

Unusual Case of Recurrent Pericardial Effusion — Erdheim-Chester Disease —

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A 62-year-old woman with a history of recurrent pericardial effusion (PE) was admitted for aggravation of dyspnea. Echocardiogram showed severe PE with hemodynamic compromise (**Figure A**). Previously she had undergone pericardiocentesis, but no specific findings were noted on cytology (e.g., malignancy or tuberculosis). The patient had pericardial window formation with pericardial biopsy, which showed lymphohistiocytic infiltration, but this was not sufficient for diagnosis.

We performed full diagnostic work-up. Positron emission tomography-computed tomography (PET-CT) showed increased fluorodeoxyglucose uptake in the pericardium, thoracic aortic wall and bilateral tibia (**Figure B1–B3**, white arrows) but bone biopsy was normal, even after being performed twice (proximal site of both tibias). Brain magnetic resonance angiography showed aneurysm of the anterior communicating artery (**Figure C**, white arrow) and coronary angiogram indicated diffuse multiple stenosis (**Figure D**), suggesting systemic vasculitis. Chest CT showed aorta wall enhancement, suggesting aortitis/periaortitis (**Figure E1,E2**, blue arrows) and interlobular septal thickening (**Figure F**). Therefore, we carried out video-assisted thoracoscopic surgery assisted wedge resection, and lung biopsy showed histiocytic infiltration and fibrosis (**Figure G**). On immunohistochemistry staining, CD68 (PG-M1;

Figure H, blue arrows) and S-100 were positive, and BRAF (VE1) was weakly positive. The pathology was compatible with Erdheim-Chester disease (ECD). The patient was discharged without PE and stable in the outpatient clinic on low-dose oral steroid.

ECD is a rare non-Langerhans histiocytic disorder. We report this ECD case, initially presenting with PE and involvement of the long bones, cardiovascular and pulmonary system.¹ Considering the various clinical manifestations and the difficulty in identifying this rare disease, meticulous diagnostic work-up is crucial for diagnosis.²

Disclosures

The authors declare no conflicts of interest.

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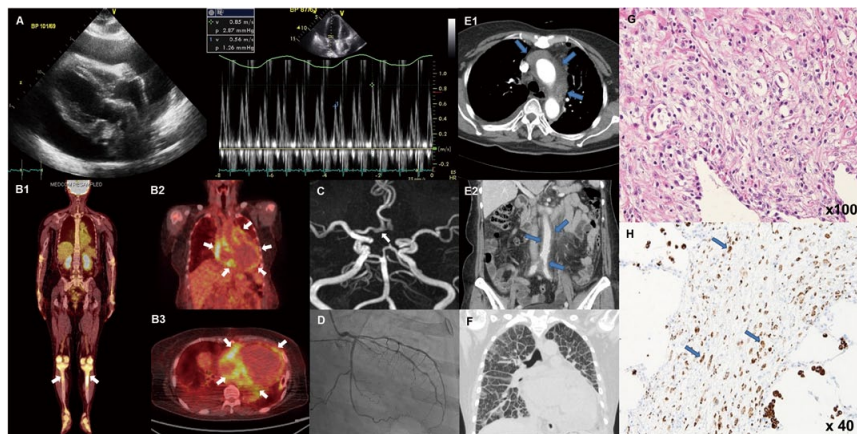


Figure. (A) Transthoracic echocardiography showed severe pericardial effusion with increased respiratory variation in mitral peak E-wave velocity. (B1–B3) Positron emission tomography-computed tomography showed increased fluorodeoxyglucose uptake in the pericardium, thoracic aortic wall and bilateral tibia (arrows). (C) Brain magnetic resonance angiography showed aneurysm of the anterior communicating artery (arrow). (D) Diffuse coronary artery stenosis on coronary angiogram, suggesting vasculitis. (E1,E2) Chest and abdominal-pelvic CT showed increased enhancement of soft-tissue infiltrations along the aorta (arrows) and (F) smooth interlobular septal thickening in both lungs. (G) On lung biopsy, hematoxylin and eosin staining showed histiocytic infiltration and fibrosis. (H) CD68 (blue arrows) and S-100 were positive, and BRAF (VE1) was weakly positive on immunohistochemistry, suggesting Erdheim-Chester disease.