A plasmacytoma is a mass composed of monoclonal neoplastic plasma cells. It can exist isolated as a solitary plasmacytoma or associated with multiple myeloma. In a recent reported series almost half of patients with GIT involvement by EMP were initially asymptomatic. These lesions were found incidentally by imaging or autopsy. Most of these lesions mimic carcinoma or lymphoma and pose difficult diagnostic challenges (7).

Most frequent sites are superior respiratory airway, pleura, lung, lymphnode, skin, subcutaneous and soft tissues, testicles and liver (8). Despite many recent progressions in diagnostic modalities, occasionally the initial manifestation of the disease may be misleading. It is therefore helpful to consider the uncommon presentation of prevalent disease. Plasma cell dyscrasias is one of the most well-known haematological malignancies. Clonal expansion of plasma cells results in diverse clinical findings such as renal failure, lytic bone lesion, anaemia, hyperviscosity syndrome and so forth. However, this disease entity rarely presents with abdominal mass due to mesenteric amyloid deposition (9). Males are commonly affected than females. Patients in the age group of 35-85 years are usually affected (4).

According to WHO plasma cell tumors are of two types: multiple myeloma and plasmacytoma. Plasmacytoma includes solitary plasmacytoma of bone or solitary EMP (4). A plasmacytoma is a mass composed of monoclonal neoplastic plasma cells. It can exist isolated as a solitary plasmacytoma or associated with multiple myeloma. In a recent reported series almost half of patients with GIT involvement by EMP were initially asymptomatic. These lesions were found incidentally by imaging or autopsy. Most of these lesions mimic carcinoma or lymphoma and pose difficult diagnostic challenges (7).

Most frequent sites are superior respiratory airway, pleura, lung, lymphnode, skin, subcutaneous and soft tissues, testicles and liver (8). Despite many recent progressions in diagnostic modalities, occasionally the initial manifestation of the disease may be misleading. It is therefore helpful to consider the uncommon presentation of prevalent disease. Plasma cell dyscrasias is one of the most well-known haematological malignancies. Clonal expansion of plasma cells results in diverse clinical findings such as renal failure, lytic bone lesion, anaemia, hyperviscosity syndrome and so forth. However, this disease entity rarely presents with abdominal mass due to mesenteric amyloid deposition (9). Males are commonly affected than females. Patients in the age group of 35-85 years are usually affected (4).

Histologically these lesions are characterized by monoclonal proliferation of plasma cells. Monoclonal proliferation can be confirmed by demonstrating single type of paraprotein in tumor cells by immunoperoxidase staining. The demonstration of single type of paraprotein in tumor cells is considered to be the confirmed evidence for making the diagnosis of plasmacytoma (4).

These tumors are highly radiosensitive however, if the neoplasm can be completely surgically resected, the long term results have been reported to be equal to those involving radiation alone. At least 70% of these patients remain disease free after 10 years and fewer than 30% show signs of recurrence or develop a further failure in form of multiple myeloma (3).

The prognosis of solitary plasmacytoma is dependent on the presence of disseminated disease. Multiple myeloma is a systemic disease having no potential for complete cure. The prognosis of plasmacytoma with multiple myeloma is very poor (10). Long term follow
up is very important as systematic disease may present later on. In the largest series of EMP, 22 patients (32%) developed multiple myeloma after a follow up of 1.8 years. In 22 patients from MD Anderson Cancer Center the median survival was 9.5 years and 56% of patients were free from systemic disease at 5 years. Prognosis of solitary EMP is generally good (4).

**Conclusion**

EMP of mesentry is a very rare entity with only 3 cases reported in the literature. It usually presents as mesenteric mass and may mimic lymphoma, carcinoma or any inflammatory pathology. Although cases of mesenteric mass are rare, treating physician, radiologist and pathologist should keep the possibility of mesenteric plasmacytoma in the differential diagnosis and patient should be thoroughly followed up for the systemic progression of the disease.

**References**


9. Asadi M. Unusual presentation of more common disease/injury: Mesenteric amyloid deposition as the initial presentation of multiple myeloma BMJ Case Reports 2011; doi:10.1136/bcr.05.2010.2977