Southwest Journal of Pulmonary, Critical Care & Sleep

Journal of the Arizona, New Mexico, Colorado and California Thoracic Societies <u>www.swjpcc.com</u>

November 2022 Imaging Case of the Month: Out of Place in the Thorax

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History of Present Illness: A 30-year-old woman presented with complaints of left-sided back pain and numbness. She denied any history of trauma.

PMH, SH, FH: No significant past medical history. She denied smoking and use of illicit substances. Her family history was largely unremarkable, positive only for a history of gastrointestinal stromal tumor affecting her father.

Medications: Her medications included fluoxetine, spironolactone, and Celebrex (celecoxib).

Physical Examination: The patient's physical examination showed her to be afebrile with pulse rate and blood pressure within the normal range.

Laboratory Evaluation: A complete blood count showed a hemoglobin and hematocrit value of 14.3 gm/dL (normal, 13.2-16.6 gm/dL) and 41.5% (normal, 38.3-48.6%) and a platelet count of 253 x 10° /L (normal, 135-317 x10°/L). The white blood cell count was normal at 6.9 x10°/L (normal, 3.4-9.6 x10°/L), with no left shift. The eosinophil count was normal. Liver function studies were entirely normal. Serum chemistries were completely within normal limits aside from a minimally elevated serum calcium level of 10.1 mg/dL (normal, 6.6-10 mg/dL).

Frontal chest radiography (Figure 1) was performed.



Figure 1. Frontal chest radiography shows normal heart size, clear lungs, no evidence of pleural effusion or peribronchial or mediastinal lymph node enlargement. Click <u>here</u> to be directed to an enlarged figure in a separate window.

Which of the following represents <u>an</u> <u>appropriate interpretation</u> of the frontal chest and lateral radiograph?

- 1. Frontal chest radiography shows normal findings
- 2. Frontal chest radiograph shows numerous small nodules
- 3. Frontal chest radiography shows rib abnormalities
- 4. None of the above
- 5. More than one of the above

Correct!

4. None of the above

Frontal chest radiography shows normal heart size and clear lungs. No mediastinal or peribronchial lymphadenopathy is evident. No pulmonary nodules are seen. The frontal chest radiograph was prospectively interpreted as negative but the image is abnormal. Closely review Figure 1. Which structure(s) is/are <u>abnormal</u>?

- 1. The heart
- 2. The pleura
- 3. The thoracic spine
- 4. The ribs
- 5. More than one of the above

Correct!

4. The ribs

The heart is not only normal in size but the contour is also normal. The costophrenic angles are sharp and no pleural abnormality is seen. The thoracic spine shows no evidence of paravertebral abnormality and the vertebral body heights appear wellmaintained. The left inferior ribs, however, appear irregular.

Which of the following represents the <u>most</u> <u>appropriate description</u> of the appearance of the ribs in Figure 1?

- 1. Lytic, destructive lesions
- 2. Callus formation
- 3. Aggressive periosteal reaction
- 4. Smooth erosive change
- 5. Osteoblastic lesions

Correct!

3. Aggressive periosteal reaction

Lytic lesions typically appear as abnormally lucent bone, sometimes manifesting as a "punched out" appearance in the bones, focal osteolysis, or cortical destruction. Callus formation appears as smooth, well-formed periosteal reaction, this morphology commonly encountered with healed rib fractures. Smooth erosive change often appears as thinning of one margin of the osseous cortex and is commonly the result of a lesion adjacent to a bone slowly enlarging and exerting pressure on the affected bone. Osteoblastic lesions manifest as abnormally dense-appearing bones, or *sclerotic*, and may be encountered in isolation, as my occur with the commonly encountered enostosis, or "bone island," as multifocal lesions in the context of metastatic disease, or in a number of metabolic bone conditions. See Figure 2.



Figure 2. Detail view of the frontal chest radiograph (Figure 1A) shows well-formed periosteal reaction (arrows) involving the left 10th-12th ribs. Click <u>here</u> to be directed to an enlarged figure in a separate window.

Using the information above and presented in Figure 2, which of the following represents <u>the least likely consideration</u> for the patient's imaging findings?

1. Osteopoikilosis

- 2. Metastatic disease
- 3. Multiple myeloma
- 4. Osteomyelitis
- 5. Lymphoproliferative disorder

Correct!

