Case Presentation

Husein Husein

Emory University Hospital Division of Nuclear Medicine and Molecular Imaging, Department of Radiology and Imaging Sciences

Introduction

62 years old female with a PMH of seborrheic dermatitis, panniculitis, smoking and arthritis presented initially for evaluation of lower back pain and a slowly increasing right buttock swelling, recently becoming painful.

ROS: Unintentional weight loss over the past several years, chronic cough, subcutaneous nodule on the left side of the abdominal wall and diffuse myalgia.

Medications: Guaifenesin, hydrocortisone cream, ammonium lactate lotion, menthol/m-salicylate cream.

Labs

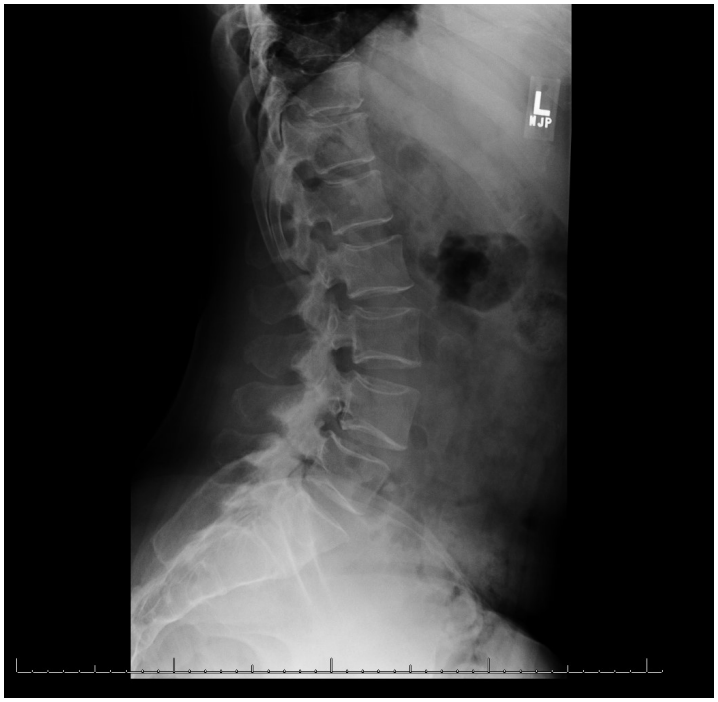
WBC	RBC	HGB	HCT	MCV	MCH	MCHC	PLT	RDW
6.2	3.78	12.1	34.7	91.8	32	34.9	281	15.1

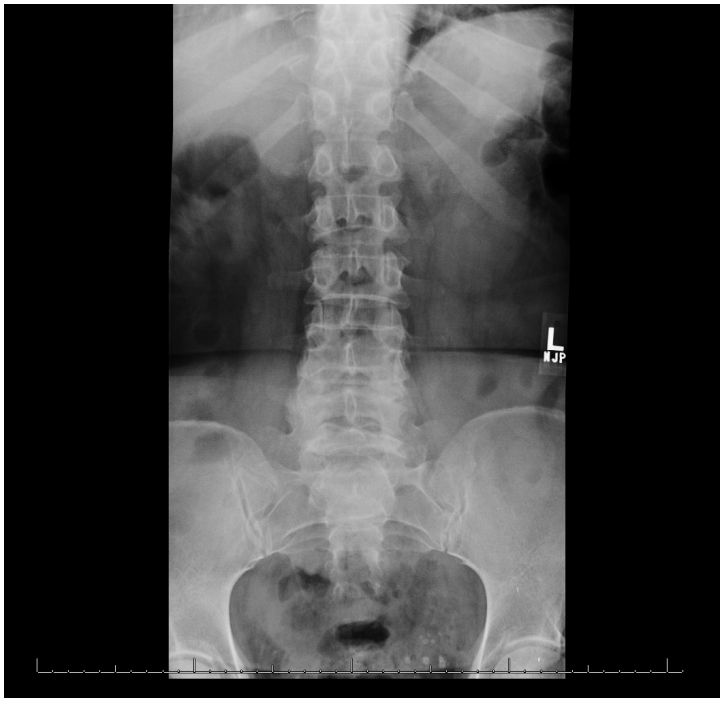
Cr	BUN	Glu	NA	K	Cl	CO2	CA	Prote	Alb	Globu
0.77	12	79	139	4.1	107	26	8.6	6.7	3.4	3.3

SERUM PROTEIN ELECROPHORESIS (SPEP)
Faint restricted band (M-spike) migrating in the gamma globulin region. Monoclonal IgG kappa present.

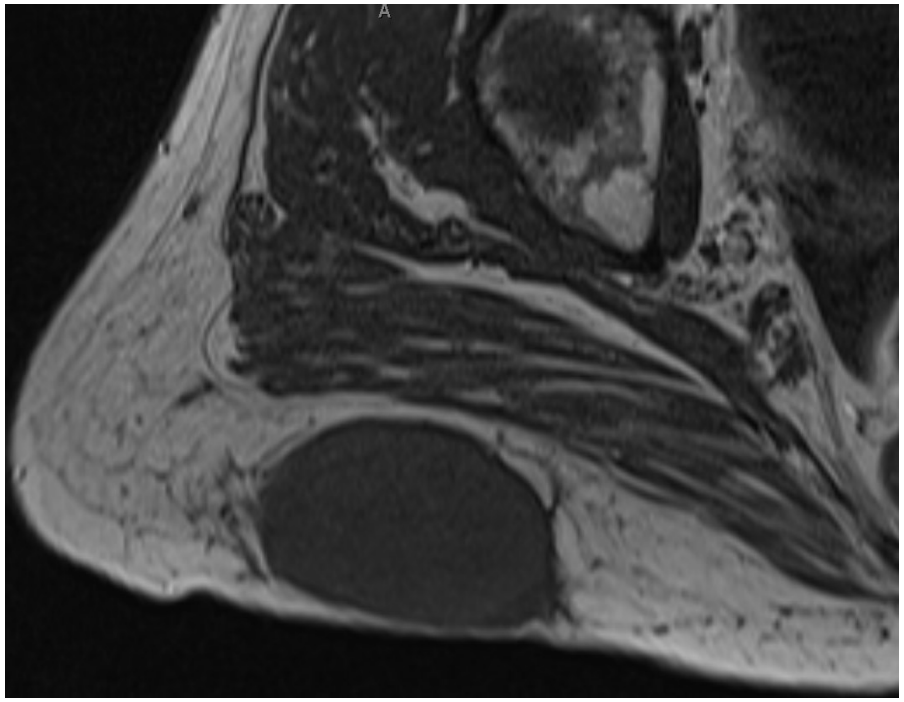
FREE KAPPA, SERUM	FREE LAMBDA, SERUM	FREE K/L RATIO (Q)
160.4H mg/L	35.6 mg/L	4.51 H

