Case Presentation

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Introduction

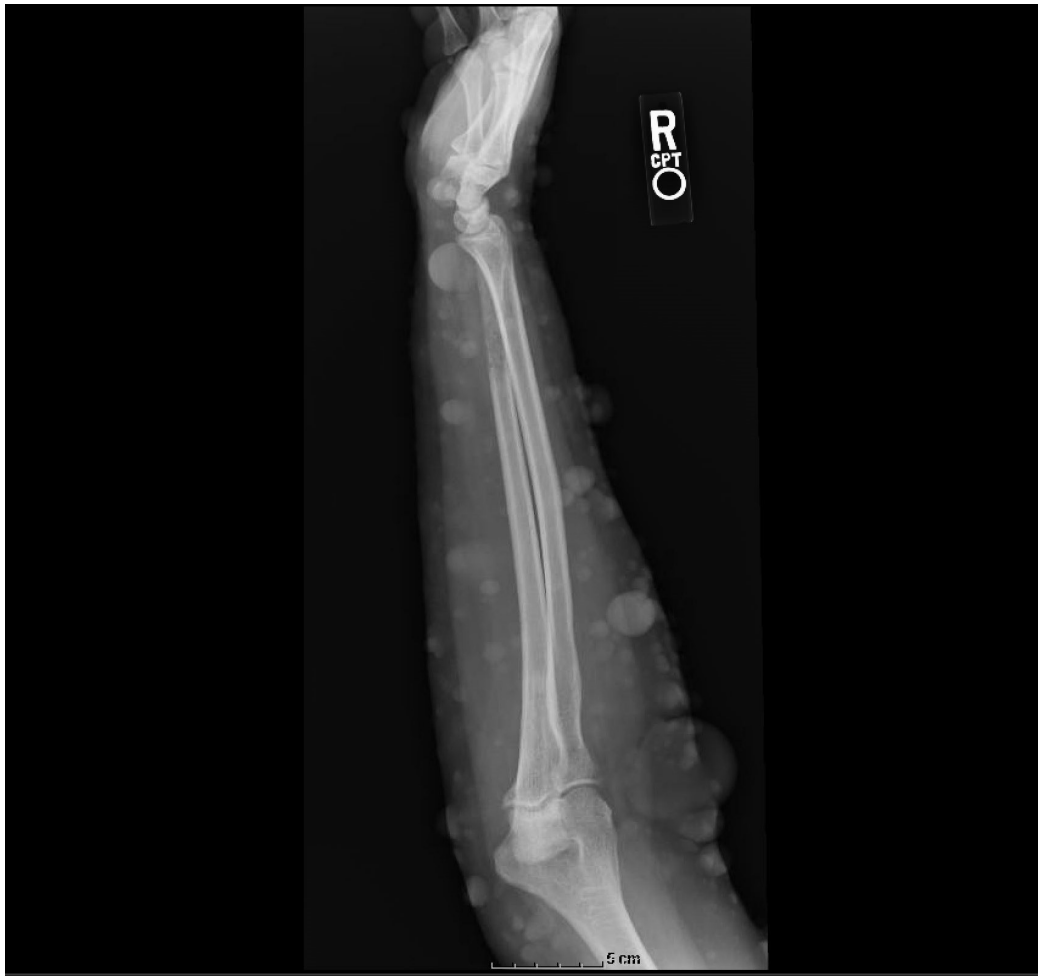
46 years old female with a PMH of NF1, HTN, anxiety and depression was seen by Orthopedic surgery in 09/14/2018 with chief complaint of right arm pain.

