

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.

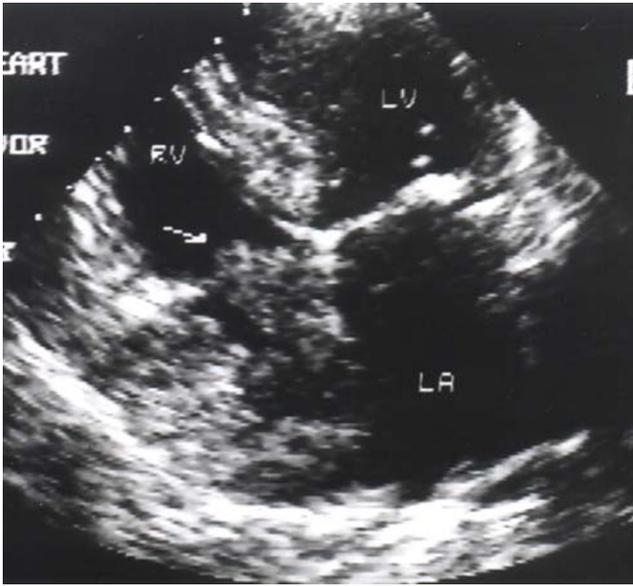


Figure 1
Apical 4 chamber view depicting a mass (arrow) within the right atrium.

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.

References

- Bruce L and McClennan D **Staging and follow-up of renal and adrenal carcinoma** *Cancer* 1991, **67**:1199-1208
- Francis IR, Smid A, Gross MD, Shapiro B, Naylor B and Glazer GM **Adrenal masses in oncologic patients: Functional and morphologic evaluation** *Radiology* 1988, **166**:353-356
- Luton J-P, Cerdas S, Billaud L, Thomas G, Guillaume B, Bertagna X, Laudat MH, Louvel A, Chapuis Y and Blondeau P **Clinical features of adrenocortical carcinoma, prognostic factors, and the effect of mitotane therapy** *N Engl J Med* 1990, **322**:1195-1201
- Schramek P, Dunser E, Bhargabha A, Hruby W and Umek H **Adrenal cortical carcinoma: Preoperative demonstration of right atrial extension by sonography and computerized tomography** *J Urol* 1985, **133**:260-262
- Lee JJ, Kupfer J, Raissi S, Geller SA and Siegel RJ **Rapid extension of left adrenocortical carcinoma into the right atrium** *J Am Soc Echocardiogr* 1998, **11**:86-88
- Godine LB, Berdon WE, Brasch RC and Leonidas JC **Adrenocortical carcinoma with extension into inferior vena cava and right atrium: Report of 3 cases in children** *Pediatr Radiol* 1990, **20**:166-168
- Friedrich MG, Dill H, Unverdorben M, Engels G, Scheele H and Bachmann K **Adrenal carcinoma with intravenous extension into the tricuspid valvular plane in a patient with patent foramen ovale** *Eur Heart J* 1994, **15**:708-709
- Bilge A, Pierre LA and Kulbertus HE **Isolated cardiac metastasis of adrenal carcinoma: Transesophageal echocardiographic features** *Am Heart J* 1996, **132**:1066-1068
- Huguet C, Caporossi M, Gavelli A, Harb J and McNamara M **Neoplastic thrombosis of the inferior vena cava involving the right atrium caused by adrenal cortical carcinoma. A new indication for vascular exclusion of the liver** *Ann Chir* 1994, **48**:364-369
- Ohnishi M, Niwayama H, Miyazawa Y, Kondon N, Imai H, Nishimoto Y, Morooka N, Watanabe S, Masuda Y and Inagaki Y **Echocardiography in patients with malignant metastatic neoplasms of the heart and great vessels** *J Cardiol* 1990, **20**:377-384
- Shahian DM, Nieh PZT and Libertino JA **Resection of atriocaval adrenal carcinoma using hypothermic circulatory arrest** *Ann Thorac Surg* 1989, **48**:421-422
- Davies RP and Lam AH **Adrenocortical neoplasm in children. Ultrasound appearance** *J Ultrasound Med* 1987, **6**:325-328
- Okazumi S, Asano T, Ryu M, Nagashima T, Odaka M, Isono K and Nishizawa T **Surgical resection of adrenal carcinoma extending into the vena cava, right atrium and ventricle: Case report and review of the literature** *Nippon Geka Gekka Zasshi* 1987, **88**:231-238
- Smith BM, Mulherin JL, Sawyers JL, Turner BI, Prager RL and Dean RH **Suprarenal vena caval occlusion. Principles of operative management** *Ann Surg* 1984, **199**:656-658
- Triebling A and Chyczewski L **Case of hormonally inactive adrenocortical cancer with metastases to the heart** *Wiad-Lek* 1982, **35**:811-814
- Weyman A **Right ventricular inflow tract** *In Principles and practice of echocardiography (Edited by: Weyman AE) Lea & Febiger, a Waverly Co. Second edition* 1994, 824-862
- Nakata A, Yagi S, Oyama K, Kida H and Sugioka G **Adrenocortical carcinoma with a giant pericardial mass** *Intern Med* 1993, **32**:438-440
- Dickens P, Poon CS and Wat MS **Sudden death associated with solitary intracavitary right atrial metastatic tumor deposit** *Forensic Sci Int* 1992, **57**:169-173
- Gindea AJ, Gentin B, Naidich DP, Freedberg RS, McCauley D and Kronzon I **Unusual cardiac metastasis in hypernephroma: The complementary role of echocardiography and magnetic resonance imaging** *Am Heart J* 1988, **116**:1359-1361
- Lestuzzi C, Biasi S, Nicolosi GL, Lodeville D, Pavan D, Collazzo R, Guindani A and Zanuttini D **Secondary neoplastic infiltration of the myocardium diagnosed by two-dimensional echocardiography in seven cases with anatomic confirmation** *J Am Coll Cardiol* 1987, **9**:439-445
- Johnson MH and Soulen RL **Echocardiography of cardiac metastases** *Am J Radiol* 1983, **141**:677-681
- Singh I, Jacobs L, Kotler MN and Ioli A **The utility of transesophageal echocardiography in the management of renal cell carcinoma with intracardiac extension** *J Am Soc Echocardiogr* 1995, **8**:245-250

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