CLINICAL IMAGE

Right atrial angiosarcoma in a pregnant woman: diagnostic and therapeutic dilemmas

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A 33-year-old pregnant woman in the 23rd week of her third pregnancy (G3P1A1: gravidity, 3; parity, 1; abortion/miscarriage, 1) was admitted to an intensive cardiac care unit due to new-onset atrial fibrillation. Laboratory results showed mild anemia, D-dimer levels of 2111 ng/ml, and β-chorionic gonadotropin levels of 6850 mIU/ml. Transthoracic echocardiography (TTE) revealed a pathological mass in the right atrium, measuring 70 × 65 mm (FIGURE 1A). The mobile fragment of the tumor had a size of 22 × 14 mm, visible in the inflow tract of the right ventricle, increasing the risk of pulmonary embolism (FIGURE 1B). Ultrasonography showed a metastasis to the sixth rib and to the liver (FIGURE 1C and 1D). Magnetic resonance imaging (MRI) revealed a tumor measuring 82 × 71 × 114 mm, infiltrating the right atrium, tricuspid valve, narrowing

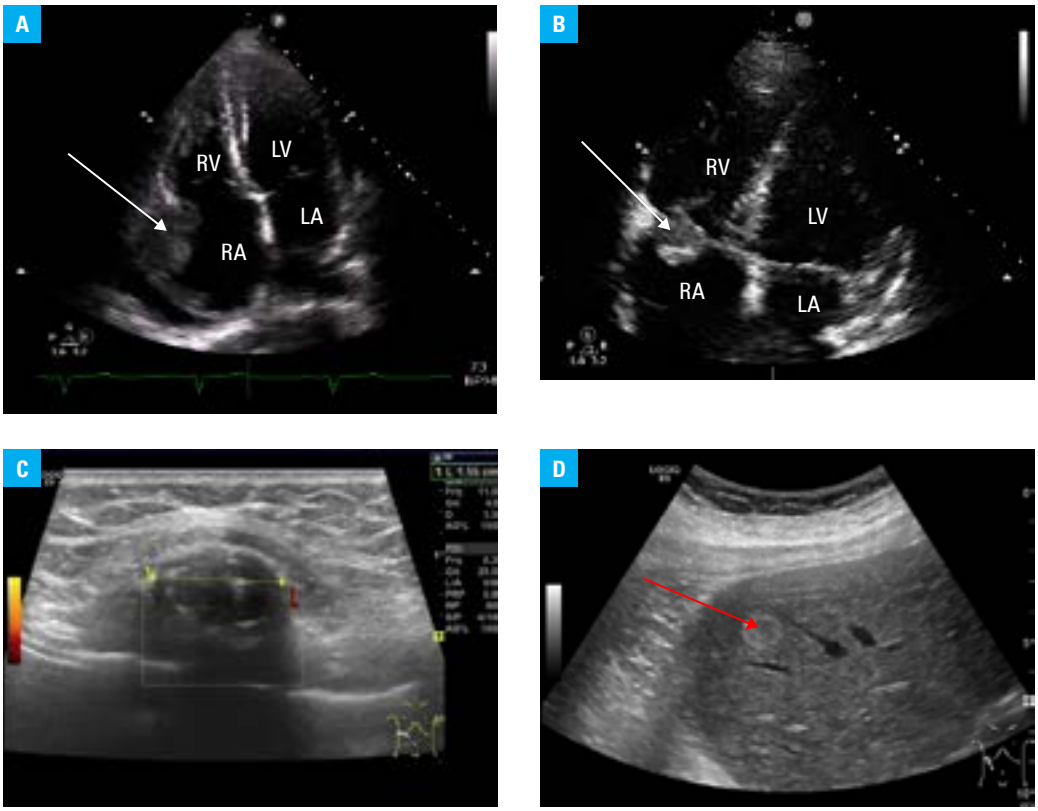

FIGURE 1 Transthoracic echocardiography (apical 4-chamber view) images showing the tumor of the right atrium (**A**, white arrow) and the mobile fragment of the tumor (**B**, white arrow); ultrasound images showing metastasis to the sixth rib (**C**) and metastasis to the liver (**D**, red arrow)
Abbreviations: see on the next page