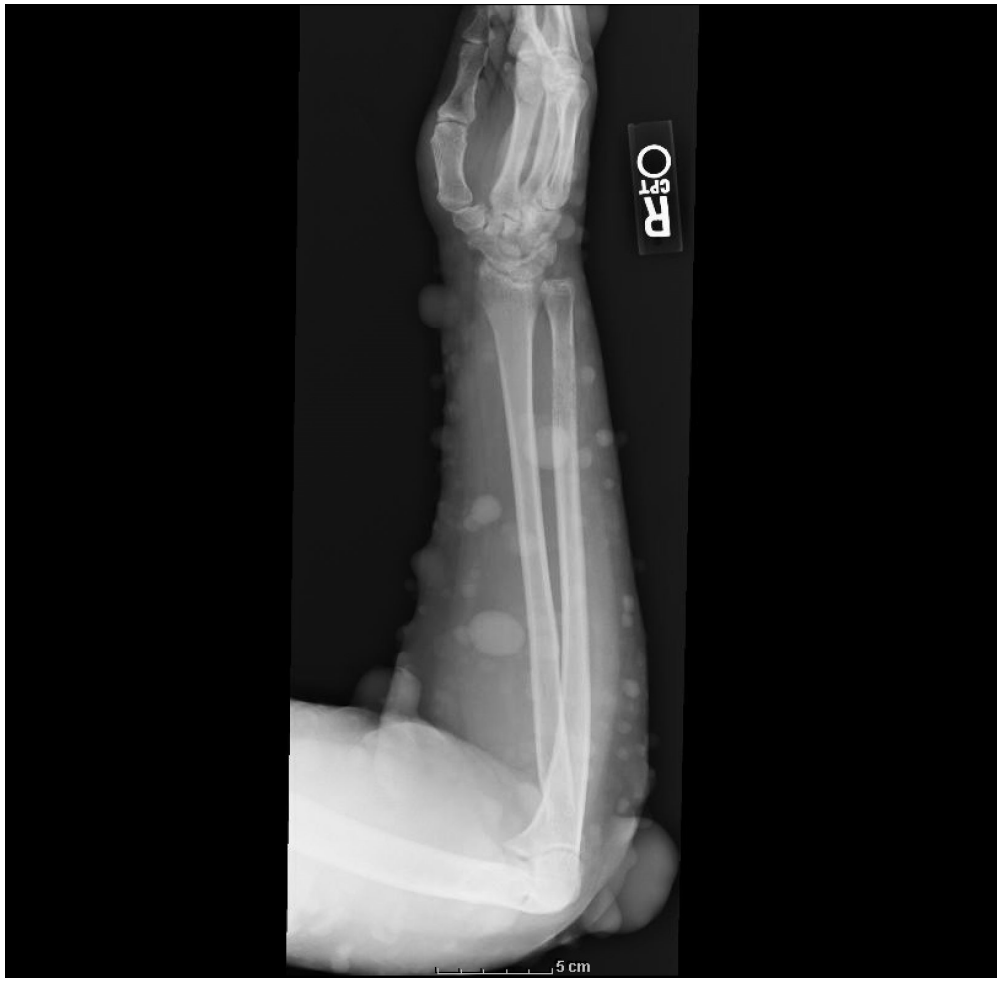
The arm pain has been gradually increasing for few months, associated with arm swelling and weakness, not responding to topical ice or pain medication. She was given antibiotics by the PCP.

ROS: Weight loss, cough, neck discomfort, fatigue, limb paresthesia and right facial partial paralysis.

Medications: Gabapentin, Duloxetine and Tramadol.

X-Ray of the right forearm, 09/14/2018






What is the differential diagnosis?

