Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2015; 3(10A):998-1001 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources) DOI: 10.36347/sjmcr.2015.v03i10.022

A Rare Presentation of Adrenocortical Carcinoma-Spontaneous Rupture with Sub capsular Splenic Hematoma

Rajeev T.P[1], Nitin Gupta[2], Debanga Sarma[3], Sasanka K. Barua[4]
[1]Proffesor & Head, [2]Resident Urology, [3]Assistant Professor, [4]Associate Professor, Department Of Urology, GMCH, Guwahati, Assam, India

***Corresponding author**
Dr. Rajeev T.P
Email: urohodgmch@gmail.com

Abstract: Adrenocortical carcinoma (ACC) is a rare endocrine malignancy with an incidence rate of 1 to 2 per million. Clinical symptoms are mainly related to an excess of steroid hormones or tumor bulk. The most common metastatic sites are the liver, lungs, lymph nodes and bone. Surgical resection remains the cornerstone of treatment. The case mentioned here has two rare characteristics. First that spontaneous rupture of ACC is very rare and secondly associated splenic sub capsular hematoma has never been reported. A 63 year old non-hypertensive male presented in the emergency department with the complaints of pain and gradually increasing lump in the left side of abdomen since last 3 months. On initial clinical examination he was diagnosed to have splenomegaly by emergency medical officer. USG showed to have splenomegaly and left suprarenal mass lesion for which he was referred to urology department. CECT of abdomen revealed heterogeneously enhancing left adrenal ruptured mass lesion with involvement of left kidney and large splenic sub capsular hematoma (13.8x3.2x14.8cms). CEMR findings correlated to CECT. Serum cortisol and urinary metanephrines were normal. Patient was planned for surgery and intraoperatively was found to have locally extensive adrenal tumor with large sub capsular splenic hematoma with left kidney involvement. Adrenalectomy, nephrectomy and splenectomy was done with the palliative intent which relieved patient symptomatically. HPE of specimen showed it to be adrenocortical carcinoma. Though spontaneous rupture of pheochromocytoma and cysts is reported in literature, to our knowledge this is the first reported case of spontaneous adrenocortical carcinoma rupture with sub capsular splenic hematoma.
Keywords: Adrenocortical carcinoma, Spontaneous rupture, splenic hematoma, Adrenalectomy, Nephrectomy.

INTRODUCTION

Adrenocortical carcinoma (ACC) is a rare endocrine malignancy with an incidence rate of 1 to 2 cases per million person-years. It has a poor prognosis and can be classified as either a functional or nonfunctional tumor [1-3]. It most commonly occurs in two age groups: children less than 5 years age and adults from 40–59 years, and is twice as common in women [1-6]. Most ACC cases are sporadic. However, familial neoplasms also exist, most commonly associated with multiple endocrine neoplasia type 1 (MEN-1), Li-Fraumeni syndrome, Beckwith-Wiedemann syndrome and Carney complex [1, 6].

Clinical symptoms are related to hormonal dysfunction, which occurs in 60–80% of cases. Most commonly it is glucocorticoid or both glucocorticoid and androgen hyper secretion [1, 4, 6]. Glucocorticoid-secreting adrenocortical tumors are responsible for the minority (10–15%) of endogenous Cushing syndrome [7].

ACCs are generally large tumors, with an average diameter of 5–20 cm and mass of 1000 Gms [1, 2, 6]. Most commonly they are diagnosed as an advanced disease (stage IV), with tumor mass extending beyond the adrenal gland and metastases (most commonly lungs, liver or lymph nodes) [1,6,8]. Spontaneous rupture of adrenocortical carcinoma is very rare. We here present a case of spontaneous rupture of adrenocortical carcinoma with sub capsular splenic hematoma.

CASE REPORT

A 63 year old non-hypertensive male presented in the emergency department with the complaints of pain and gradually increasing lump in the left side of abdomen since last 3 months. Initially the symptoms were mild and non bothersome to the patient, which later became severe, and patient consulted with his family physician but was not relieved on medications and patient came to emergency department for further management.

There was no history of trauma. On initial clinical examination pallor was present, heart rate was 84/mins, BP was 136/84 mm Hg and he was diagnosed to have splenomegaly by emergency medical officer. USG showed to have splenomegaly and left suprarenal mass lesion for which he was referred to urology department. CECT of abdomen revealed heterogeneously enhancing left adrenal ruptured mass lesion with involvement of left kidney and adjacent organs and large splenic sub capsular hematoma of approximately 13.8 x 3.2 x 14.8 cms (Figure 1). Contrast enhanced MRI findings showed ill defined heterogeneously enhancing mass lesion invading adjacent organs with large sub capsular hematoma (Figure 2).