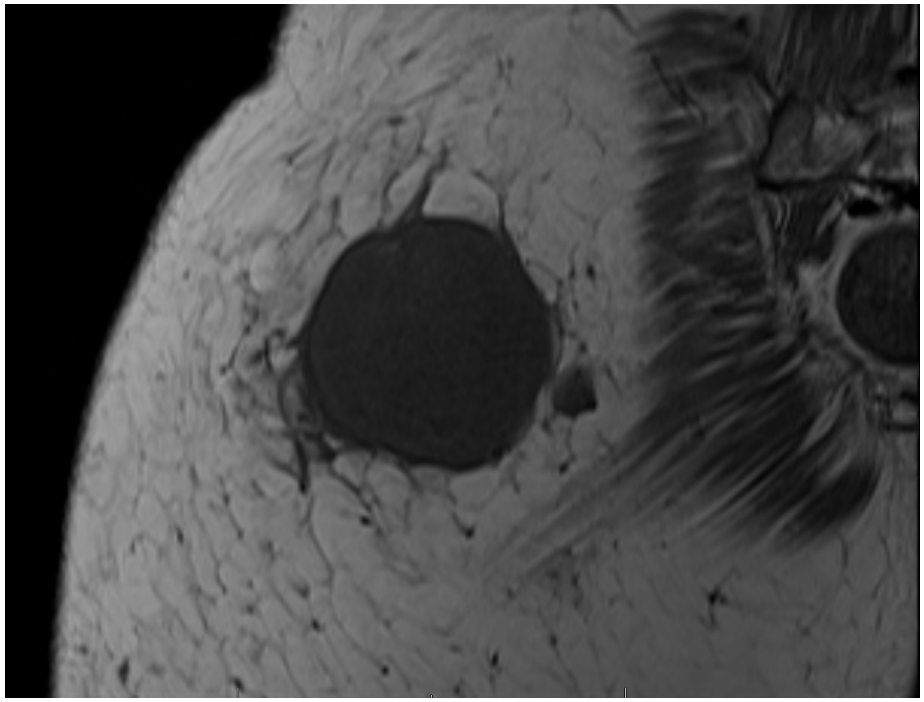
Lumbar spine x-ray, 10/02/2018





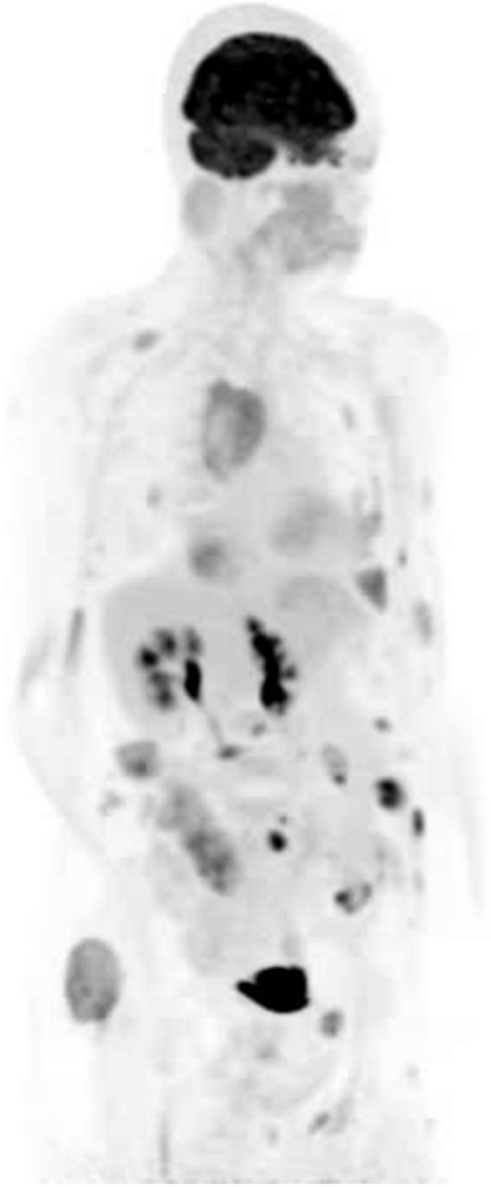
MRI of the pelvis 10/03/2018



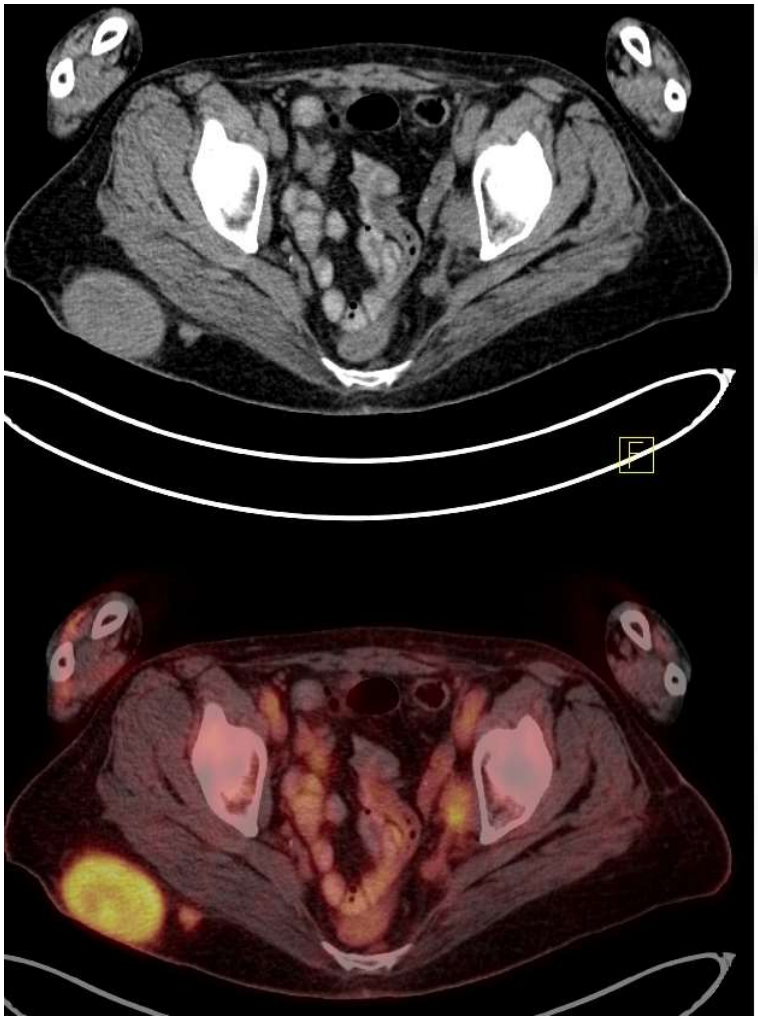


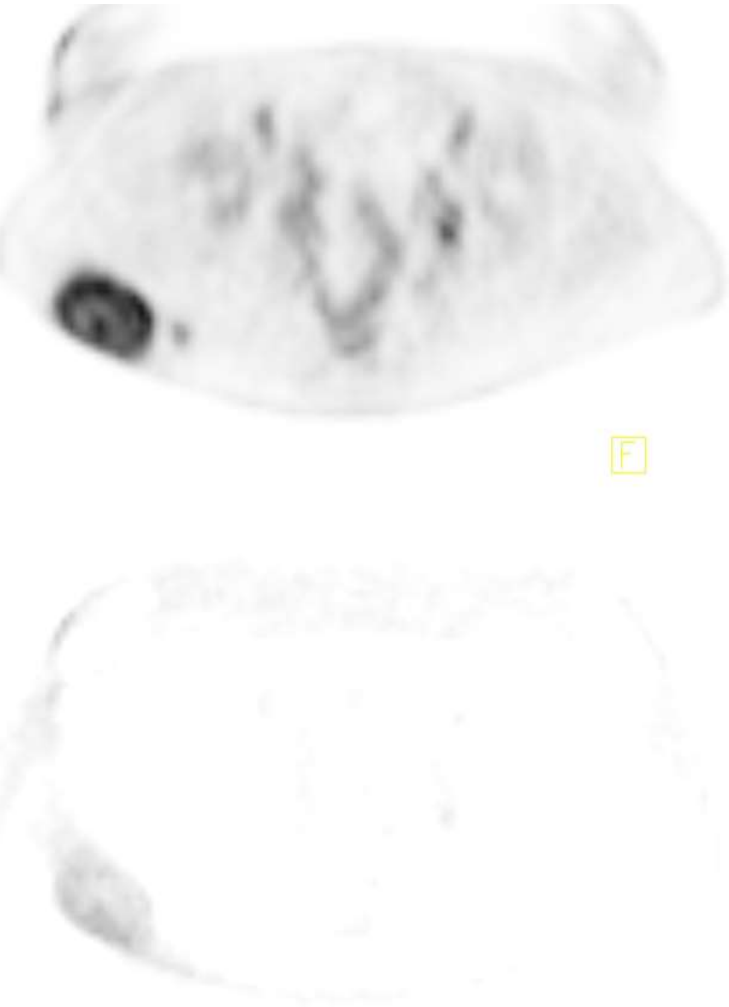
PET-CT

10/17/2018

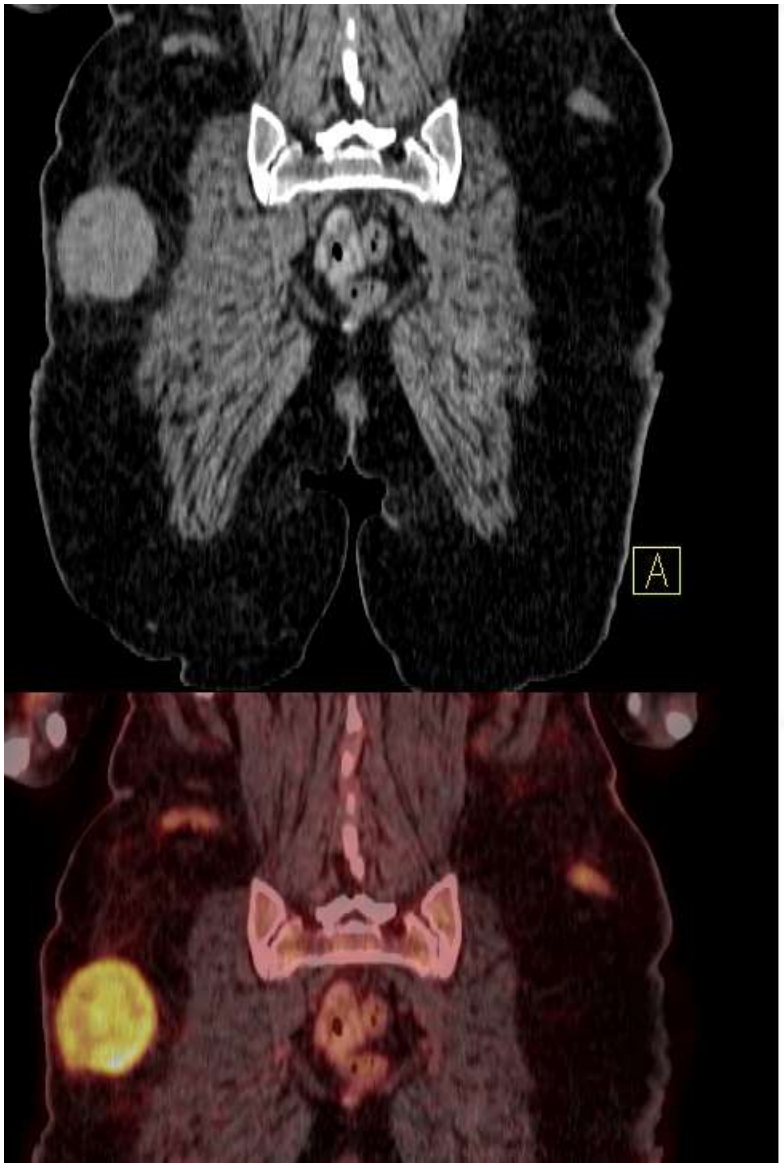


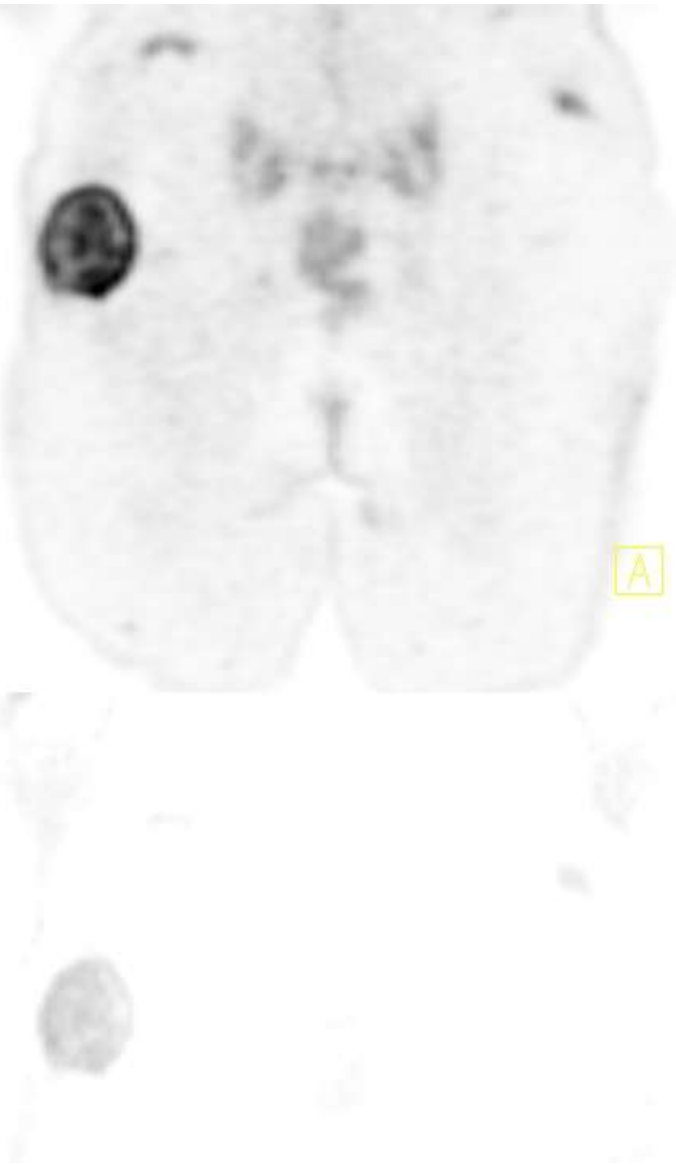
PET-CT
10/17/2018



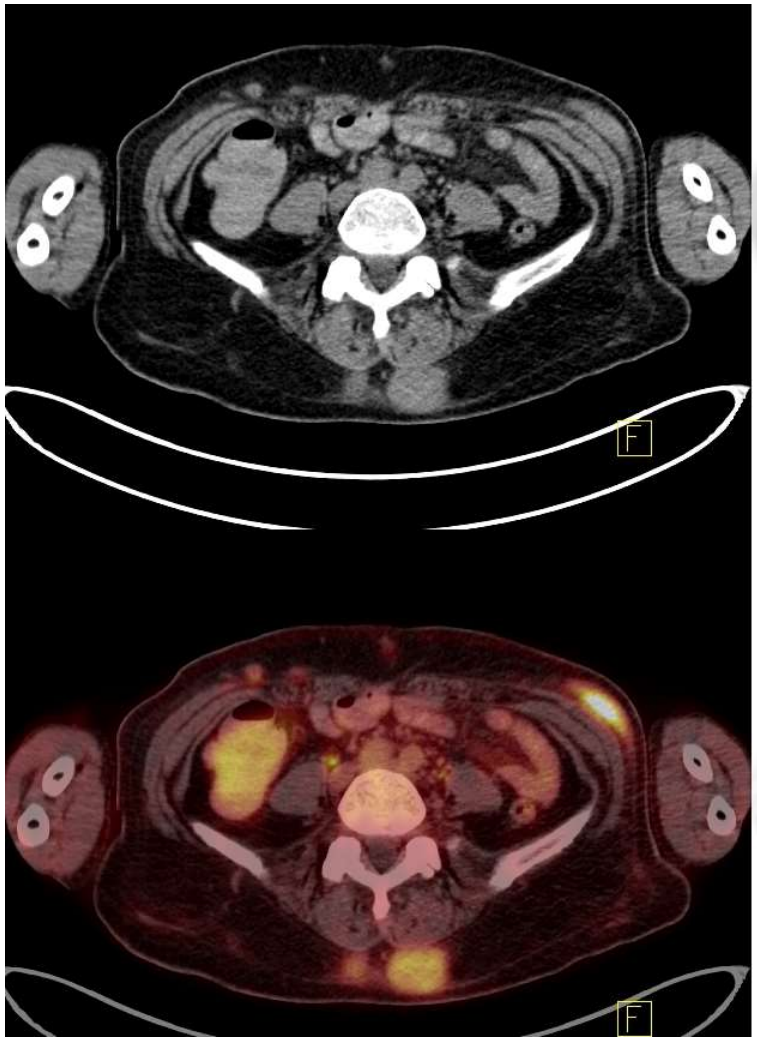


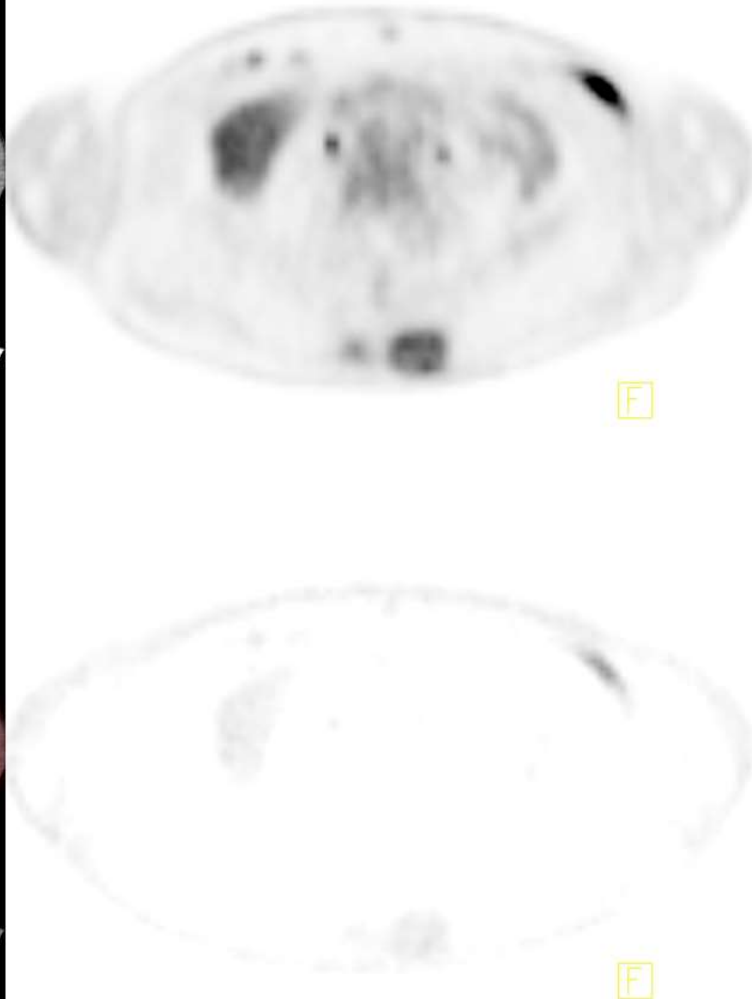
PET-CT
10/17/2018



