Cardiac Erdheim-Chester

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A 59-year-old woman presented with an 8-month history of neurologic disturbance. On examination, she had eyelid xantelasmas, dyspnea and diminished heart sounds. Chest X-ray showed cardiomegaly. Pericardial effusion was confirmed on two-dimensional echocardiogram. Laboratory findings revealed an elevated erythrocyte sedimentation rate. Contrast-enhanced MDCT scan of the chest showed on axial reformation, pericardial and pleural effusion (stars) and infiltration of the atrioventricular groove and right atrial wall as a soft-tissue pseudomass of low intensity (arrow). RA: right atrium, RV: right ventricle, LA: left atrium, LV: left ventricle

Picture 1A. Contrast-enhanced MDCT scan of the chest on axial reformation showed pericardial and pleural effusion (stars) and infiltration of the atrioventricular groove and right atrial wall as soft-tissue pseudomass of low intensity (arrow). RA: right atrium, RV: right ventricle, LA: left atrium, LV: left ventricle

Immunohistochemistry of the pericardial biopsy demonstrated the presence of non-Langerhans histiocytes markers, positivity for CD68, and negativity for CD1a and PS-100 protein, confirming the diagnosis of ECD (Picture 1B).

Picture 1B. Immunohistochemistry of pericardial biopsy showed non-Langerhans histiocytes cells which exhibited marked CD68.

Picture 1C. Anteroposterior radiographs of two legs showed symmetric osteosclerosis of both tibias.

(Picture 1B). Anteroposterior radioagaphs of two legs shows pathognomonic bilateral symmetrical diaphyseal

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osteosclerosis (Picture 1C). The patient was treated with oral
corticosteroids and interferon-α, with clinical stability and
less dyspnea and three months of receding.

Erdheim-Chester disease is a non-Langerhans cell histio-
cytosis of unknown etiology (1). It’s a multi-systemic dis-
eease that characteristically affects long bones bilaterally and
symmetrically. Cardiovascular involvement can affect all the
cardiac layers and the aorta (“coated aorta”). The diagnosis
is based on the presence of symmetric bilateral osteosclero-
sis of the metaphysis and diaphysis of the long bones, and
confirmed by histologic examination which shows spumous
histiocytes CD68+, PS100+/-, CD1a- (2).
The prognosis is extremely variable and is often worse
when there is a cardiovascular and/or central nervous system
involvement. The treatment is controversial; it is often based
on corticosteroids and/or interferon alpha (3).

References

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