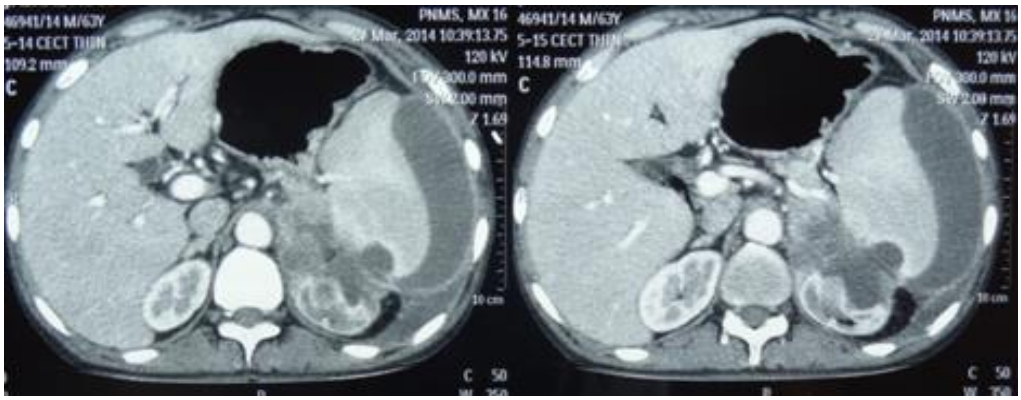

Fig 1: CECT – Ruptured adrenal tumor with sub capsular hematoma

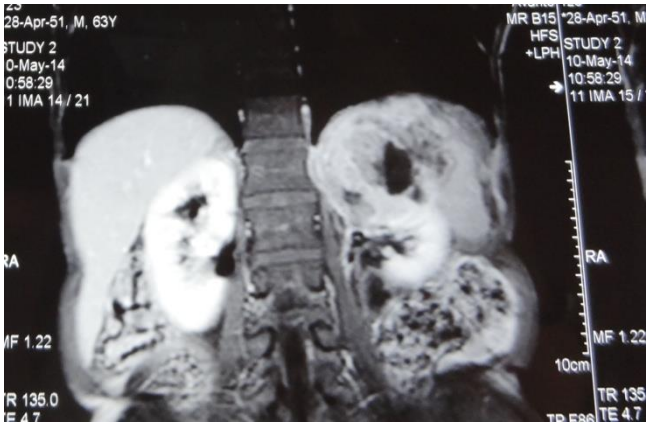

Fig 2: Contrast enhanced MRI

Total leucocyte count was 7.92 x 1000, hemoglobin was 7.1 g/dl, platelet count was 173 x 1000 and differential count was normal. His renal function tests, liver function tests and chest X ray were normal. Serum cortisol and 24 hour urinary metanephrines were done to evaluate the functional status of the lesion and both were within normal limits. (10.7 micrograms/deciliter and 47.9 micrograms/liter respectively). As patients' symptoms were severe and he was not relieved of pain even on opioid analgesics, he was planned for emergency surgery and intraoperatively was found to have ill defined ruptured locally extensive large adrenal tumor with large sub capsular splenic hematoma with left kidney involvement (Figure 3). After explaining the condition to attendants, adrenalectomy, left nephrectomy and splenectomy were done with the palliative intent. Post operative period was uneventful and patient was relieved symptomatically. HPE of specimen showed it to be adrenocortical carcinoma (Figure 4). Patient was planned for adjuvant therapy which he denied. He was on follow up for 6 months without any symptoms after which he was lost to follow up.

