A 50 year old woman comes in for evaluation of swelling in her face and neck which has progressed over the past three weeks. The patient reports a history of patchy red areas with itching in the upper and lower extremities. She reports no recent changes in environmental exposure. Her blood pressure is 140/80, his temperature is 97.2, his pulse is 88, and his respiratory rate is 14. On physical examination, there is erythema over the face, neck, and upper chest. The patient’s face and neck appear swollen and show venous prominence. The patient has a few scattered red patches on the skin of her extremities. A chest radiograph was obtained.

Which of the following imaging studies is the most appropriate next step in further evaluation?
(a) positron emission tomography (PET) scan of the whole body
(b) computed tomography (CT) of the chest
(c) ultrasound (US) of the neck and both upper extremities
(d) magnetic resonance (MR) imaging of the chest
Which of the following imaging studies is the most appropriate next step in further evaluation?

(a) positron emission tomography with fluoro-deoxy glucose (PET-FDG) scan of the whole body
(b) computed tomography (CT) of the chest
(c) ultrasound (US) of the neck and both upper extremities
(d) magnetic resonance (MR) imaging of the chest

The chest radiograph shows a mediastinal mass (arrow). Computed tomography (CT) of the chest (b) is probably the best next step in further evaluation, and is the correct answer.

Positron emission tomography with fluoro-deoxy glucose (PET-FDG) scan of the whole body (a) is a method of determining metabolically active tissue. It is generally used in the evaluation of known malignancy (e.g., known lung cancer, esophageal cancer, colorectal cancer, head and neck cancer, and melanoma – see www.cms.gov) and evaluation of solitary pulmonary nodules. It is generally performed after and not before other cross sectional imaging studies, and (with the exception of a solitary pulmonary nodule) after a diagnosis has been obtained. Therefore, (a) is incorrect. US of the neck and both upper extremities (b) may be useful to evaluate the deep venous system in this patient if the chest radiograph was normal, but the chest radiograph shows an obvious mass that needs further evaluation and the first priority is to further characterize this lesion. Therefore, (b) is incorrect. MR imaging of the chest (d) is generally performed when contrast-enhanced CT of the chest cannot be performed in a patient with a known iodine-based contrast allergy, or when CT has been performed but has not provided enough information, but is usually not done prior to CT of the chest (in the absence of a contrast allergy) and (d) is incorrect.
The patient underwent imaging:

Imaging questions:
1) What type of study is shown?
2) Are there any abnormalities?
3) What is the most likely diagnosis?
4) What is the next step in management?
Imaging questions:

1) What type of study is shown? Chest CT. Panel A is an axial scan done at the level of the aortic arch filmed at “bone” windows, and Panel B is a coronal study at the location of the reference line in Panel A.

2) Are there any abnormalities? Yes. There is extensive abnormal soft tissue with stippled calcification in the mediastinum surrounding the aorta and superior vena cava (arrows in A) and there is narrowing of the superior vena cava by the (partially calcified) mass (arrow in B).

3) What is the most likely diagnosis? Fibrosing mediastinitis with associated superior vena cava compression.

4) What is the next step in management? Referral to a thoracic surgeon.

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PATIENT DISPOSITION, DIAGNOSIS, AND FOLLOW-UP

It was assumed on the basis of the clinical presentation (erythema nodosum and superior vena cava compression) along with the classic radiographic picture of a calcified mediastinal mass on the CT study that the patient had fibrosing mediastinitis (also known as sclerosing mediastinitis or mediastinal fibrosis) as a sequelae of infection with Histoplasma capsulatum. In these cases, biopsy is generally not performed, because biopsy of an area dense fibrosis and calcification with extensive engorged collateral vessels may lead to uncontrollable bleeding. The patient was referred to a thoracic surgeon who, in turn, referred the patient to an interventional radiologist. The interventional radiologist placed a stent across the superior vena cava and this eliminated the facial swelling, erythema, and venous prominence.
SUMMARY

Presenting symptom: In patients with face and upper extremity swelling and venous prominence, superior vena cava compression should be suspected. Such swelling may be accompanied by cough, chest pain, or dysphagia.

Imaging work-up: The first step in the imaging evaluation should be a two view chest radiograph, as was obtained in this case. A chest CT is usually performed next. If the chest radiograph is negative, there may still be a lesion of the mediastinum causing superior vena cava obstruction, and if the chest radiograph shows a mass, CT demonstrates the degree and location of superior vena cava obstruction.

Establishing the diagnosis: When a patient has classic clinical features of superior vena cava syndrome, with face and neck swelling and venous congestion (with or without cough, chest pain, or dysphagia) and classic CT findings of extensive calcifications of the mediastinum with associated compression of the superior vena cava, the diagnosis of fibrosing mediastinitis usually does not require a biopsy. Biopsy of the dense, calcified mediastinal tissue (which frequently has extensive collaterals) may lead to uncontrolled bleeding.

Take-home message: The first step in imaging patients with clinical features of superior vena cava syndrome is two view chest radiography. This is often followed by a chest CT, which allows demonstration of the degree and location of superior vena cava obstruction as well as demonstrating the classic calcified soft tissue mass allowing presumptive diagnosis.

FURTHER READING
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