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A case report of an atypical presentation of a case of pheochromocytoma

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Abstract: *Despite all technical progress in modern diagnostic methods and treatment modalities of pheochromocytoma, early consideration of the presence of these tumors remains the pivotal link towards the best possible outcome for patients. A timely diagnosis and proper treatment can prevent the wide variety of potentially catastrophic cardiovascular and other complications. Here is presented the case report of a patient with an atypical presentation of the condition.*

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I. Introduction

Some of the most common causes for palpitations include cardiac conditions such as mitral valve prolapse and atrial fibrillation, stimulants such as caffeine and alcohol, certain over the counter medications, pregnancy and around the peri menopausal period.[1] Certain of the causes for menstrual irregularities along with pain abdomen include pelvic inflammatory disease, endometriosis and ovarian cysts.[2] Pregnancy is one of the only condition where above three mentioned symptoms, i.e. palpitations, amenorrhoea and pain abdomen are seen together. Pheochromocytoma is a tumor of the adrenal medulla, presenting with at least three of the four symptoms of headache, palpitations, diaphoresis and hypertension.[3] It is uncommon for the condition to present with pain abdomen and menstrual abnormalities.

II. Case Report

An eighteen-year-old female patient presented with complaints of pain abdomen for 6 months, amenorrhoea. Patient also gave history of occasional spells of palpitations since a period of eight years. Patient attained her menarche at the age of sixteen years and has had only two menstrual cycles since then. Pain was localised in the right lower abdomen, on and off(episodic), dull aching type. Patient also gives history of palpitations since last 8 years, initially occurring in intervals of around 5-6 months, with frequency progressively increasing. Ultrasound of the abdomen showed a solid-cystic mass above the right kidney, probably arising from the right suprarenal and displacing the inferior vena cava anteromedially. Furthermore, a contrast CT of the abdomen showed similar findings with a 9.3x4.5x11.2 cm solid cystic lesion in the suprarenal location with early heterogenous enhancement in the arterial phase with maximum enhancement in the portal phase with delayed washout. The cystic component showed septations within. The right suprarenal gland could not be visualised separately. 24-hour urine catecholamine measurement revealed markedly elevated epinephrine and norepinephrine levels. Therefore, under the provisional diagnosis of pheochromocytoma, medicine, surgical gastroenterology and endocrinology consultations were sought and the patient was started on alpha and beta blocker drugs. After optimization, the patient was taken up for open right adrenalectomy. Intraoperatively, a mass arising from the right suprarenal was noted, measuring 14x12x10cm, adherent to the inferior surface of the liver and abutting the inferior vena cava. The mass was dissected out after ligation of multiple feeding vessels and hemostasis was ensured. The mass had an appearance as shown. Post-operative period was uneventful. Histopathology showed ZellBallen pattern of cells, hyaline globules, nuclear smudging and lipid degeneration, hence suggestive of a diagnosis of pheochromocytoma.

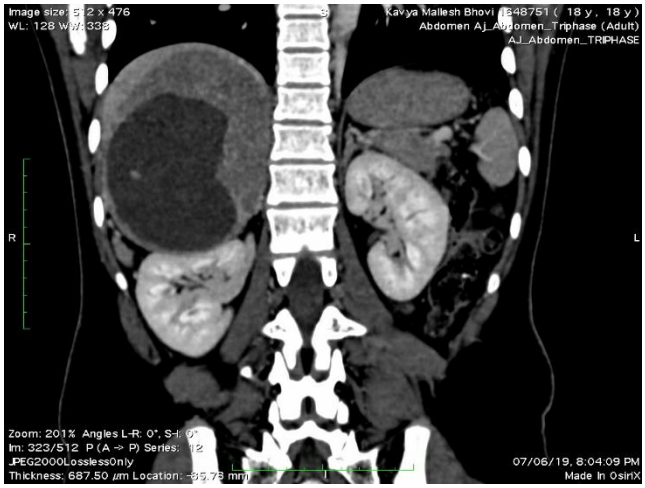

Fig.1: CT scan image showing solid cystic lesion in the suprarenal region with maximum enhancement in the venous phase.

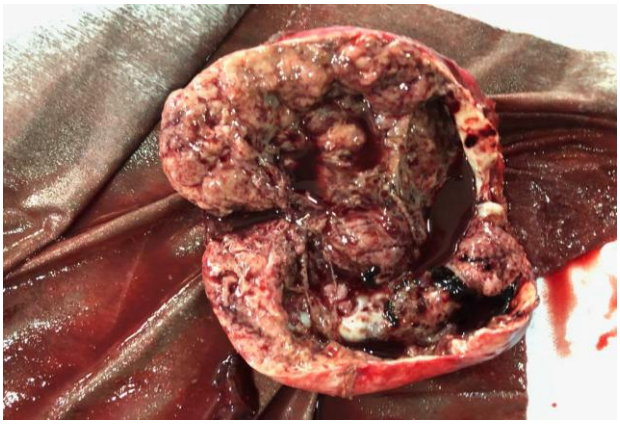

Fig.2: Surgically resected pheochromocytoma

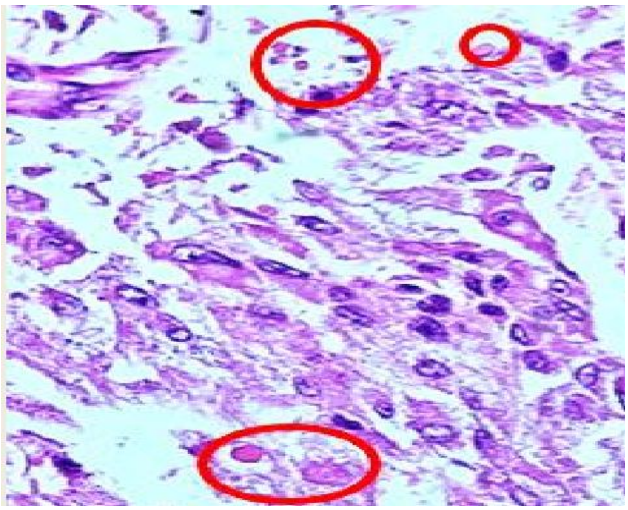

Fig.3: HPE showing ZellBallen pattern of cells, hyaline globules, nuclear smudging and lipid degeneration

III. Discussion

Pheochromocytoma is a rare tumor of the chromaffin cells of the adrenal gland with excessive catecholamine secretion causing symptoms. Classical symptoms of pheochromocytoma include accelerated hypertension, headache, palpitations, and diaphoresis.[4] A high index of suspicion is necessary to not miss a diagnosis of pheochromocytoma. Atypical presentation is seen in around 10 percent of the cases, and can take various forms. Cerebral ischemia leading to neurological features is a rare manifestation of pheochromocytoma, causes being severe hypertension with deranged cerebrovascular autoregulation, catecholamine-induced

vasospasm of cerebral vessels or cardiomyopathy, and embolization from a left ventricular thrombus.[5] There are very few case reports of pheochromocytoma presenting with menstrual disturbances.[6] There exist case reports of patients presenting with atypical features such as refractory anxiety and panic disorder.[7] Hence the need for a surgeon or physician to keep in mind the possibility of this particular disorder and be aware of its possible presentations.

IV. Conclusion

Pheochromocytoma is a rare disease but with high mortality if it is not being diagnosed early. Surgical removal is curative in ninety percent of the patients. The possibility of long term cure in the majority- if diagnosed early and treated surgically, and lethal consequences if untreated or detected late merits the need for high index of suspicion for this diagnosis. Also, this places emphasis on the need for the knowledge of atypical presentations of the disease.

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