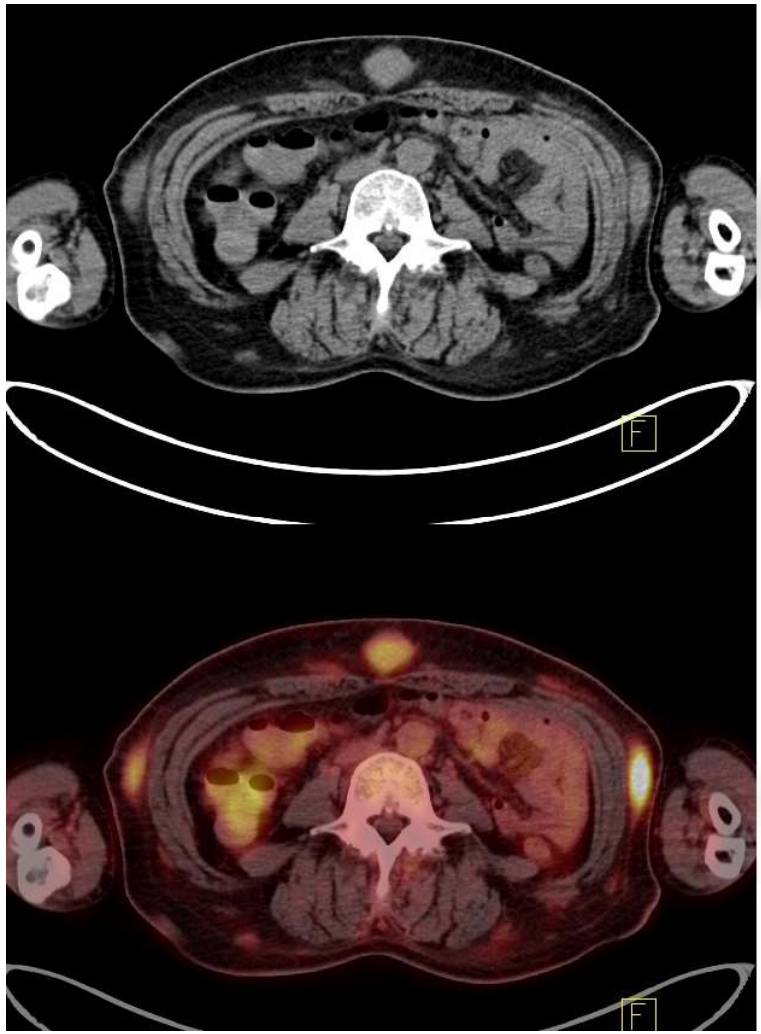


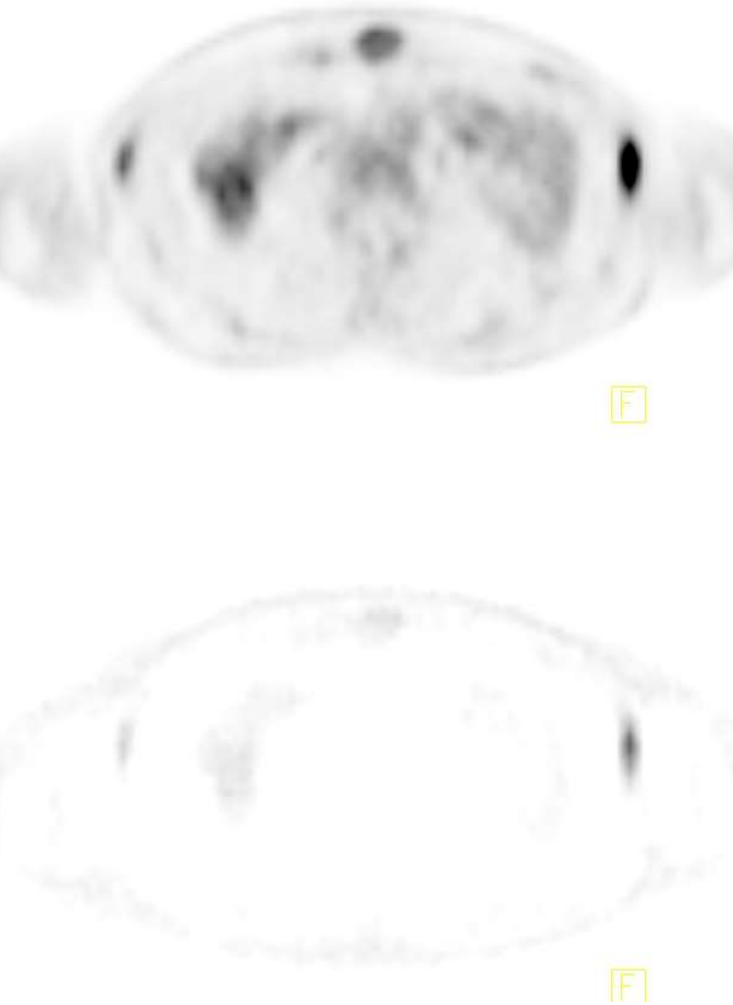
PET-CT
10/17/2018



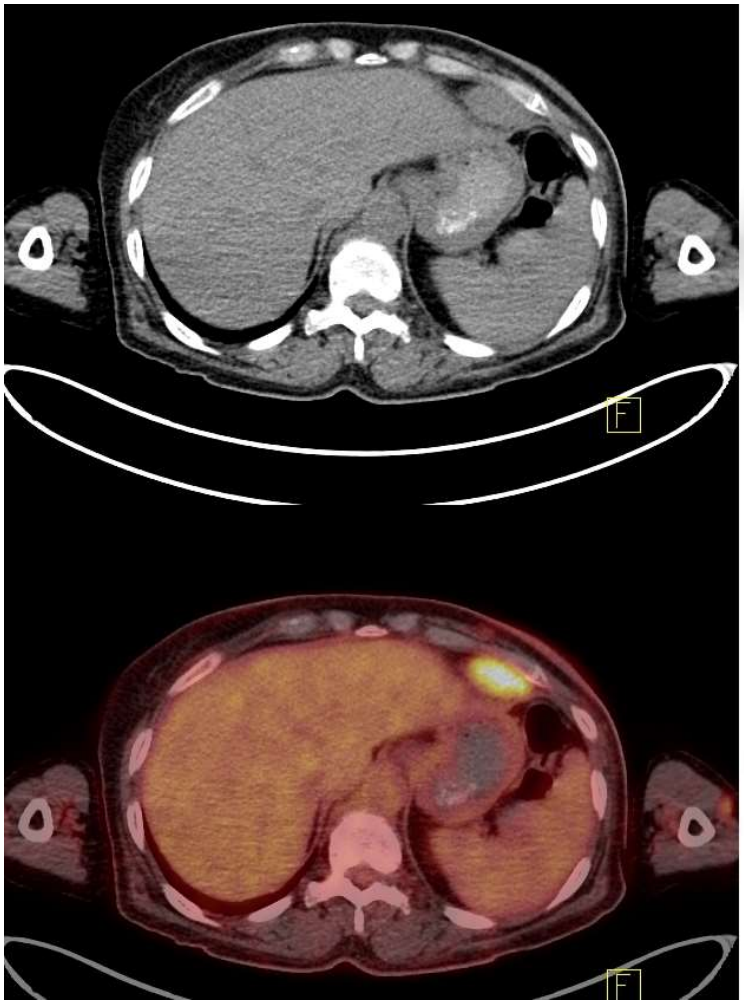


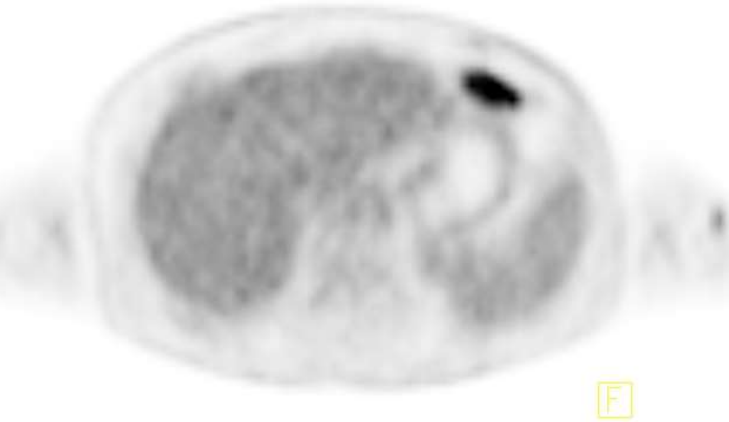
PET-CT
10/17/2018



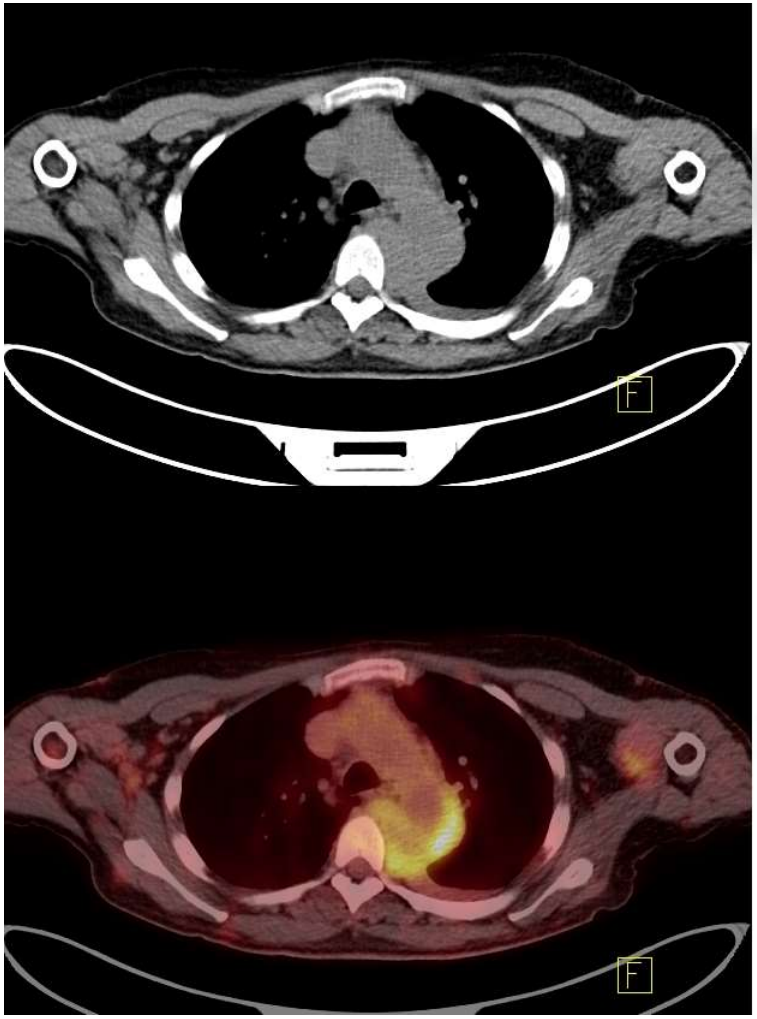


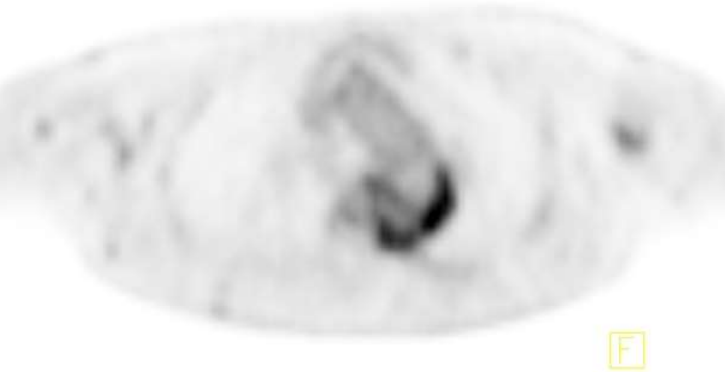
PET-CT
10/17/2018





PET-CT
10/17/2018






What is the differential diagnosis?

---- SURGICAL PATHOLOGY ----

MEDICAL RECORD | FLOW CYTOMETRY (AP)

