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*International Journal of Recent Scientific Research*
*Vol. 6, Issue, 2, pp. 2743-2745, February, 2015*

*International Journal of Recent Scientific Research*

ISSN: 0976-3031

RESEARCH ARTICLE

# MALIGNANT PHEOCHROMOCYTOMA PRESENTED WITH SPONTANEOUS RUPTURE AND HEMORRHAGE

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**ARTICLE INFO**

***Article History:***

Received 14th, January, 2015
Received in revised form 23th, January, 2015
Accepted 13th, February, 2015
Published online 28th, February, 2015

***Key words:***

Adrenal Pheochromocytoma, hemorrhage, adrenal tumor

**ABSTRACT**

Pheochromocytoma classically presents with the triad of paroxysmal headaches, palpitations and diaphoresis accompanied by hypertension. Rarely do they present with spontaneous rupture, hemorrhage and abdominal catastrophe. If unrecognized this may lead to death very rapidly. In our case 40 year male patient presented with pain in abdomen since 2 days which was sudden in onset and increasing in intensity. The patient is hypertensive and diabetic since 5 years and is on medication. On examination the patient had sweating, tachycardia, hypotension, abdominal tenderness and guarding. Computed Tomography scan revealed a large right adrenal mass with fluid collection in right peri-anterior paranerphric spaces, perimass and right para retrocolic space.

On X-ray chest a solitary pulmonary nodule in right middle zone (? metastases) was seen. Mass was operated. The histopathological examination showed features of malignant pheochromocytoma with hemorrhage and capsular destruction. We are presenting this case for its clinical, radiological and histopathological features, as malignant pheochromocytoma presenting as rupture is extremely rare.



## INTRODUCTION

Pheochromocytoma meaning dusky brown tumor is generally benign tumor arising from the adrenal medulla. Adrenal pheochromocytomas are uncommon catecholamine secreting tumors. The prevalence of pheochromocytoma in patients with hypertension is 0.1-0.6% (Omura M *et al*, 2004).Most pheochromocytomas are slow growing and benign, but 5-10 % of the tumors are malignant, invasive and metastasizing. Spontaneous rupture of adrenal pheochromocytoma is extremely rare, with only about 50 cases reported in literature (Kobayashi T *et al*, 2005)

### Case Presentation

A 40 year old male patient presented with pain in abdomen since 2 days which was sudden in onset and increasing in intensity. The patient took treatment for the same outside but his complaints worsened so he was referred to our emergency department. The patient is a known hypertensive and diabetic since 5 years and was on medications. On clinical examination the patient had tachycardia, hypotension and sweating. Abdominal tenderness and guarding was present. His urinary Vanillyl Mandelic Acid levels were raised (13.1 mg). His complete blood counts, liver function tests were normal. His Computed Tomography scan revealed a large adrenal mass with fluid collection in right peri- anterior paranerphric spaces, perimass, right para retrocolic space. Other adrenal (left), liver, thyroid showed no evidence of tumor. His chest X-ray showed a solitary pulmonary nodule in right mid zone. There was no significant family history.

The mass was surgically resected and the patient's vitals were managed with required blood transfusion, fluids and supportive care. The post operative period of the patient was uneventful. Resected specimen was sent to our department for histopathological examination.

### Gross

Received right suprarenal mass measuring 8.5x6x4.5 cm with irregular destruction of capsule covered with large blood clot and friable grey brown material (figure1). On cut section it showed irregular dark grey brownish, friable mass invading and destroying capsule, with extensive areas of hemorrhage and necrosis.

### Microscopy

Multiple sections showed adrenal tissue with tumor composed of neoplastic cells arranged in nests, diffuse sheets, clusters and Zell ballen appearance (figure2). Tumor cells showed moderate to high nuclear

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plemorphism and having eosinophilic granular cytoplasm (figure3). Extensive areas of hemorrhage, necrosis, high mitotic activity, with increased cellularity, capsular and vascular invasion was noted. This final diagnosis of malignant pheochromocytoma with rupture was made.

