Recurrent pericardial effusion with unexplained central diabetes insipidus, and hydronephrosis

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A 67-years-old female with aggravated dyspnea for a month admitted. Four months ago, she had pericardiocentesis due to pericardial effusion (PE). She had been treated with central diabetes insipidus (DI) and hyperprolactinemia with pituitary adenoma for more than ten years. Also, she had been taken ureteral stent insertion several times because of bilateral hydronephrosis (HN) with urinary tract infection. As PE with hemodynamic compromise figured out by transthoracic echocardiogram (TTE), we did pericardiocentesis (Figure 1A). Pericardial fluid was exudate. Either bacterial or mycobacterial culture was not diagnostic. After a weak, follow-up TTE showed recurrent PE and we did pericardial window formation. The pathology of pericardium showed histiocytic infiltration with fibrosis, which suggesting Erdheim-Chester Disease (ECD). Real time PCR showed BRAF V600E mutation. To support the diagnosis of ECD, we did several exams. We could diagnose ECD from following results, which suggestive of organ involvement of ECD: Cardiac Magnetic Resonance Imaging (MRI), diffuse pericardial enhancement with thickening (Figure 1B); Positron Emission Tomography - Computed Tomography, fluorodeoxyglucose uptake along pericardium and both distal femoral and distal tibia (Figure 1C); Abdominopelvic Computed Tomography, soft tissue infiltration at bilateral renal pelvis and left proximal ureter (Figure 1D); Brain MRI, subtle T2 high signal intensity in pons (Figure 1E). ECD is a rare disease that involves multiple organs with infiltration of non-Langerhans histiocytes. Clinical manifestation of ECD has broad spectrum including skeletal, endocrine, cardiovascular, neurologic, urologic symptoms and signs. Pathologically produced histiocytes can infiltrate pituitary gland, which leads to endocrine manifestations such as hyperprolactinemia and central DI. Also, it infiltrates retroperitoneal space, which leads to HN caused by ureteral compression. In the light of this case, central DI and recurrent HN can be considered as one of clinical manifestations of ECD. We can learn a lesson that clinical manifestations of EnCD may not be revealed simultaneously.

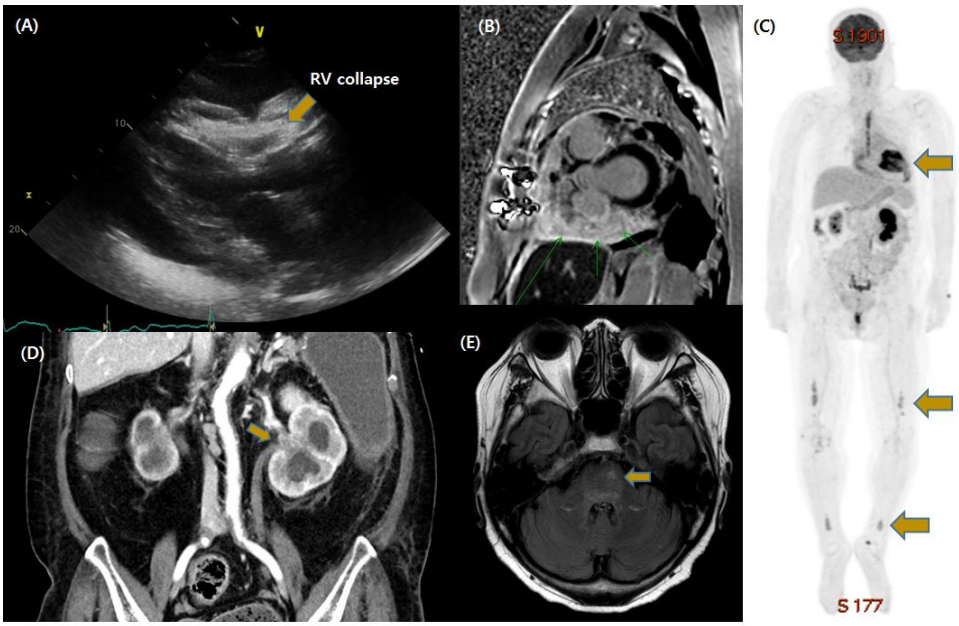

Figure 1. **A)** TTE Parasternal long axis view, large amount of PE with hemodynamic compromise, **B)** Cardiac MRI, diffuse pericardial enhancement with thickening, **C)** PET-CT, FDG uptake along pericardium and both distal femoral and distal tibia, **D)** APCT, soft tissue infiltration at bilateral renal pelvis and left proximal ureter, **E)** Brain MRI, subtle T2 high signal intensity in pons