PATHOLOGY REPORT Accession No. FL 18 2990

Submitted by: Date obtained: Dec 17, 2018 00:00

Specimen (Received Dec 17, 2018):
bone marrow aspirate

BRIEF CLINICAL HISTORY:

PREOPERATIVE DIAGNOSIS:

OPERATIVE FINDINGS:

POSTOPERATIVE DIAGNOSIS:

PATHOLOGY REPORT Accession No. FL 18 2990

Specimen: BONE MARROW. FLO 18 2990
Specimen Collection Date: Dec 17, 2018@10:45

FLOW CYTOMETRY INTERPRETATION/SUMMARY:

Flow cytometric immunophenotyping of this bone marrow aspirate specimen fails to identify any definitive immunophenotypically aberrant hematolymphoid populations.

Correlation with clinical/radiographic and additional laboratory data - including, but not limited to routine chemistry, protein electrophoresis, immunofixation, serum free light chain, marrow morphology, and cytogenetic study findings (reported separately) - is appropriate.

PATHOLOGY REPORT

Rapid on site preliminary evaluation, anterior abdominal wall, TP#1:
-Atypical plasmacytoid cells are present, additional cores requested for ancillary studies

Gross description:
The specimen is received in two parts.

Part # 1. The specimen is received in formalin labeled with patient name, social security number and further designated on the container and the requisition as "anterior abdominal wall, U/S guided FN BX ". The specimen consists of two needle core biopsies measuring 1.2 cm in greatest length and 0.1 cm in diameter. Specimen is wrapped in lens paper and entirely submitted in cassette 1A.

Part # 2. The specimen is received in formalin labeled with patient name, social security number and further designated on the container and the requisition as "left thigh, U/S guided FN BX ". The specimen consists of two needle core biopsies measuring 1.5 cm in greatest length and 0.1 cm in diameter. Specimen is wrapped in lens paper and entirely submitted in cassette 2A.

