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Case Study

Role of PET-CT in POEMS Syndrome - Case Study (AFIRI-MH)

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Abstract

POEMS syndrome is a lymphoproliferative disease, a paraneoplastic disorder. POEMS (polyneuropathy, organomegaly, endocrinopathy, M-protein and skin changes). Moreover its other important traits include sclerotic bone lesions, thrombocytosis, pappill-oedema, clubbing, castlemans disease [1]. The radiological findings of bone lesions differ from those in myeloma in the sclerotic appearance of focal lesions [2,4]. In the majority of patients the diagnosis is confirmed by the identification of monoclonal plasma cells obtained through biopsy of an osteosclerotic lesion; as such, a whole-body X-ray survey is essential to detect focal lesions [5-7].

Keywords: *POEMS Syndrome; POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, M-Protein and Skin Changes); PET-CT*

Introduction

POEMS syndrome is a lymphoproliferative disease, a paraneoplastic disorder.

History

A 75 years old male with complaints of joint pains, weakness of whole body, hyperpigmentation, weight loss and deranged LFT's. Plasma electrophoresis shows presence of M-proteins.

Clinical symptoms

- Complaints: Eczema, weight loss, joint pains, weight loss and burning micturition.
- Examination: Systemic examination was otherwise unremarkable.

Investigations

- BCP: WNL, LFT's: Increased ALT count, RFT's: WNL
- USG abdomen: Bilateral renal cysts
- CXR: Normal.

Diagnosis

Plasma electrophoresis shows M-protein suggestive of POEMS syndrome keeping in view the major and minor criterion for it.

PET-CT scan for disease characterization

8.47 mCi (^{18}F) - fluorodeoxyglucose was administered intravenously via left hand. To allow for distribution and uptake of radiotracer, the patient was allowed to rest quietly for 52 minutes in a shielded room. Imaging was performed on an integrated 64-slice PET/CT scanner, with scanning from whole body. Blood glucose at the time of the injection was measured at 107 mg/dL. Serum creatinine is 115 µmol/L. CT scanning was performed with intravenous contrast material. All reported uptake values are maximum SUVs unless stated otherwise.

Discussion

Reference hepatic SUV = 4.1.

Sections through brain show physiological parenchymal uptake with no focal lesion. Activity within the glottic region, tonsils and salivary glands is physiological. Size stable insignificant discrete non-avid subcentimetre size bilateral level II and V cervical nodes. No avid or size significant supraclavicular nodes.

Size stable avid discrete subcentimetre size bilateral hilar and mediastinal nodes at right lower paratracheal-4R, right inferior tracheobronchial, right hilar-10/11R and left hilar-10L nodal stations, likely reactive. No avid or size significant axillary nodes noted. For reference, right lower paratrachel-4R node 0.6 cm 3.4 SUV (previously same size 3.2 SUV), right hilar-10R 0.8 cm 7.5 SUV (previously same size 8.7 SUV), left hilar-10L 0.6 cm 4.5 SUV (previously same size 5.4 SUV) nodes. Size stable multiple fairly defined subcentimetre size metabolically insignificant bilateral pulmonary nodules, likely old inflammatory. For reference, right upper lobe apical segment nodule 0.3 cm size. No pleural or pericardial effusion.

Bosniak type I non-avid bilateral multiple renal cysts. For reference, left lower pole renal cyst 2.4 cm size. Liver, spleen, pancreas, adrenals and gallbladder are unremarkable. Bowel shows physiological tracer uptake. No avid or size significant para-aortic, pelvic sidewall or inguinal nodal disease. Pelvic viscera outlines normally. No ascites. Stable appearing proximal left fibular focal expansion, with subtle sclerosis, 3.2 cm size 1.4 SUV, remains metabolically insignificant. Stable appearing non-avid spinal degenerative changes. No destructive osseous lesion or abnormal marrow uptake.

Conclusion

The PET-CT scan of the patient reveals, Stable appearing proximal left fibular focal expansion, with subtle sclerosis, remain metabolically insignificant. Stable appearing spinal degenerative changes. Size stable hilar and mediastinal nodes at right lower paratracheal-4R, right inferior tracheobronchial, right hilar-10/11R and left hilar-10L nodal stations, likely reactive. Size stable level II and V cervical nodes, remain metabolically insignificant.

Size stable multiple pulmonary nodules, likely old inflammatory [15,16]. Bosniak type I multiple renal cysts.

Understanding links between the monoclonal lambda plasma cell disorder and resulting proinflammatory cytokine milieu is fundamental to determining POEMS syndrome pathophysiology. Similarities to chronic inflammatory demyelinating polyradiculoneuropathy and some other monoclonal proliferative diseases makes POEMS misdiagnosis common [8,9]. A range of treatments are available, and more work to identify pathogenic mechanisms and treatment targets and prognostic scores will further enable treatment stratification for optimum outcomes [10,11].

^{18}F-FDG PET/CT is a useful tool for evaluating patients with suspected POEMS syndrome [12-14]. ^{18}F-FDG PET/CT may contribute to the diagnosis, evaluation, and follow-up of patients with POEMS syndrome by providing systematic findings of the bone lesions, lymphadenopathy, liver or spleen involvement, serous cavity effusion, and the metabolic status of the lesions [16].

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