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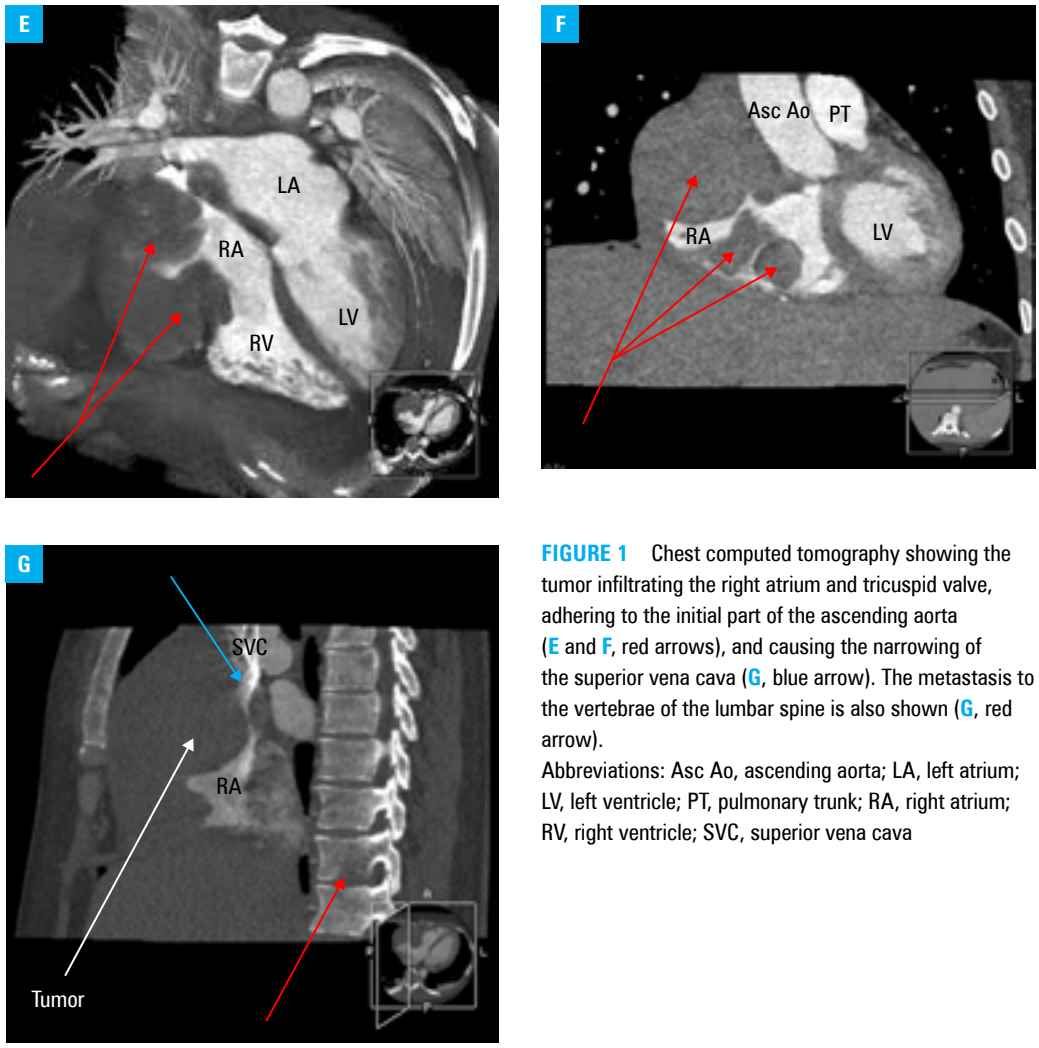

FIGURE 1 Chest computed tomography showing the tumor infiltrating the right atrium and tricuspid valve, adhering to the initial part of the ascending aorta (**E** and **F**, red arrows), and causing the narrowing of the superior vena cava (**G**, blue arrow). The metastasis to the vertebrae of the lumbar spine is also shown (**G**, red arrow).
Abbreviations: Asc Ao, ascending aorta; LA, left atrium; LV, left ventricle; PT, pulmonary trunk; RA, right atrium; RV, right ventricle; SVC, superior vena cava

the superior vena cava, and adhering to the initial part of the ascending aorta. Additionally, it showed numerous scattered metastatic lesions in the skeletal system: ribs, sternum, pelvis, and lumbar and thoracic vertebrae. After pregnancy, the patient was examined by chest computed tomography angiography (FIGURE 1E–1G), which confirmed our result obtained by MRI and ultrasonography. Trepanobiopsy of the sixth rib was performed. A histopathological examination showed angiosarcoma.

After the introduction of betamethasone in the 27th week of pregnancy, a Cesarean section was performed. The patient gave birth to a daughter weighing 1020 g, with an Apgar score of 5 and 7 points at 1 and 10 minutes, respectively. TTE performed on the newborn showed significant atrial septal defect type II (ASD II) and patent ductus arteriosus (PDA). The daughter was discharged from the hospital in good condition, weighing 2990 g. In an oncology center, the mother received the eight cycle of chemotherapy (ifosfamide, adriamycin, vincristine) and radical radiotherapy of the lumbar spine. Our patient died due to neoplastic complications 13 months after the diagnosis. Her child develops normally after surgical correction of PDA and ASD II.

Literature data regarding cardiac angiosarcoma in pregnancy are limited mostly to case reports and case series. Cardiac angiosarcoma is characterized by rapid growth, resulting in a mass effect when the tumor obstructs cardiac output, or in local invasion, embolization, or systemic manifestations.[1] Most tumors (about 90%) are located in the right atrium, originating from the lateral wall; the second most frequent location is the left atrium, followed by the right ventricle, and finally the left ventricle.[2,3] Because of the preferential right-sided location, patients with angiosarcoma often present with symptoms of heart failure and superior vena cava syndrome.[4] Surgical resection with or without adjuvant chemotherapy or radiotherapy is the main treatment, and complete excision is the most important prognostic factor.[4]

Our patient was consulted in 2 different cardiac surgery centers and she was referred only for chemotherapy and radiotherapy. After the diagnosis of angiosarcoma, an elective Cesarean section in the 27th week of pregnancy allowed delivery to a child with low weight at birth with good prognosis and early start of the mother's chemotherapy and radiotherapy. This is a rare case of the very unusual coincidence of pregnancy with asymptomatic primary malignant heart tumor, the first symptom of which was paroxysmal atrial fibrillation.

REFERENCES

1 Azimi NA, Selter JG, Abott JD, et al. Angiosarcoma in a pregnant woman presenting with pericardial tamponade - a case report and review of the literature. Angiology. 2006; 57: 251-257.

2 Amonkar GP, Deshpande JR. Cardiac angiosarcoma. Cardiovascular Pathology. 2006; 15: 57-58.

3 Simpson L, Kumar SK, Okuno SH, et al. Malignant primary cardiac tumors: review of a single institution experience. Cancer. 2008; 112: 2440-2446.

4 Saraiva F, Antunes M, Providencia LA. Cardiac angiosarcoma in a pregnant woman: a case report and review of the literature. Acta Cardiologica. 2012; 67: 727-731.