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Case Report

A case of adrenal cortical carcinoma with inferior vena cava thrombus extending into the right atrium: a two staged procedure

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ABSTRACT

We present a case of adrenal cortical carcinoma (ACC) with tumor thrombus involving the inferior vena cava (IVC) and right atrium in a morbidly obese, middle-aged female. ACC is a rare type of cancer with a poor outcome. Most cases present with metastasis at the time of initial presentation. This patient presented with breathing difficulty and flank pain. With endocrinology evaluation and collaborative effort of multidisciplinary teams, the patient successfully underwent staged procedures of atrial clot evacuation after thoracotomy with cardiopulmonary bypass (CBP) along with inferior vena cava thrombectomy (IVCT) and left radical nephroadrenalectomy. The two staged procedure reduces the mortality when compared with a single staged procedure. The patient underwent atrial clot evacuation, performed by cardiothoracic surgery team under CBP and deployment of IVC filter by interventional radiologist as a first staged procedure. At six months of follow-up, the patient had locoregional disease spread but with good functional status. This case report highlights that even a high-volume disease, with proper planning by an experienced surgical team, can be operated successfully with an acceptable post-surgery quality of life for the patients.

Keywords: Adrenal cortical carcinoma, Atrial clot evacuation, Inferior venacava thrombectomy, Nephroadrenalectomy

INTRODUCTION

Adrenal cortical carcinoma (ACC) is a very unusual type of cancer that affects 0.7 to 2 persons per million populations. The poor prognosis, aggressive characteristics, and possible recurrence warrant investigations, meticulous staging, and surgical resection with follow up and possible chemotherapy or radiotherapy.[1-3] Most patients may present with symptoms arising from metastasis. Even with complete resection, survival is usually less than a year. We present a case of adrenal cortical carcinoma (ACC) with metastasis involving left Kidney and extensive tumor thrombus involving inferior vena cava (IVC) and extending up to the right atrium.

CASE REPORT

A 34-year-old known diabetic, hypertensive, morbidly obese lady came to our outpatient with complaints of breathlessness on exertion and left flank pain. An endocrine evaluation was done. Pheochromocytoma and functional adrenal tumors were ruled out with metabolic workup. CECT Abdomen and Chest revealed a large, extensively vascular left adrenal tumor (8×7×6 cms) with infiltration into the upper left kidney and with tumor thrombus in inferior vena cava (IVC), right atrium, and in the pulmonary artery branches. No IVC wall enhancement was seen (Figure 1 A-C).

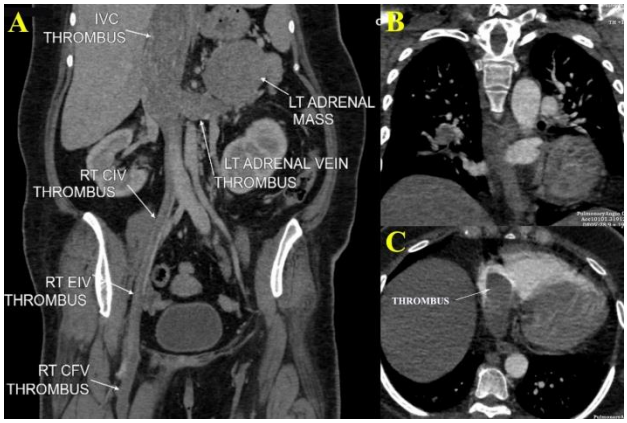

Figure 1: (A) Contrast enhanced CT of abdomen and chest showing left adrenal tumor, tumor thrombus in IVC and bland thrombus in right external iliac vein and right common femoral vein; CECT chest showing tumor thrombus (B) in right atrium and (C) pulmonary artery.