Surgical Pathology Report

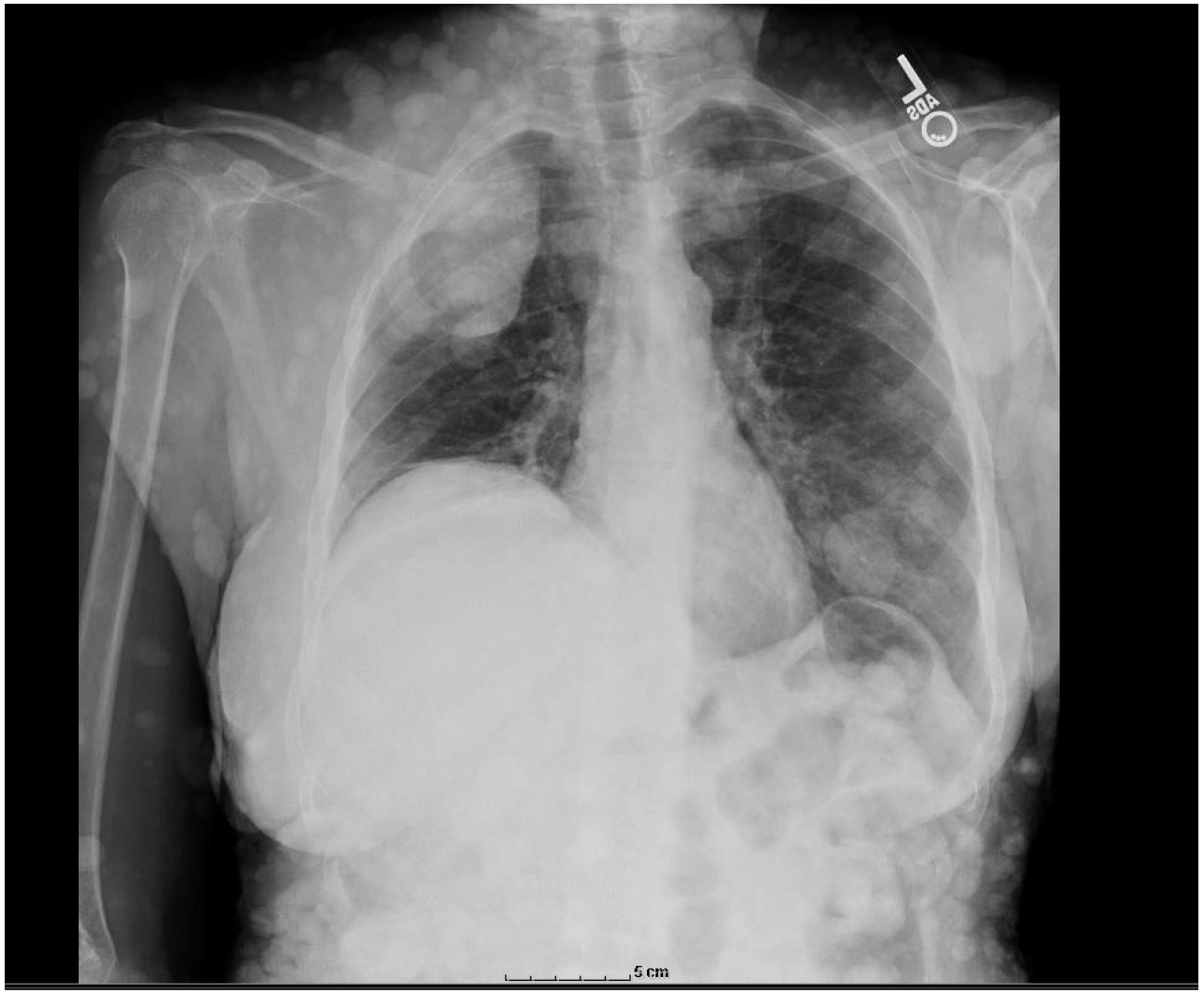
Pathologic Diagnosis

A. SOFT TISSUE, RIGHT ULNA, BIOPSY:
- ANGIOSARCOMA.

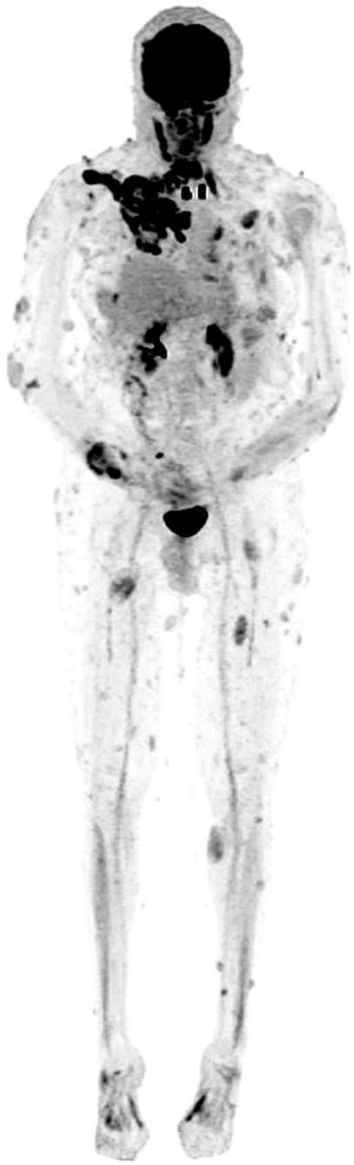
B. BONE, RIGHT ULNA, BIOPSY:
- ANGIOSARCOMA.

C. SOFT TISSUE, RIGHT ULNA, BIOPSY:
- ANGIOSARCOMA.
- IMMUNOSTAINS ARE POSITIVE FOR CD31 AND ERG, NEGATIVE FOR S100, CD45, CD3, CD20, DESMIN, AND AE1/AE3.

