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Case Report

Unusual clinical presentation and complications following preoperative embolization of a large adrenal tumor

Puneet Garg, Ankita Aggarwal, Venkatram Krishnan*, Sunil Kumar Bajaj, Ritu Nair Misra

Department of Radiodiagnosis, Vardhaman Mahavir Medical College and Safdarjung Hospital, New Delhi, India

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***Correspondence:**
Dr. Venkatram Krishnan,
E-mail: kvktram@gmail.com

ABSTRACT

Authors present a case of a young female presenting with secondary amenorrhea which on further investigation revealed a large intra-abdominal mass, likely arising from the left adrenal gland. As the tumor was highly vascular with large feeders, she was referred for pre-operative embolization to reduce the blood loss during surgery. Post embolization, the patient suffered from an unusual complication of tumor rupture along with excessive secretion of catecholamines resulting in myocarditis and myocardial infarction. Patient ultimately died of the myocardial infarction. Preoperative embolization of a large, hypervascular adrenal mass lesion is not devoid of unusual complications like tumor rupture and subsequent cardiovascular complications even if the tumor is hormonally inactive. This complication is extremely rare and has never been reported in adrenal tumors after embolization.

Keywords: Catecholamines**,** Embolization, Myocardial infarction, Pheochromocytoma, Rupture

INTRODUCTION

Pheochromocytomas are tumors arising from chromaffin cells of adrenal or extra-adrenal sites. They are typically hyper vascular and show large arterial feeders. They may clinically present with a wide variety of signs and symptoms based on their biochemical activity.[1] Several variations in imaging appearance can also be seen.[2] Preoperative embolization is an endovascular procedure commonly performed in order to reduce risk of excessive intraoperative blood loss that is usually associated with minimal risk of complications, especially if the tumor does not show clinical or biochemical evidence of hormone production.[3] Here author present a case of preoperative embolization of a large adrenal pheochromocytoma which did not show any evidence of hormone production during the initial work up of the patient. However, post procedure author encountered the unusual complication of tumoral rupture with possible catecholamine-induced myocardial infarction and cardiogenic shock which was totally unexpected considering the absence of any evidence of hormone production by the tumor.

CASE REPORT

Authors present a case of 22-year-old female patient who presented to the hospital with chief complaints of secondary amenorrhea for the past eight months. Routine gynecological examination, hormonal assays and hysterosalpingography were within normal limits. Patient also underwent ultrasonography of the abdomen which revealed a normal uterus and bilateral ovaries; however, there was a large mass on the left side of the abdomen situated anterior to the left kidney.

The mass was well-defined, heterogeneously hypoechoic with multiple high flow vascular channels. CT

angiography of the abdomen was done for further assessment which revealed a large (8 x 8 x 8 cm), well encapsulated intra-abdominal mass lying anterior to the left kidney and caudal to the pancreatic tail (Figure 1 and Figure 2).

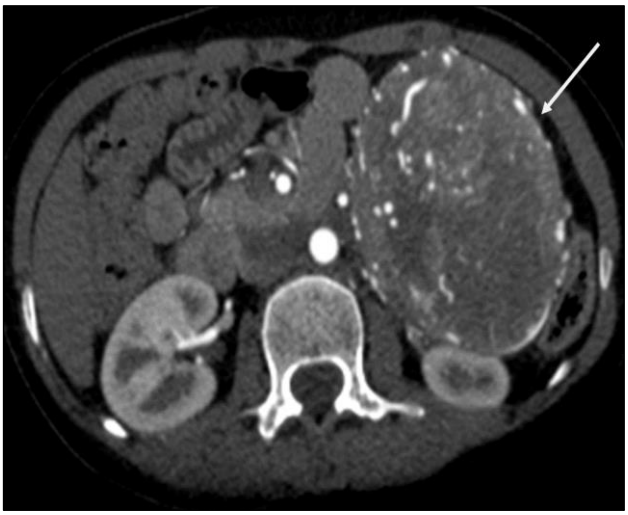

Figure 1: Axial CT image in arterial phase of contrast enhancement showing a well-defined rounded soft tissue mass lesion on left side lying anterior to left kidney and showing early enhancement and multiple intratumoral vessels (solid white arrow).