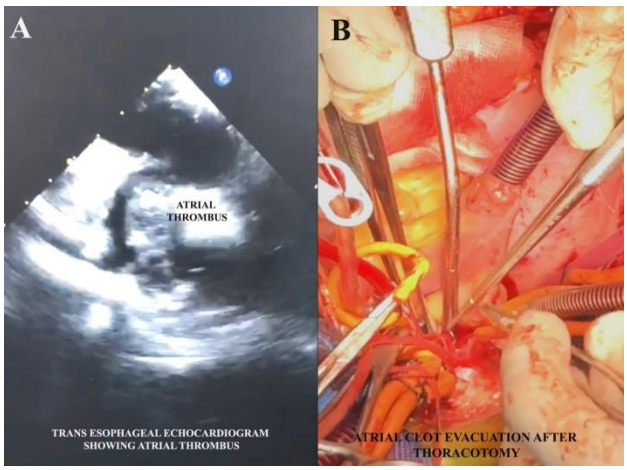

Figure 2: (A) Intraoperative transesophageal echocardiogram showing thrombus in right atrium. (B) Thoracotomy and atrial clot evacuation.

Cardiothoracic and vascular surgeon's opinion was obtained and mortality risk of 30-40% was explained. Informed, written consent was obtained for the surgery. She successfully underwent thoracotomy, atrial clot evacuation and supradiaphragmatic tumor clot evacuation from IVC under CBP and Intervention radiologist deployed IVC filter (Figure 2). She was managed in the intensive care unit (ICU) for 2 days and she underwent open left Radical Nephroadrenalectomy with IVCT on POD-2. (Figure 3) The entire procedure was uneventful. She was managed in ICU till postoperative day POD-5. Enhanced recovery after surgery protocol was followed. The bilateral drains were removed on POD-4 and 5. Foley catheter was removed on POD-9. Histopathology report suggested ACC (pT4 Nx Mx). Tumor embolus from the right atrium was consistent with malignant epithelioid neoplasm with lymphovascular invasion. The patient was discharged on POD-10 and was started on chemotherapy after obtaining a medical oncologist's opinion. Follow up PET CT at 3 months and 6 months revealed hypermetabolic mediastinal and lung nodes and hypermetabolic lesions in right perinephric region with mild pleural effusion. She is under medical oncologist care till the current date.

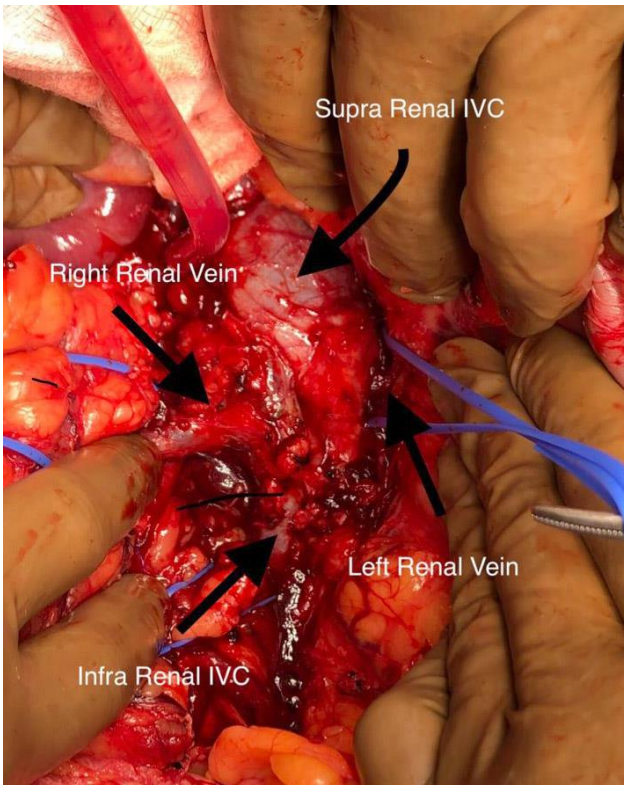

Figure 3: Control achieved while doing left nephroadrenalectomy.