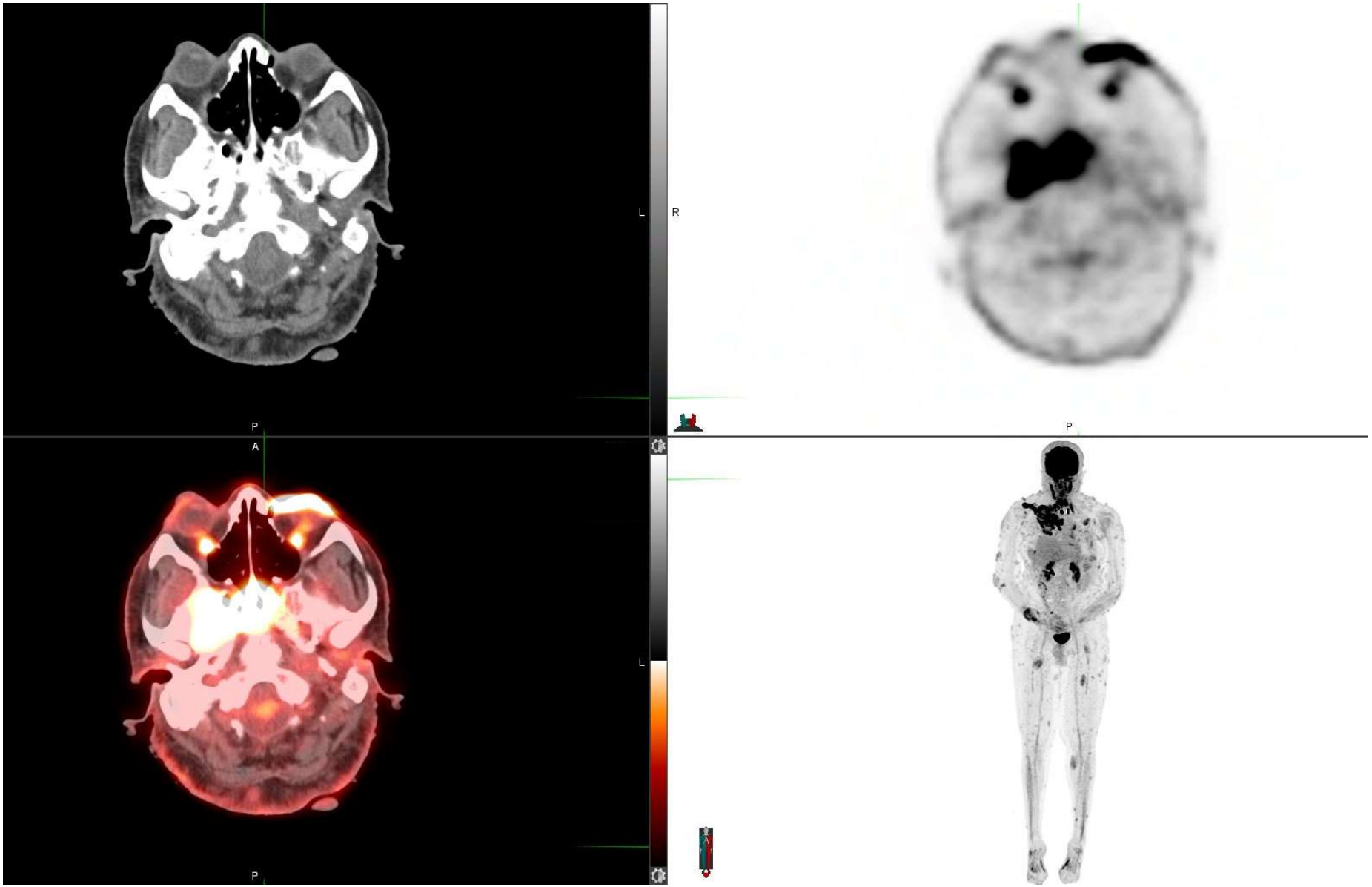
Chest x-ray, 09/21/2018



PET/CT



10/26/2018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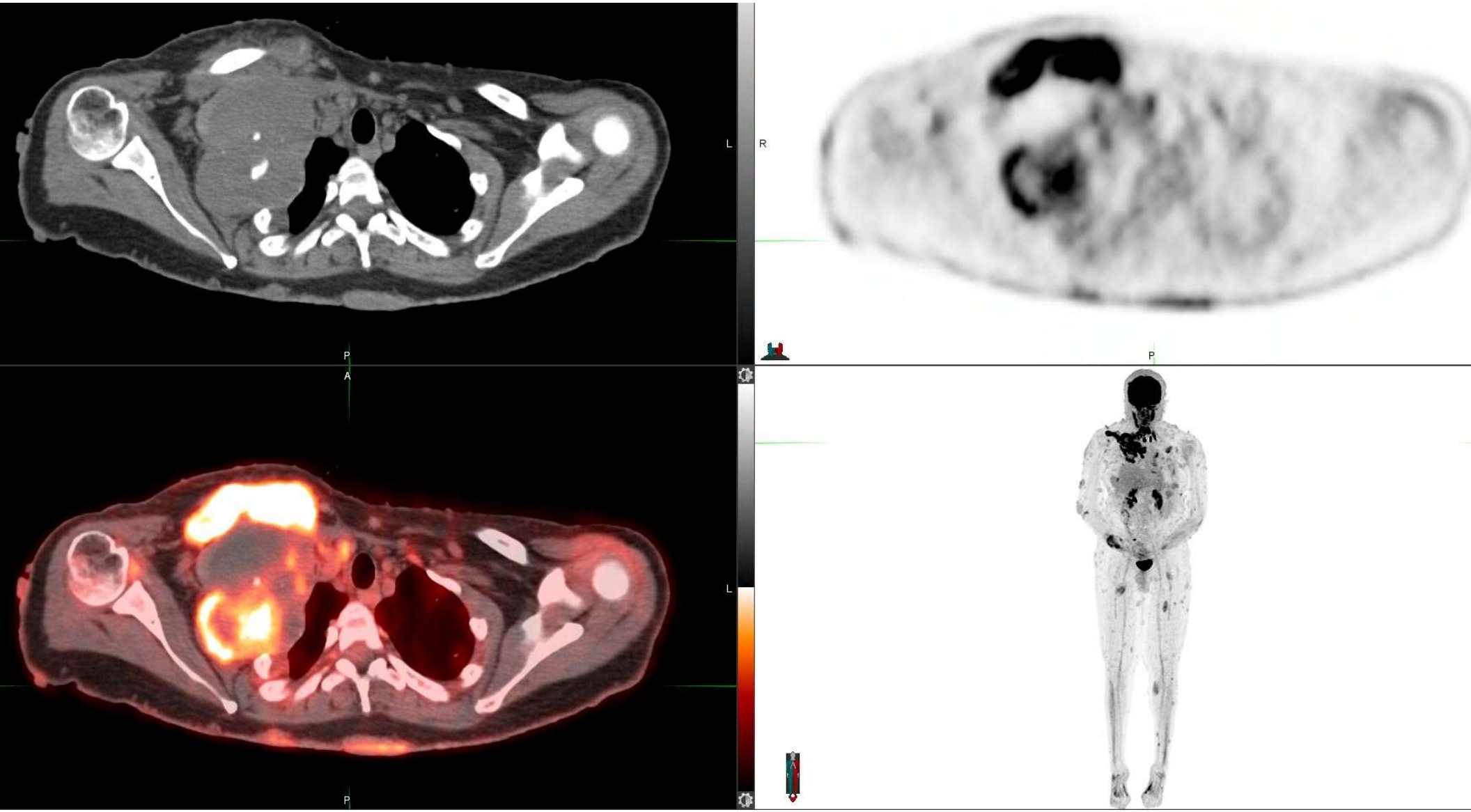