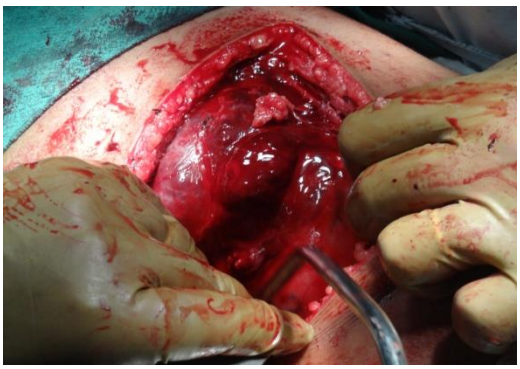

Fig 3: Intraoperative image – Ruptured adrenal tumor

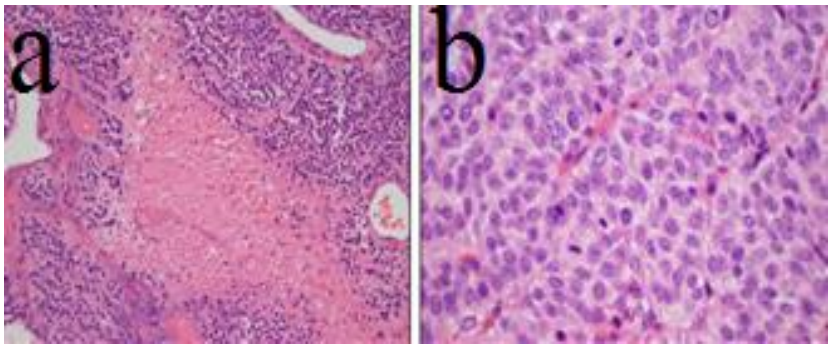

Fig 4: Adrenal cortical carcinoma. (a) Central necrosis within the tumor tissue. (b) ACC cells with prominent nuclei and nucleoli

DISCUSSION

Adrenocortical carcinoma (ACC) is a rare malignancy with poor prognosis. Patients usually present with abdominal pain or with symptoms related to the mass effect or hormonal activity of the tumor, mainly steroids. The most common metastatic sites are the liver, lungs, lymph nodes and bone. Surgical resection remains the cornerstone of treatment. Spontaneous rupture of adrenocortical carcinoma is very rare. Lu CY *et al.;* in their review on spontaneous rupture of adrenocortical carcinoma reported that only 5 such cases were reported in the literature and theirs was the sixth reported case that is not related to any preceding traumatic incidents or predisposing disease. They also reported that tumor size greater than 10 cm in adults is a predisposing factor for spontaneous rupture [9]. Other adrenal lesions reported in the literature for spontaneous rupture are pheochromocytoma and pseudo cyst [10, 11].

CONCLUSION

Adrenocortical carcinoma is a rare malignancy with aggressive behavior. Though spontaneous rupture of pheochromocytoma, adrenal cysts and angiomyolipoma is reported in literature, to our knowledge this is the first reported case of spontaneous rupture of adrenocortical carcinoma with sub capsular splenic hematoma.

REFERENCES

1. Weiss LM, Bertagna X, Chrousos GP, Kawashima A, Kleihues P, Koch CA, *et al.;* Adrenal Cortical Carcinoma. In Pathology and Genetics of Tumors of Endocrine Organs. Edited by DeLelils RA. Lyon: IARC; 2004:139–142.
2. Icard P, Goudet P, Charpenay C, Andreassian B, Carnaille B, Chapuis Y, *et al.;* Adrenocortical carcinomas: surgical trends and results of a 253-patient series from the French association of endocrine surgeons study group. World J Surg 2001; 7:891–897.
3. Sasaki K, Desimone M, Rao HR, Huang GJ, Seethala RR; Adrenocortical carcinosarcoma: a case report and review of the literature. Diagn Pathol 2010, 5:51.
4. Luton JP, Cerdas S, Billaud L, Thomas G, Guilhaume B, Bertagna X, *et al.;* Clinical-features of adrenocortical carcinoma, prognostic factors, and the effect of mitotane therapy. New Engl J Med 1990, 17:1195–1201.
5. Wajchenberg BL, Pereira MAA, Medonca BB, Latronico AC, Carneiro PC, Alves VAF, *et.al.;* Adrenocortical carcinoma - clinical and laboratory observations. Cancer 2000, 4:711–736.

6. Lafemina J, Brennan MF: Adrenocortical carcinoma: past, present, and future. J Surg Oncol 2012, 5:586–594.
7. Shahani S, Nudelman RJ, Nalini R, Kim HS, Samson SL; Ectopic corticotropin-releasing hormone (CRH) syndrome from metastatic small cell carcinoma: a case report and review of the literature. Diagn Pathol 2010, 5:56.
8. Correa P, Chen VW; Endocrine gland cancer. Cancer 1995, 1:338–352.
9. Lu Cy, Yen TH, Hsieh ML, Chen Y; Spontaneous rupture of adrenocortical carcinoma: a coincidence or a tendency? Clinical nephrology, 2009; 72(2): 147-150.
10. Hanna JS, Spencer PJ, Savopoulou C, Kwasnik E, Askari R; Spontaneous adrenal pheochromocytoma rupture complicated by intraperitoneal hemorrhage and shock. World Journal of Emergency Surgery 2011 6:27.
11. Rahim Mahmodlou, Neda Valizadeh; Spontaneous rupture and hemorrhage of adrenal pseudocyst presenting with acute abdomen and shock. Iran J Med Sci. 2011; 36(4): 311–314.