

Case Report

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Extension of adrenocortical carcinoma into the right atrium – echocardiographic diagnosis: A case report

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Abstract

Background: Adrenocortical carcinoma is a rare, highly malignant tumor. Cardiac involvement of the tumor is very rare. Echocardiography facilitates the evaluation of the cardiac involvement of the tumor.

Case Presentation: We describe a patient with an adrenal tumor. Transthoracic echo showed its extension into the right atrium. Accordingly, a combined abdominal and cardiac operation was performed, monitored by transesophageal echocardiography.

Conclusion: This case highlights the importance of echocardiography in revealing the cardiac involvement by this tumor and in planning the operative procedure.

Background

Adrenocortical carcinoma is a rare and a highly malignant neoplasm [1] It grows rapidly and tends to metastasize to the liver and lungs and to invade the kidney, renal veins and the inferior vena cava [2] Dissemination of the tumor occurs in 82% of the patients with a median survival of 14.5 months [3]

Cardiac involvement of adrenal carcinoma is very rare; less than 20 cases have been described [4–15]. The tumor may extend through the inferior vena cava into the right atrium. The main therapeutic approach is adrenalectomy and extraction of the tumor from the inferior vena cava and the right atrium. This requires a precise evaluation of the extent of spread of the tumor, a meticulous pre-operative planning and a combined team including general and cardiac surgeons and a cardio-pulmonary bypass team.

Echocardiographic imaging is of paramount importance in the evaluation of this rare tumor.

Case Presentation

75 year-old woman was hospitalized because of dyspnea and bilateral leg edema. The patient had a history of liver cirrhosis. Physical examination revealed a patient without distress. Jugular venous pulse and heart sounds were normal. A soft murmur compatible with tricuspid regurgitation was heard. Ascites, splenomegaly and bilateral leg edema were noted.

Laboratory findings revealed pancytopenia, hypoalbuminemia, and elevated transaminase levels. Abdominal ultrasound revealed a cirrhotic liver with an enlarged portal vein, splenomegaly and ascites. A solid lesion inside the inferior vena cava was evident. A large lesion was seen on the right adrenal gland consistent with a tumor.

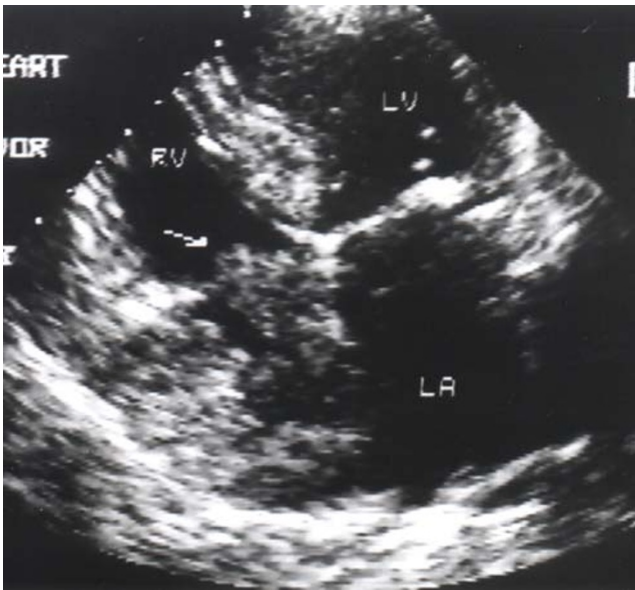


Figure 1
Apical 4 chamber view depicting a mass (arrow) within the right atrium.

Echocardiography revealed a normal size and function of the left side structures. A large uniform, echodense, immobile mass (area- 3.3 cm²), attached to the interatrial septum was seen in the right atrium (Figure 1). Doppler study revealed moderate tricuspid regurgitation. The inferior vena cava was mildly enlarged and an elongated mass attached to its wall was seen.

The patient was operated and combined cardiac and abdominal procedures were performed, monitored by transesophageal echocardiography, which did not add any additional information. She underwent right adrenalectomy and nephrectomy. Inferior vena cava and right atrium were exposed and a large mass extending from the hepatic portion of the inferior vena cava to the right atrium was removed.

Histologic examination revealed an adrenal carcinoma and a large thrombus containing malignant cells. Postoperative course was complicated by massive bleeding and coagulopathy and the patient died few hours after the operation.

Conclusions

We describe a rare case of adrenocortical carcinoma that extended to the inferior vena cava and the right atrium. Tumors that affect the right atrium include primary neoplasms and secondary tumors such as hypernephroma, hepatoma, testicular sarcoma and melanoma [16].

Adrenal carcinoma may also extend to the renal veins and the inferior vena cava, usually accompanied by a thrombus. Right atrial involvement is very rare [4–15].

Most of the described cases of adrenal carcinomas extending to the right atrium including the present case were right-sided [4,6,7,9–15]. This is explained by the direct course of the right adrenal vein to the inferior vena cava.

Other forms of cardiac involvement include infiltration of the inter-atrial septum, and a malignant pericardial effusion [8,17].

The main clinical manifestations of vena caval and right atrial extension of the tumor include peripheral edema, ascites and hepatomegaly as in our case. A patient with bouts of dyspnea and cyanosis due to a patent foramen ovale and a right atrial tumor was described [7]. Sudden death, presumably caused by right ventricular inflow obstruction was also reported [18].

Due to the tendency of the tumor to disseminate, a precise pre-operative assessment is essential. Evaluation should involve various additional modalities such as CT, [4,6–8] venography and MRI [7,19]. Echocardiography is a very useful tool in the assessment of cardiac tumors [5,7,8,10,13,16,17,19–22]. In the present case transthoracic echo identified the cardiac involvement, leading to change in the operative approach by adding cardiac surgeons to the operation.

Echocardiography can define the various patterns of cardiac extension with excellent anatomic correlations [20,21]. Transesophageal echo can provide high quality images of the inferior vena cava and right atrial involvement, and direct the surgeons in the removal of the neoplasm from both sites [8,22]. In cases in which a good quality transthoracic study is positive for cardiac involvement, transesophageal study can be performed only intraoperatively, to monitor and direct surgery in the operative arena. A patent foramen ovale should be looked for before putting the patient on by-pass.

We suggest that adrenal carcinoma, although very rare, should be included in the differential diagnosis of right atrial tumors. Once an adrenal tumor is detected, the work-up should include echocardiography, which is of a paramount importance in the evaluation of cardiac involvement, operative planning and follow-up.

Competing interests

None declared. There are no financial or other relations that could lead to a conflict of interest.

Authors' Contribution

BR drafted the manuscript. YR drafted the manuscript. DH performed the echocardiographic study and drafted the manuscript several times. All authors read and approved the final manuscript.

Abbreviations

LA – Left Atrium; LV – Left Ventricle; RV – Right Ventricle.

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Written consent was obtained from the patient's relatives for publication of study.

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