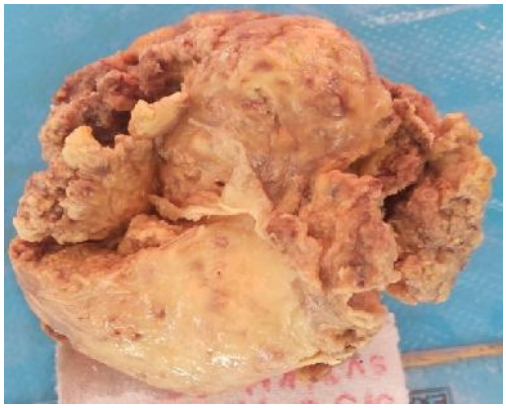

**Figure 1** gross photograph showing adrenal mass with rupture

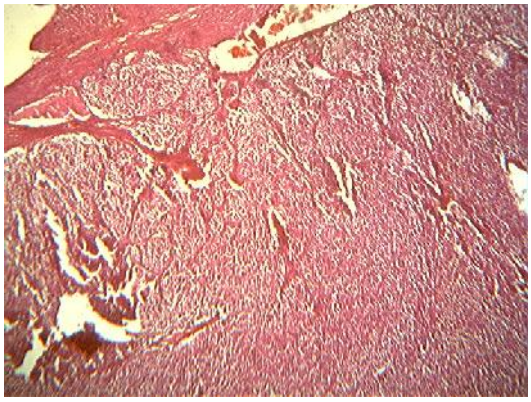

**Figure 2** Photomicrograph showing pheochromocytoma with neoplastic cells arranged in nests, diffuse sheets, clusters and Zell ballen appearance(H&E stain, 40X)

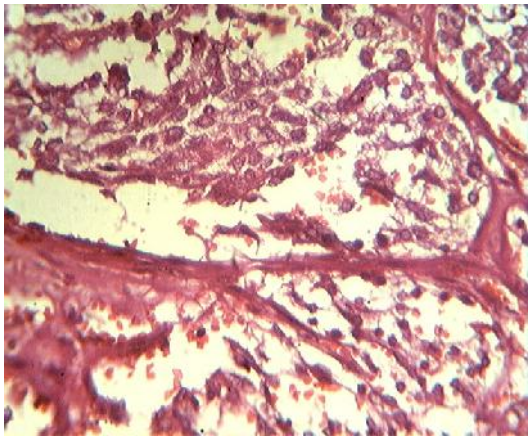

**Figure 3** microphotograph by Photomicrograph, high power view(H & E stain, 400X)

## DISCUSSION

Adrenal pheochromocytoma are rare catecholamine secreting yellowish brown tumors composed of chromaffin cells derived from embryonic neural crest cells. They are traditionally referred to as 10% tumors (10% being bilateral, malignant, extra-adrenal, in children, hereditary). The average annual incidence rate of pheochromocytoma was approximately 2- 8 per million population (Stenson G *et al*,1986), with an average age of diagnosis being fifth decade of life, with roughly equal sex distribution.

Clinically patient presents usually with triad of paroxysmal headaches, palpitation and diahopresis accompanied with marked hypertension. In addition to these symptoms pallor, flushing, sweating, anxiety, chest or abdominal pain, blurring of vision and transient electrocardiographic changes may be seen (Adler J *et al*, 2008).

Our case presented with sudden onset of abdominal pain, hypertension and tachycardia which gradually went on worsening. On radiological evaluation it was found to be right adrenal mass with hemorrhage and rupture which was managed with emergency with all medical and surgical help.

Spontaneous hemorrhage within pheochromocytoma resulting in capsular rupture and retroperitoneal or intra abdominal hemorrhage is extremely rare with only 50 documented cases in the literature between 1944-2010 (Kobayashi T *et al*, 2005 & Mohamed HA *et al* 2003)

The exact mechanism of rupture is unknown but high blood pressure attributable to massive release of catecholamine into the circulation is probably associated with vasoconstriction in the tumor and subsequent necrosis, hemorrhage and increased intracapsular pressure. Also rapid tumor growth may contribute to increased intracapsular pressure and tear.

On gross pathology pheochromocytoma typically presents as solitary , unilateral and multicentric mass that is often sharply circumscribed. In larger tumors there may be degenerative, necrotic, cystic or hemorrhagic changes.

On microscopy variety of architectural pattern can be seen. The most common is nesting and anastomosing trabecular pattern. In our case it was malignant pheochromocytoma with capsular rupture and large hemorrhages. On radiological evaluation there was evidence of metastasis in right lung. The patient was adequately managed with all required medical, supportive and surgical treatment.

**Summary**

Malignant pheochromocytoma presenting with hemorrhage and rupture is extremely rare. With proper clinical and radiological evaluation, if promptly diagnosed and managed it will be life saving for the patient.

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**How to cite this article:**

Jagtap SV *et al*. Malignant pheochromocytoma presented with spontaneous rupture and hemorrhage*International Journal of Recent Scientific Research Vol. 6, Issue, 2, pp. 2743-2745, February, 2015*

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