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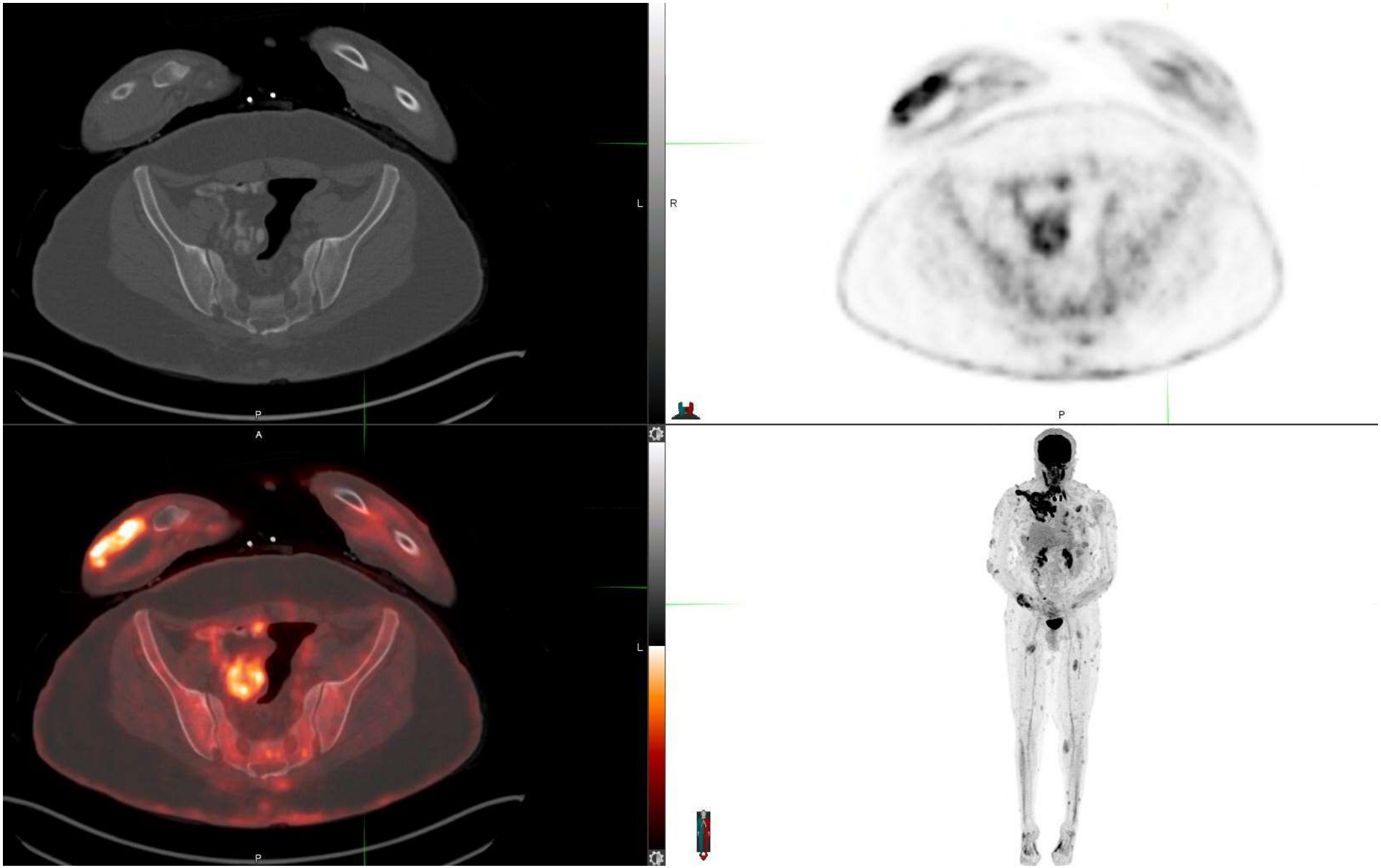
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Follow-up

