Interesting image

Pulmonary epithelioid hemangioendothelioma: Nuclear medicine and 18F-FDG PET/CT findings

Hemangioendotelioma epitelioide pulmonar: Medicina Nuclear y hallazgos de 18F-FDG PET/CT

M. Yang*, B.D. Nguyen, M.C. Roarke

Department of Radiology, Mayo Clinic, AZ 85259, USA

A 46-year-old man, with a remote history of lower extremity deep venous thrombosis, consulted the emergency room for acute shortness of breath, right-sided chest pain and left knee pain. The patient also complained of a 3-month chronic thoracic pain and 30-lbs weight loss. Physical examination revealed clubbed fingers (Fig. 1). Chest CT angiography showed no pulmonary embolism, but demonstrated mediastinal lymphadenopathy with right middle and lower lung consolidation. 99mTc-MDP whole body bone scan exhibited features of hypertrophic osteoarthritis (HOA) at long bones and zygomatic processes, suspicious for paraneoplastic manifestation of lung malignancy (Fig. 2A). Staging 18F-FDG PET/CT study showed FDG-avid lesions corresponding to the right hilar/perihilar consolidative mass (SUV 8.4), subcarinal (SUV 6.1) and left hilar adenopathy (SUV 5.7), as well as right diaphragmatic and bilateral pleural lesions (Fig. 3A–C). Transbronchial right middle lobe biopsy, based on the most FDG-avid lesion, and immunohistochemistry staining showed a CD31-positive lesion consistent with pulmonary epithelioid hemangioendothelioma (PEH).

PEH is a rare vascular tumor with epithelioid appearance. According to the 2004 World Health Organization Classification, PEH is regarded as low- to intermediate-grade vascular neoplasm. It affects preferentially the adult with female gender predominance. There are three distinctive CT patterns of PEH: lung nodules, pulmonary reticulonodular opacities, and diffuse infiltrative pleural thickening, all depicted in our patient. The latter pattern mimics malignant pleural mesothelioma. In our case, the mediastinal, bilateral hilar, lung parenchymal, bilateral pleural, and right sub-diaphragmatic findings, best depicted by PET/CT imaging, indicated an advanced stage of the disease. In prior report, PET/CT finding was used as an indicator for tumor resection. In our unresectable PEH, PET-CT provided valuable staging information and biopsy guidance.

HOA, previously reported with PEH, is a paraneoplastic syndrome characterized by periosteal new bone formation, clubbed fingers, arthralgia and arthritis. It is usually regarded as an ancillary finding encountered in chronic pathological lung processes and primary thoracic malignancies. It commonly involves appendicular bones, and less frequently affects the skull, scapula and clavicle. Our case presented the typical clinical manifestations of HOA, including clubbed fingers, left knee pain, as well as characteristic bone scintigraphic features including the uncommon bilateral zygomatic involvement. The bilateral zygomatic processes and proximal femoral bones demonstrate cortical hyperostosis on transmission CT without abnormal FDG avidity on PET (Fig. 2B and C). In summary, in this case presentation, the complementary imaging features of bone scintigraphy and PET/CT show the advanced stage of PEH of the lung with its related osseous paraneoplastic manifestations.
Fig. 2. Bone scintigraphy image shows typical HOA along bilateral femurs, tibia and zygomatic process (A, arrows). Axial CT attenuation correction images show focal hyperostosis involving bilateral zygomatic processes (B, arrows) and bilateral proximal femurs (C, arrows). None of the lesions are FDG avid.

Fig. 3. ^18^F-FDG PET-CT MIP image demonstrates hypermetabolic lymph nodes in right hilar, subcarinal and left hilar regions (A, arrows). Axial emission, fused and CT attenuation correction images of the chest show right middle lobe hypermetabolic consolidative lesion, corresponding with biopsy site (B, arrows). Axial emission, fused and CT attenuation correction demonstrate hypermetabolic nodule in right subdiaphragmatic region (C, arrowheads) and hypermetabolic lesion involving pleural space (C, arrows).
References