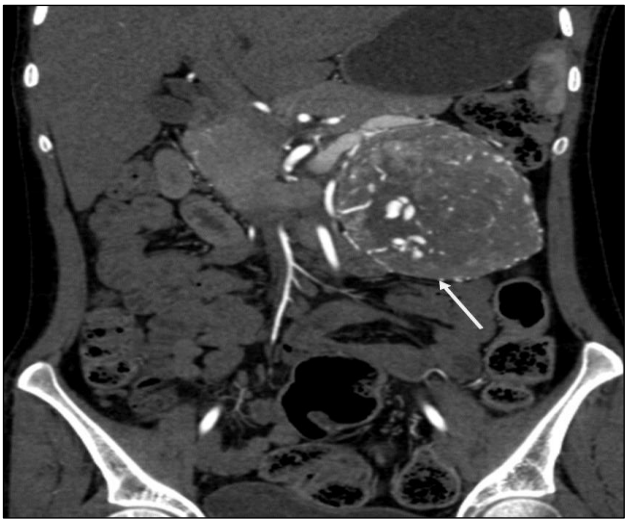

Figure 2. Coronal image showing the mass lesion with intense vascularity (straight white arrow) and displacing the pancreas and bowel loops around it.

Position of left kidney was unaltered and left adrenal gland was not separately visualized. Marked tumoral enhancement was seen with a central area of necrosis. Multiple intra-tumoral vessels were seen. The tumor derived blood supply from an ascending branch of the inferior mesenteric artery and lateral branch of the abdominal aorta above the left main renal artery (Figure3).

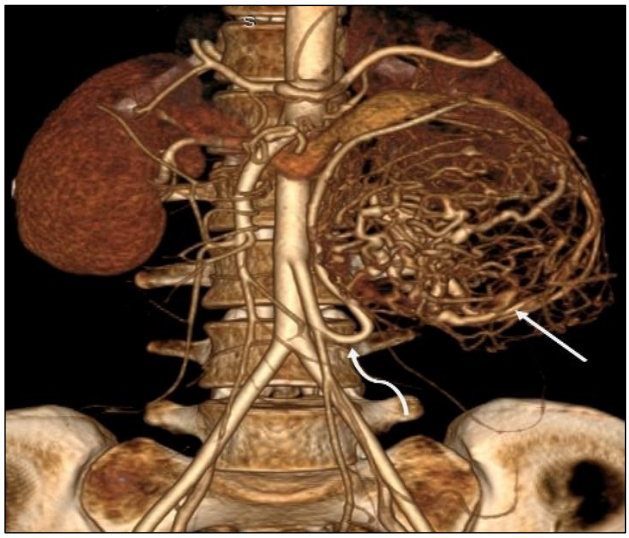

Figure 3: Volume rendered image showing intense tumoral vascularity (straight white arrow) and hypertrophied inferior mesenteric artery supplying the mass (curved white arrow).

Small arterial twigs were also noted arising from anterior division of left renal artery at hilum. The tumor showed persistent enhancement in the portal venous phase and the venous drainage of the tumor was seen into the left renal vein. In view of these radiologic findings, a diagnosis of hyper vascular retroperitoneal mass likely arising from left adrenal gland was made and a tentative diagnosis of pheochromocytoma was given.

Patient denied any history of cephalgia, sweating or palpitations. Urinary VMA levels were 40 µmol per 24 hours, serum metanephrine level was 21.7 IU and serum cortisol were 14 IU, which were within normal limits. MIBG scanning could not be done due to poor economic status and lack of affordability of the patient. Percutaneous biopsy was avoided due to the hypervascular nature of the tumor. Patient was referred to interventional radiology unit for preoperative embolization by the surgical oncology team.

After normal preliminary tests, with anesthesia team ready for the back-up, the patient was taken for preoperative embolization in digital subtraction angiography (DSA) lab. Right common femoral access was secured, and 5 French renal double curve and sidewinder catheter were utilized to obtain angiograms from celiac trunk, superior mesenteric artery, inferior mesenteric artery, left renal artery and lateral aortic branches including lumbar arteries.