Decision made to start Paclitaxel and Gemcitabine. Received 1st dose on 10/29/2018 and 2nd on 11/05/2018.

Angiosarcomas respond to first line chemotherapy but those arising from/ associated with malignant peripheral nerve sheath tumors (MPNST) in the context of neurofibromatosis 1, are less sensitive to chemotherapy.

Angiosarcoma and Malignant Peripheral Nerve Sheath Tumor with Neurofibromatosis Type 1.

Neurofibromatosis type 1 (NF1) is an autosomal dominant disorder with the incidence rate of 1 in every 3500 individuals.

Main characteristics are “café-au-lait” spots, cutaneous neurofibromas, and hamartomas of the iris (Lisch nodules).

NF1 patients have increased risk of developing benign and malignant tumors (neurofibromas, gliomas, malignant peripheral nerve sheath tumors (MPNSTs), and nonneural tumors (pheochromocytoma, myelogenous leukemia, and multifocal gastrointestinal stromal tumors).

Malignant Peripheral Nerve Sheath Tumor and Angiosarcoma with Neurofibromatosis Type 1.

Malignant peripheral nerve sheath tumor (MPNST) and angiosarcoma are rare tumors, even in those suffering from NF1.

Both histological types can be present in the same tumor mass in patients with NF1.

Some authors described these two entities within the same tumor mass, since heterologous differentiation in the form of angiosarcoma may occur in MPNST, more commonly in patients with NF1.

Malignant Peripheral Nerve Sheath Tumor and Angiosarcoma with Neurofibromatosis Type 1.

Reviewed 8 cases of angiosarcoma in patients with NF1 and found that 62.5% of them (5/8) were MPNST related, meaning they arouse as a sarcomatous transformation of a previously existing neurofibroma.

References

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