Extension of adrenocortical carcinoma into the right atrium – echocardiographic diagnosis: A case report
Boaz Rosen¹,², Yoseph Rozenman¹,² and David Harpaz*¹,²

Address: ¹The Heart Institute, E Wolfson Medical Center, Holon, Tel Aviv, Israel and ²Sackler School of Medicine, Tel Aviv University, Tel Aviv, Israel
Email: Boaz Rosen - rosenbo_99@yahoo.com; Yoseph Rozenman - rozenman@remon-medical.co.il; David Harpaz* - dharecho@netvision.net.il
* Corresponding author

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.
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.

Acknowledgments
We would like to thank Lori Mandelzweig, MPH, for editorial assistance.

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

References
1. Bruce L and McClennan D Staging and follow-up of renal and adrenocortical carcinoma Cancer 1991, 67:1199-1208
2. 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
3. Luton J-P, Cerdas S, Billaud L, Thomas G, Guilhaume B, Bertagna X, Laudat MH, Louvel A, Chapuis Y and Biondau P Clinical features of adrenocortical carcinoma, prognostic factors, and the effect of mitotane therapy N Engl J Med 1990, 322:1195-1201
4. 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 Ural 1985, 133:260-262
5. 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
6. 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
7. 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
8. Bilge A, Pierre LA and Kulbertus HE Isolated cardiac metastasis of adrenocortical carcinoma: Transesophageal echocardiographic features Am Heart J 1996, 132:1066-1068
9. 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
10. 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 metastastic neoplasms of the heart and great vessels J Cardio 1990, 20:377-384
11. Shahian DM, Nieh PZT and Libertino JA Resection of atrio caval adrenocortical carcinoma using hypothermic circulatory arrest Ann thorac Surg 1989, 48:421-422
12. Davies RP and Lam AH Adrenocortical neoplasm in children. Ultrasound appearance J Ultrasound Med 1987, 6:325-328
13. 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 Gekkipai Zasshi 1987, 88:231-238
14. Smith BM, Mulherin JL, Sawyers JL, Turner BL, Prager RL and Dean RH Suprarenal vena cava occlusion. Principles of operative management Ann Surg 1984, 199:656-658
15. Triebeling A and Chyczewski L Case of hormonally inactive adrenocortical cancer with metastases to the heart Wiad-Lek 1982, 35:811-814
16. Weyman A Right ventricular infow tract In Principles and practice of echocardiography (Edited by: Weyman AE) Lea & Febiger, a Waverly Ca. Second edition 1994, 824-862
17. Nakata A, Yagi S, Oyama K, Kida H and Sugiioka G Adrenocortical carcinoma with a giant pericardial mass Intern Med 1993, 32:438-440
18. Dickens P, Poon CS and Was MS Sudden death associated with solitary intracavitary right atrial metastatic tumor deposit Forensic Sci Int 1992, 57:169-173
19. Gindea AJ, Gentin B, Naiddih 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
20. 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
21. Johnson MH and Soulen RL Echocardiography of cardiac metastases Am J Radiol 1983, 141:677-681
22. Singh I, Jacobs L, Kottler 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

http://www.cardiovascularultrasound.com/content/1/1/5

Publish with BioMed Central and every scientist can read your work free of charge

"BioMed Central will be the most significant development for disseminating the results of biomedical research in our lifetime."
Sir Paul Nurse, Cancer Research UK

Your research papers will be:
• available free of charge to the entire biomedical community
• peer reviewed and published immediately upon acceptance
• cited in PubMed and archived on PubMed Central
• yours — you keep the copyright