Angiograms revealed major tumor vascularity from the inferior mesenteric artery and a direct branch from the aorta likely the middle adrenal artery (Figure 4 and Figure 5). Super selective cannulation of these vessels was done using a 2.7 French microcatheter (Progreat, Terumo) and the vessels were embolized with Gelfoam

slurry till stasis was observed with marked decrease in tumoral vascularity (Figure 6 and Figure 7).

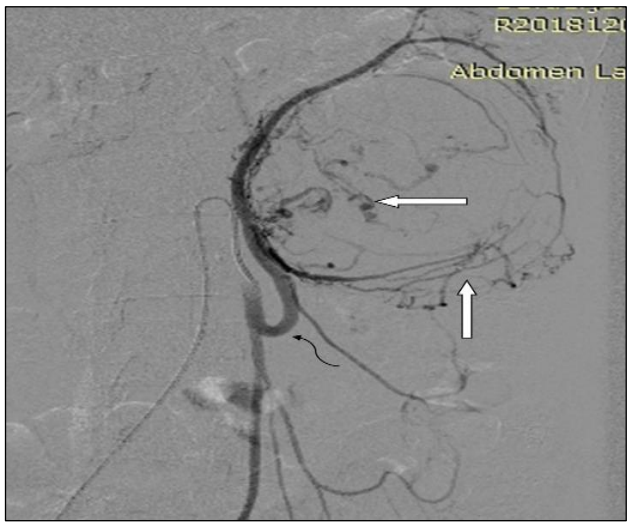

Figure 4: DSA image showing inferior mesenteric selective angiogram showing intra and peritumoral hypervascularity (straight white arrows) and hypertrophied inferior mesenteric artery (curved black arrow).

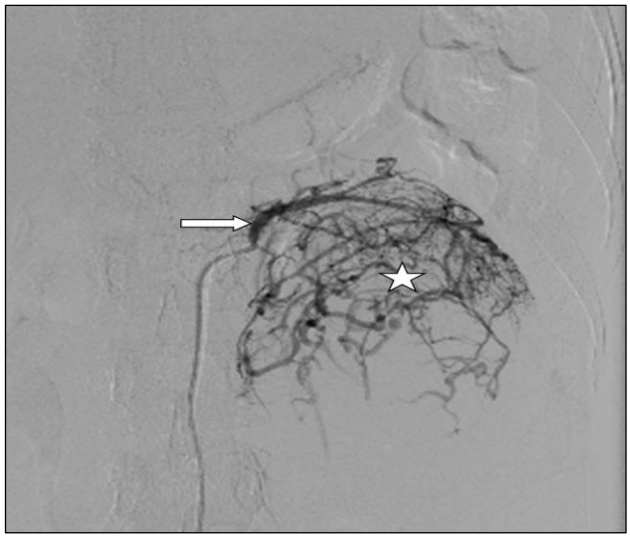

Figure 5: DSA image showing selective angiogram from left middle adrenal artery (straight white arrow) with tumoral hypervascularity (white star).

No arteriovenous shunting was observed within the tumor. Minor arterial supply to the tumor was also observed from the celiac trunk, superior mesenteric artery and left renal artery, however, embolization of these vessels was avoided due to difficulty in super selective cannulation and high risk of non-target embolization. The patient maintained a blood pressure of 110/70 mm Hg with SpO2 of 99% on room air throughout the procedure.

The patient complained of excessive vomiting and abdominal pain which was expected due to post-embolization tumoral ischemia and was managed with IV injection of tramadol and ondansetron.

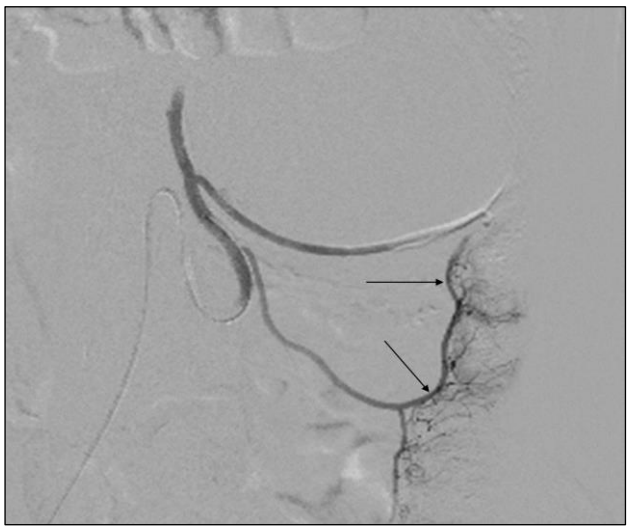

Figure 6: Post embolization angiogram shows significantly reduced vascularity and tumoral blush with normal opacification of descending colon vasa recta (straight black arrows).