DISCUSSION

ACCs are rare, highly hostile cancers, with a poor prognostic outcome with overall 5-year survival of less than 15%.[1] In a population of 1–2 million people, 0.7 to 2 cases of ACCs have been recognized.[2,3] The bimodal age distribution is seen (< 5 years and between 40-50 years), with female preponderance (F:M=1.5:1). Around 50% of ACCs are functional tumors producing excess hormone (cortisol, androgens, aldosterone, estrogen) and may present with Cushing's syndrome, severe hypertension, or refractory hypokalemia. Women may present with oligomenorrhea, acne, hirsutism, or virilization. High suspicion of ACC is required if symptoms progress rapidly. Some patients present with symptoms of the mass itself such as pain. Few are asymptomatic. Initial workup should be directed towards the stratification of functional, non-functional, and malignant tumors. Peri and postoperative management depend on the functional status of the tumor.[4] Blood workup includes plasma cortisol, basal ACTH, 24-h urinary free cortisol, serum potassium, aldosterone: renin ratio, 17-OH-progesterone, androstenedione, and testosterone. checking blood pressure, evaluating postural hypotension is also important.[3] computed tomography (CT) and magnetic

resonance imaging (MRI) can characterize the tumors, but extra-adrenal metastasis is often difficult to be differentiated by them, warranting 18F-FDG PET. ACC is more probable in cases of local invasion, usually involving the kidneys, IVC, or lymph nodes, which is better evaluated by MRI. The European Network for the study of adrenal tumors and the TNM classification are used for disease stratification.[2,3] For resectable, invasive ACCs, open surgery is preferred where collateral resection of invaded organs has to be done. Since our patient had IVC and right atrial thrombus involvement, atrial clot evacuation and supra-diaphragmatic IVCT were done followed by open nephroadrenalectomy with infra-diaphragmatic IVCT as staged procedures. Laparoscopic adrenalectomy (LA) is reserved for small adrenal tumors. In a margin negative, localized ACC, LA is as successful as open adrenalectomy. A multivariate analytical study states localized lymphadenectomy improved prognosis.[5] Recurrence is always a possibility that is closely followed-ups with CT, MRI, or PET scans for which reoperation or chemotherapy can be given. In cases where radical surgical resection is not possible, chemotherapy can be beneficial.[1,3] Radiotherapy is used as an adjuvant or as palliation in cases of unresectable tumors.[6] The origin of the tumor and definitive diagnosis are obtained by histopathology and immunohisto-chemistry. Robotic adrenalectomy is useful in larger tumors, partial or bilateral adrenalectomies, and in truncal paragangliomas.[7] Molecular analyses such as nuclear transcription factor, steroidogenic factor-1 will aid in diagnosis soon.[1,3]

CONCLUSION

ACCs are rare and robust prospective studies are the need of the hour. This case is presented to show that with early investigations, a patient-tailored approach and proper follow up, even such a difficult case with grave prognosis can have a decent quality of life and better overall survival.

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REFERENCES

1. Fassnacht M, Terzolo M, Allolio B, Baudin E, Haak H, Berruti A, et al. Combination chemotherapy in advanced adrenocortical carcinoma. N Engl J Med. 2012;366(23):2189-97.
2. Langenhuijsen J, Birtle A, Klatte T, Porpiglia F, Timsit MO. Surgical management of adrenocortical carcinoma: Impact of Laparoscopic Approach, Lymphadenectomy, and Surgical Volume on Outcomes- A Systematic Review and Meta-analysis of the Current Literature. Euro. Urol Focus. 2016;1(3):241-50.
3. Zini L, Porpiglia F, Fassnacht M. Contemporary management of adrenocortical carcinoma. Euro Uro. 2011;60(5):1055-65.
   Mayr R, Pycha A, Burger M. The perioperative management of metabolically active tumors. Euro Uro Focus. 2016;1(3):258–60.
4. Reibetanz J, Jurowich C, Erdogan I, Nies C, Rayes N, Dralle H, et al. Impact of lymphadenectomy on the oncologic outcome of patients with adreno-cortical carcinoma. Ann Surg. 2012;255(2):363-9.
5. Polat B, Fassnacht M, Pfreundner L, Guckenberger M, Bratengeier K, Johanssen S, et al. Radiotherapy in adrenocortical carcinoma. Canc. 2009;115(13):2816-23.
6. Nomine-Criqui C, Germain A, Ayav A, Bresler L, Brunaud L. Robot-assisted adrenalectomy: indi-cations and drawbacks. Upd Surg. 2017;69(2):127-33.

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