1. Osteopoikilosis

A number of the conditions listed would be expected to cause aggressive periosteal reaction metastatic disease, multiple myeloma, osteomyelitis, and lymphoproliferative disease. Osteopoikilosis results in circumscribed rounded sclerotic foci- enostoses, or "bone islands which have a non-aggressive appearance.

A viral panel, including testing for SARS-CoV-2, was negative.

Which of the following represents <u>the most</u> <u>appropriate next step</u> for the patient's management?

- 1. Enhanced chest CT
- 2. ¹⁸FDG-PET scan
- 3. ⁶⁸Ga-PSMA-11 PET/CT
- 4. ⁶⁸Ga-pentixafor PET/CT
- 5. ⁶⁸Ga-DOTA-FAPI-04 PET/CT

Correct!

1. Enhanced chest CT

Enhanced chest CT would be an appropriate next step in the evaluation of this patient's chest wall abnormalities. Chest MRI would also be an appropriate choice but was not one of the listed options. ¹⁸FDG-PET could prove useful at some point in this patient's evaluation, but is premature as the use of ¹⁸FDG-PET is often more rewarding following anatomic characterization. ⁶⁸Ga-PSMA-11 PET/CT is a modality typically used for assessment for prostate carcinoma. ⁶⁸Ga-pentixafor PET/CT is a modality which may be employed for endocrinologic disturbances, notably Cushing syndrome or aldosterone-secreting tumors. ⁶⁸Ga-DOTA-FAPI-04 PET/CT is a modality that employs fibroblast activation protein inhibitors (FAPI) labeled with radionuclides that be used for

the diagnosis and treatment of various malignancies.

The patient underwent enhanced chest CT (Figure 3).

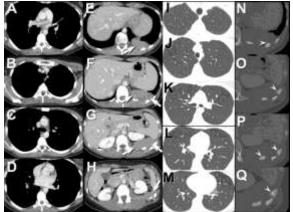


Figure 3. Enhanced axial chest CT displayed in soft tissue windows (A-H), lung windows, and bone windows (N-Q). Click <u>here</u> to be directed to an enlarged figure in a separate window.

Which of the following represents <u>an</u> <u>appropriate interpretation</u> for this examination?

- 1. Enhanced chest CT shows extensive mosaic perfusion
- 2. Enhanced chest CT shows a mass related to the chest wall
- 3. Enhanced chest CT shows multifocal bronchiectasis
- 4. Enhanced chest CT shows multifocal lymph node enlargement
- 5. Enhanced chest CT shows normal findings

Correct!

2. Enhanced chest CT shows a mass related to the chest wall

The enhanced chest CT sows that the lung parenchyma appears normal. There is no bronchiectasis and lung attenuation is homogeneous- there is no mosaic perfusion. No peribronchial or mediastinal lymph node enlargement is present. A homogeneous solid mass is present in the left posterior interior thorax. Based on the appearance on the chest CT Which of the following represents <u>an</u> <u>appropriate interpretation</u> for this examination?

- 1. The lesion probably arises from the subpleural lung
- 2. The lesion probably arises from the pleura
- 3. The left probably arises from chest wall soft tissues
- 4. The lesion probably arises from the posterior mediastinum
- 5. More than one of the above

Correct!

5. More than one of the above

The close contact with the chest wall, extension of the lesion into the adjacent chest wall musculature between the rib interspaces (Figure 4R-U), and the rib periosteal reaction all suggest that the lesion arises from the chest wall soft tissues, although a pleural origin is possible. The features of the lesion are not consistent with a lung origin, and the lesion is lateral to the lateral margin of the posterior mediastinum and therefore does not originate from this compartment. A prior whole-body screening MR from one year earlier was located for comparison (Figure 4).

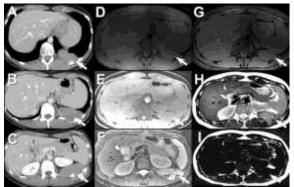


Figure 4. A previous whole-body screening MR 1 year earlier compared to the enhanced chest CT. Axial enhanced chest CT images

(A-C) through the left posterior inferior thoracic mass (arrows). Selected images from the whole-body unenhanced MR exam (D-I) show the lesion (arrows) was present one year earlier and has probably enlarged somewhat during the interval. Click <u>here</u> to be directed to an enlarged figure in a separate window.

