TREATMENT OF ERDHEIM-CHESTER SYNDROME: INTERFERON-ALPHA AND FUTURE PERSPECTIVES

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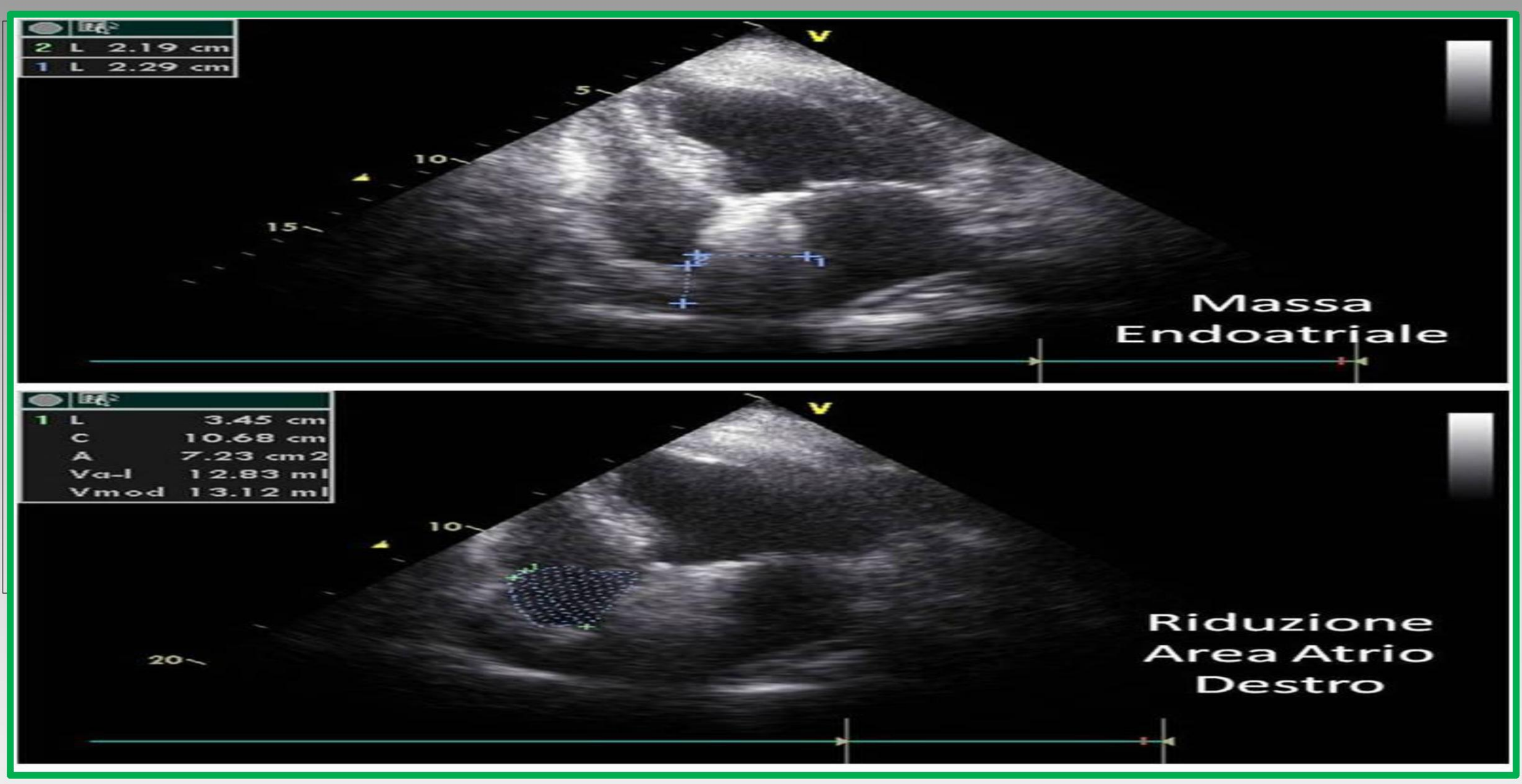
OBJECTIVES

Erdheim-Chester disease is a rare form of histiocytosis characterized by xantomatosa multiorgan tissue infiltration by histiocytes CD68 + / CD1. Bone involvement is the most common symptom. Half of the patients also have exophthalmos, pulmonary and retroperitoneal fibrosis , diabetes insipidus and involvement of the central nervous cardiovascular and renal system.

METHODS

A 77 years man , came to our observation for 'increase of renal function index (creatinine 2.0mg/dl), marked asthenia and hypertension. MRI of the upper abdomen documented the presence of pathological tissue infiltrating the renal sinus fat up to the region of the joint pielo ureter, surrounding the renal parenchyma bilaterally and pelvis sleeve. The x-ray of the long bones showed the presence of medullary osteosclerosis. The echocardiographic examination documented the presence of abundant pericardial effusion (570cc), a cloth epicardial and masses atrial on the interatrial septum . After performing brain MRI was diagnosed with Erdheim-Chester syndrome with renal, cardiac, bone long involvement , without pulmonary and cerebral localization. Was initiated therapy with interferon-α before, with clinical worsening general and with interleukin-1 receptor antagonist after.



CONCLUSIONS

At present there is no specific therapy for Erdheim-Chester syndrome, many authors suggest as first line treatment interferon-α, which in our case has yielded poor results. Among the future perspectives may play an important role in receptor antagonist interleukin-1.

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Immune and inflammatory mechanisms

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