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Pericardial Synovial Sarcoma: An Incidental Case of Primary Cardiac Tumor

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Abstract

Pericardial synovial sarcoma is a very rare malignant soft-tissue tumor. It is mandatory to exclude metastatic origins. The diagnosis is based on multidisciplinary approach, including morphology, immunostaining and cytogenetic analysis.

Keywords: Sinovial Sarcoma; pericardial; TLE-1; SYT

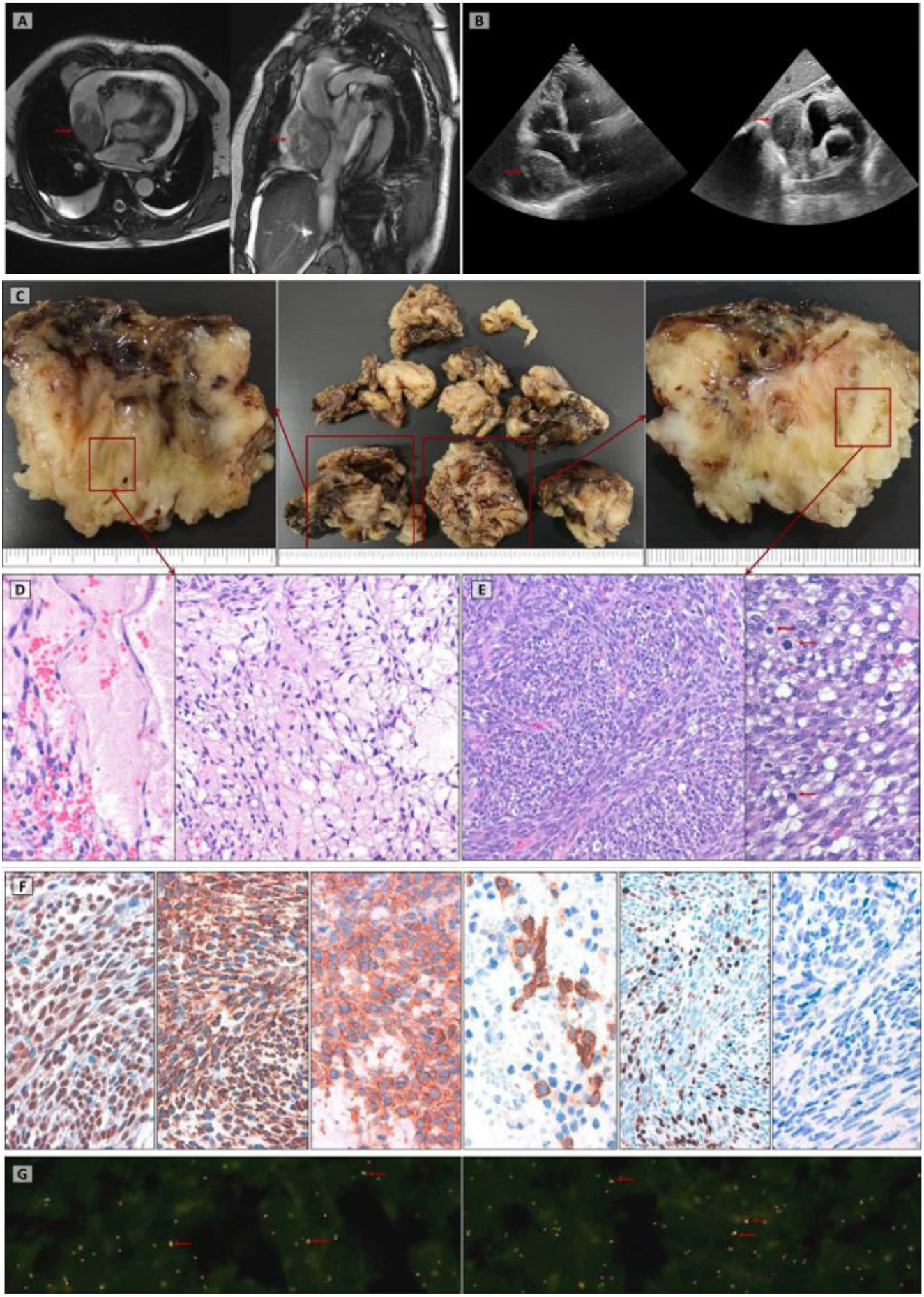
A 71-year-old man was admitted for constipation, nausea, and right abdominal pain. The abdominal computed tomography showed incidental pericardial effusions. The cardiac magnetic resonance showed a 6.2 x 7.8 cm right retroatrial intra-pericardial mass with irregular borders, no atrial wall infiltration, and pericardial effusion (Panel A).

Echocardiography demonstrated a mass contacting and imprinting of the posterior right atrial wall, without occluding cavae veins (Panel B).

These findings suggested a metastasis or primary pericardial neoplasia. Complete resection surgery was performed.

Macroscopically, a fragmented tumor measuring 7 cm, weighing 200 g, was observed. In section, it showed whitish solid areas alternating with others of gelatinous consistency (Panel C). Histologically, it was a malignant proliferation with myxoid hypocellular (Panel D) and storiform patterns, composed of spindle-shaped cellularity, with frequent mitoses (Panel E). On immunostaining, the cells were positive for TLE-1, CD99, bcl-2, EMA, high Ki-67, and negative for CKAE1-AE3 (Panel F, left to right) among others. Fluorescent in-situ hybridization was positive for SS18 (SYT) gene rearrangement on chromosome 18q11, a t(X;18) translocation equivalent (Panel G). The diagnosis was pericardial synovial sarcoma (PSS). After eight months, the patient is asymptomatic.

Primary PSS is a rare malignant soft-tissue tumor of unknown incidence, difficult diagnosis, and poor prognosis.The particularity of this case stems from the absence of cardiorespiratory symptoms regardless of the size. Complete resection of the tumour appeared to be an independent prognostic factor associated with survival. When surgical resection is unsuccessful, multimodal management improves patient outcomes.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