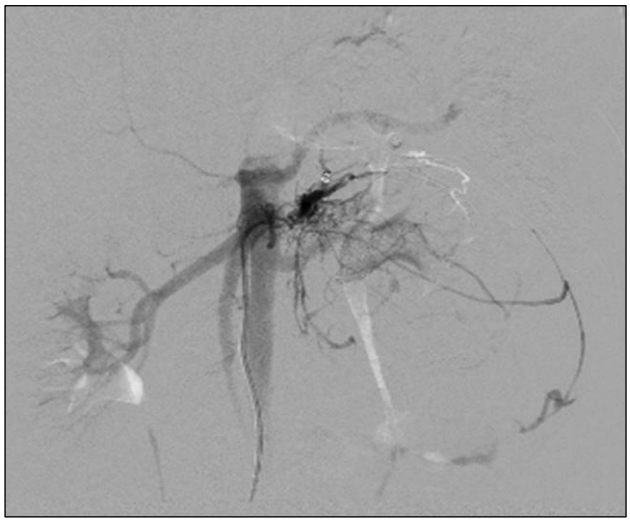

Figure 7: Post embolization angiogram from middle adrenal artery shows significantly reduced tumoral vascularity.

Intravenous fluid was started at the rate of 150 mL per hour and patient shifted to high dependency unit in oncology surgery ward after removal of the femoral sheath.

During observation in the ward, patient complained of vomiting and persistent abdominal pain with maintained vitals; however, after 6 hours, patient had fresh complaints of chest pain and respiratory distress. SpO2 dropped down to 60% in room air and she was intubated emergently and shifted to the intensive care unit (ICU). In the ICU, mechanical ventilation was started on

continuous positive airway pressure (CPAP) mode and oxygen saturation improved to 85%. On auscultation bilateral basal crepitations were noted and possibility of aspiration pneumonitis, pulmonary edema or pulmonary embolism was considered. Arterial blood gas analysis revealed metabolic acidosis with the base deficit of 10. Patient was stabilized overnight; however, in the morning she developed hypotension with tachycardia, abdominal distention with decreased bowel sounds and oliguria.

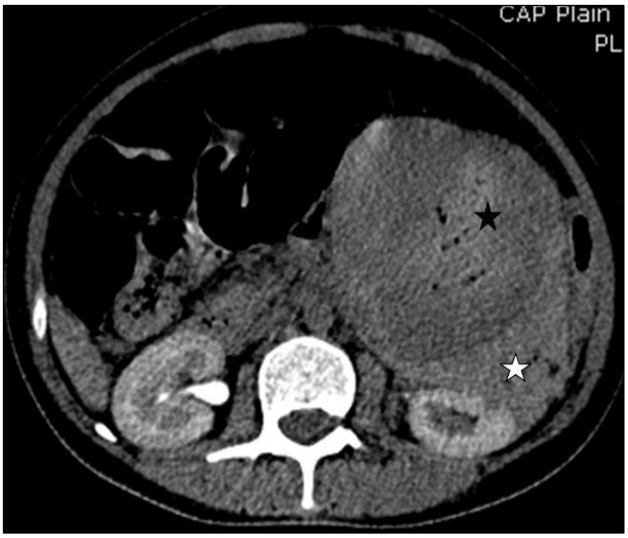

Figure 8: Non contrast CT axial image showing perinephric hematoma (white star), intratumoral hemorrhage (black star).