DIAGNOSIS

1) Anterior abdominal wall (fine needle biopsy):

- Kappa predominant plasma cell infiltrate consistent with a plasmacytic neoplasm

- See comment

2) Left thigh (fine needle biopsy):

- Kappa predominant plasma cell infiltrate consistent with a plasmacytic neoplasm

- See comment

Comment: The biopsies show similar features.

Corresponding flow cytometric immunophenotyping for the abdominal wall biopsy (FLO-19-205) is precluded because of insufficient quantity of B cells present in the analysis on a limited antigen panel. Flow cytometric immunophenotyping for the left thigh specimen (FLO-19-204), using a limited antigen panel because of low overall specimen cellularity, fails to identify any clonal B cell populations.

Previous anatomic pathology findings of a "plasmacytic neoplasm consistent with plasmacytoma or multiple myeloma within dense fibrous tissue" involving right buttock soft tissue (see SP18-7440 Mayo Medical Laboratories consultation report in VISTA imaging) and radiographic/PET studies showing multiple FDG avid masses above and below the diaphragm are noted. Taken together, the findings of multiple plasmacytomas would be compatible with a diagnosis of multiple myeloma. Although no diagnostic accompanying neoplastic lymphocytic component is observed by flow cytometry and lymphocytes are not prominent by IHC, diagnostic considerations may also include a low grade B cell lymphoma - such as a marginal zone lymphoma (MZL) - with marked plasma cell differentiation (as can be seen in cutaneous MZL, for example) and distinction between MZL and plasmacytoma may not be possible by morphological assessment alone.

Multiple Solitary Extramedullary Plasmacytomas

- **Plasmacytoma:** Rare plasma cell dyscrasia presenting as tumors growing within the soft tissue or within the axial skeleton.
- Characterized by the neoplastic proliferation of a single clone of plasma cells, typically producing a monoclonal immunoglobulin.
- The International Myeloma Working Group lists three types:
  - Solitary plasmacytoma of bone (SPB)
  - Extramedullary plasmacytoma (EP)
  - Multiple plasmacytomas that are either primary or recurrent.

British Journal of Haematology, 2003, **121**, 749–757

Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: a report of the International Myeloma Working Group

THE INTERNATIONAL MYELOMA WORKING GROUP*

Received 2 September 2002; accepted for publication 27 November 2002

Summary. The monoclonal gammopathies are a group of disorders associated with monoclonal proliferation of plasma cells. The characterization of specific entities is an area of difficulty in clinical practice. The International Myeloma Working Group has reviewed the criteria for diagnosis and classification with the aim of producing simple, easily used definitions based on routinely available investigations. In monoclonal gammopathy of undetermined significance (MGUS) or monoclonal gammopathy, unattributed/unassociated (MG[u]), the monoclonal protein is < 30 g/l and the bone marrow clonal cells < 10% with no evidence of multiple myeloma, other B-cell proliferative disorders or amyloidosis. In asymptomatic (smouldering) myeloma the M-protein is ≥ 30 g/l and/or bone marrow clonal cells ≥ 10% but no **r**elated **o**rgan or **t**issue **i**mpairment (ROTI)(end-organ damage), which is typically manifested by increased **c**alcium, **r**enal insufficiency, **a**naemia, or **b**one lesions (CRAB) attributed to the plasma cell proliferative process. Symptomatic myeloma requires evidence of ROTI. Non-secretory myeloma is characterized by the absence of an M-protein in the serum and urine, bone marrow plasmacytosis and ROTI. Solitary plasmacytoma of bone, extramedullary plasmacytoma and multiple solitary plasmacytomas (± recurrent) are also defined as distinct entities. The use of these criteria will facilitate comparison of therapeutic trial data. Evaluation of currently available prognostic factors may allow better definition of prognosis in multiple myeloma.

Keywords: classification, monoclonal, gammopathies, multiple myeloma.

Signs and symptoms

- Back pain
- Rhinorrhea, epistaxis and nasal obstruction (~ 85% of extramedullary plasmacytoma presents within the upper respiratory tract mucosa).
- Palpable mass (Extramedullary plasmacytomas may also occur in virtually any organ including the gastrointestinal tract, central nervous system, urinary bladder, thyroid, breasts, testes, parotid gland or lymph nodes).

Diagnosis

- Lacks increased blood calcium, decreased kidney function, too few red blood cells in the bloodstream, and multiple bone lesions.
- Serum protein electrophoresis: monoclonal spike.
- Skeletal surveys: no other primary tumors within the axial skeleton.
- MRI can be used to assess tumor status and may be advantageous in detecting primary tumors that are not detected by plain film radiography.
- PET-CT is beneficial in detecting extramedullary tumors in individuals diagnosed with SPB.

Prognosis

- Most cases of SPB progress to multiple myeloma within 2–4 years of diagnosis, but the overall median survival for SPB is 7–12 years.
- 15% of SPB and 50–65% of extramedullary plasmacytoma are disease free after 10 years.
- 30–50% of extramedullary plasmacytoma cases progress to multiple myeloma.

Treatment

- Multiple plasmacytomas may be treated by tumoricidal radiation if there is no evidence of multiple myeloma.
- Large numbers of solitary plasmacytomas or recurrent lesions at short intervals are an indication for systemic therapy such as autologous stem cell transplantation.

References

- International Myeloma Working Group (June 2003). "Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: a report of the International Myeloma Working Group". *Br. J. Haematol*. 121 (5): 749–57.

- Soutar R, Lucraft H, Jackson G, et al. (March 2004). "Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma". *Br. J. Haematol*. 124 (6): 717–2.

- Weber DM (2005). "Solitary bone and extramedullary plasmacytoma". *Hematology Am Soc Hematol Educ Program*: 373–6.

- Dimopoulos M, Terpos E, Comenzo RL, et al. (September 2009). "International myeloma working group consensus statement and guidelines regarding the current role of imaging techniques in the diagnosis and monitoring of multiple Myeloma". *Leukemia*. 23 (9): 1545–56.