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Clinical Image

Posterior Mediastinal PEComa: Rapid Response to Sirolimus Therapy

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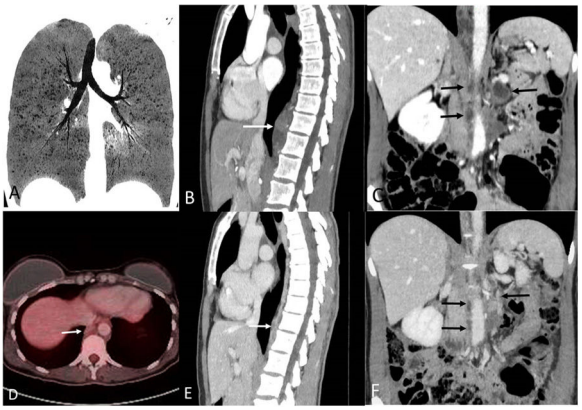

Fig. 1. (A) Coronal minimum intensity projection (minIP) thoracic CT image (lung window) shows multiple small cystic lesions in both lungs. (B) Sagittal thoracic CT image (mediastinal window) shows a low-attenuation prevertebral mass (arrow). (C) Coronal abdominal CT image shows low-attenuation lesions in the retroperitoneum (arrows). (D) Axial fused lower thoracic PET/CT image shows minimal FDG uptake by the posterior mediastinal lesion. (E) and (F) Sagittal thoracic (E) and coronal abdominal (F) CT images show an almost complete resolution of the masses (arrows) 3 months after sirolimus therapy was started.

A 47-year-old woman who had undergone resection of a large retroperitoneal perivascular epithelioid tumor (PEComa) six weeks earlier was unexpectedly found to have multiple lung cysts, mediastinal and retroperitoneal masses on whole-body computed tomography (CT) imaging (Fig. 1A–C). The mediastinal masses had the same features on CT as the retroperitoneal ones (low-attenuation with some cystic areas) and did not show significant fluorodeoxyglucose (FDG) uptake on positron emission tomography (PET)/CT (Fig. 1D). The patient was followed without treatment for 15 months, during which she developed a right chylothorax, a slow but progressive growth of some of the mediastinal and retroperitoneal lesions, and mild left flank pain. Once sirolimus therapy was started, the flank pain and the right pleural effusion disappeared and mediastinal and retroperitoneal lesions showed an almost complete resolution (Fig. 1E, F).

PEComa represents a rare mesenchymal neoplasm with variable biological behavior that occasionally occurs in patients with tuberous sclerosis complex. Although PEComas can occur in any part of the human body, the retroperitoneum and the uterus are the most affected sites. Mediastinal PEComas are extremely rare, most commonly involve the anterior mediastinum, and show nonspecific imaging features.[1] A recent paper has shown that sirolimus (a mammalian target of rapamycin [mTOR] inhibitor) may stabilize lung function, decrease PEComas growth, and stop new cyst formation, but to our knowledge response demonstration on imaging of mediastinal PEComas has not been published.[2]

References

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