Which of the following represents <u>an</u> <u>appropriate interpretation</u> for this examination?

- 1. There has been *no change* in the left posterior inferior thoracic lesion
- 2. The left posterior inferior thoracic lesion was previously *larger*
- 3. The left posterior inferior thoracic lesion was previously *smaller*
- 4. The left posterior inferior thoracic lesion was *absent*
- 5. No conclusion can be made as the MR examination did not include the area of interest

Correct!

3. The left posterior inferior thoracic lesion was previously smaller

Although the left posterior inferior thoracic lesion was not reported prospectively, the lesion was present on that study and was smaller. Interval growth between the previous whole-body **MR** and the chest **CT** has likely occurred.

Testing for coccidioidomycosis was unrevealing.

Which of the following represents <u>the most</u> <u>appropriate next step</u> for the patient's management?

- 1. Video assisted thoracoscopic surgical biopsy
- 2. ¹⁸FDG-PET scan
- 3. Bronchoscopy with transbronchial biopsy
- 4. Transthoracic needle biopsy
- 5. Open resection

Correct!

4. Transthoracic needle biopsy

Percutaneous transthoracic needle biopsy is the most appropriate choice. The lesion does not arise from the lung and is very peripherally located, and therefore a bronchoscopic approach is not likely to be rewarding. Both open resection and videoassisted thoracoscopic surgical biopsy would be capable of establishing a diagnosis but are needlessly aggressive at this point. ¹⁸FDG-PET scan may prove useful in this patient's evaluation at some point, particularly if malignancy is established, but at this juncture neither increased not absent glucose utilization within the lesion will later management.

Then patient underwent ultrasound-guided transthoracic needle biopsy (Figure 5) of the left posterior inferior thoracic lesion.

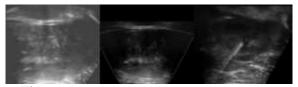


Figure 5. Ultrasound of the left posterior inferior thoracic mass shows the lesion to have mixed echogenicity, predominantly hypoechoic, without through sound transmission to suggest cystic change, and showing some internal vascularity. The final image shows the needle placed within the lesion during transthoracic percutaneous biopsy. Click <u>here</u> to be directed to an enlarged figure in a separate window.

Which of the following findings at biopsy would be <u>inconsistent</u> with the imaging appearance?

- 1. Solitary fibrous tumor of the pleura
- 2. Fibrosarcoma
- 3. Splenic tissue (herniation of spleen through a diaphragmatic rupture)
- 4. Lymphoma
- 5. Desmoid tumor

Correct!

3. Splenic tissue (herniation of spleen through a diaphragmatic rupture)

Review of Figure 4 shows that the spleen resides in its normal subdiaphragmatic position in the left upper quadrant and there is no evidence of diaphragmatic hernia. The solid, homogeneous nature of the lesion is consistent with a mesenchymal neoplasm, including sarcoma or desmoid tumor arising from the chest wall. A solitary fibrous tumor of the pleura is also a possibility, as is a lymphoproliferative lesion. Figure 6 shows a number of processes with an imaging appearance similar to the presented case.

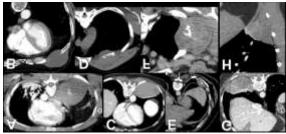


Figure 6. Lesions / processes similar in appearance at imaging to the presented case: **A.** Intrathoracic splenic herniation following blunt abdominal trauma. **B.** Desmoid tumor of the anterior chest wall. **C.** Pseudotumor.

D. Lymphoma. E. Fibrosarcoma arising from the pleural (calcification is evident). F. Sarcoma. G and H. Axial and sagittal images in a patient with solitary fibrous tumor of the

pleura. Click <u>here</u> to be directed to an enlarged figure in a separate window.

The percutaneous biopsy showed spindle cell tumor, with subsequent staining showing features consistent with aggressive fibromatosis / desmoid tumor.

Diagnosis: Aggressive fibromatosis / desmoid tumor.

Surgical consultation was conducted and the surgeon felt that surgery would not be efficacious given the potential for local recurrence after resection. Germ line testing was recommended, given the association of desmoid tumors with familial adenomatous polyposis syndrome. Treatment wit hormone manipulation, non-steroidal antiinflammatory agents, and local cryoablation were planned in favor of radiation therapy.

References

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