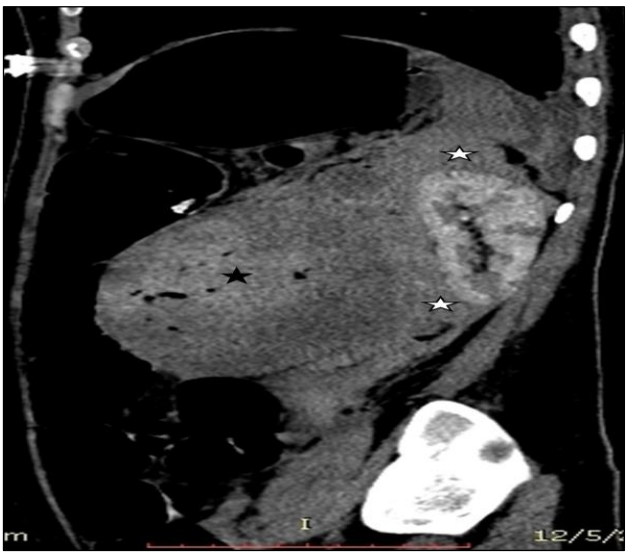

Figure 9: Sagittal CT image showing perinephric hematoma (white star) and intratumoral hemorrhage (black star).

Emergency non-contrast computed tomography scan of the chest and abdomen was done which showed ground glass haziness in bilateral lower lobes with minimal left-sided pleural effusion. Abdominal sections revealed, besides paralytic ileus, vicarious excretion of the contrast from the distended gallbladder and persistent nephrogram in both kidneys with small peripheral cortical infarcts. The tumor showed internal air foci (likely due to gelfoam embolisation) with hyperdensity suggestive of hemorrhage. Peritumoral, perinephric, retroperitoneal and pelvic hematomas were also observed which suggested tumoral rupture (Figure 8 and Figure 9).

Patient also underwent 2-D echocardiography which showed dilated left atrium and ventricle with significant hypokinesia and ejection fraction of 25%. ECG revealed tachycardia and ST segment changes consistent with ischemic pattern. Troponin T was positive, suggestive of acute myocardial infarction. Patient was started on inotropic support but subsequently developed ventricular fibrillation. CPR was started and patient was revived twice but succumbed during the third attempt.

Plausible explanation of this series of events could be tumoral rupture with possible massive release of catecholamines post-embolization causing myocardial ischemia and cardiogenic shock with pulmonary venous hypertension and pre-renal failure. Patient's relative did not consent for the autopsy so histopathological diagnosis of the tumor could not be obtained.

DISCUSSION

Pheochromocytoma is a catecholamine secreting tumor from the chromaffin cells of adrenal or extra-adrenal sites. Interestingly, it shows wide variation in its presentation from an incidental finding to a classic triad of episodic headache, sweating, and tachycardia.

The diagnosis of pheochromocytoma depends on the identification of an appropriately located mass on imaging with accompanying clinical and biochemical confirmation. The classic clinical manifestation includes hypertension, which may be episodic or refractory, in association with the triad of symptoms of palpitations, headaches, and diaphoresis. The diagnostic evaluation should include measurement of plasma metanephrine levels and of 24-hour urinary catecholamine levels. Indiscriminate biopsy of pheochromocytomas may trigger a catastrophic crisis and must be avoided. Unusual symptoms of secondary amenorrhea and diabetes mellitus due to pheochromocytoma have been reported in the literature by various authors which were controlled by alpha and beta blocker medications.[1]

Smaller neoplasms tend to be solid, whereas larger lesions are often hemorrhagic or cystic. Clinically silent pheochromocytomas also tend to be larger.[2] Other reported pathologic features include necrosis, calcification, and fibrosis. These gross features of pheochromocytomas have correlative radiologic features. Pheochromocytomas typically enhance avidly but can be heterogeneous or show non-enhancing regions due to cystic changes. When early intense enhancement occurs, it reflects the capillary-rich framework of the tumor.

Pheochromocytomas, can, however, demonstrate different and variable washout patterns and may, therefore, be confused with either adenomas or metastases.

Preoperative adrenal tumor embolization has been attempted by many interventional radiologists in the past. It has known to cause reduction in the size of the tumor, reduction in the vascularity before surgery, suppression of the hormonal axis and renal hormone production and, in particular circumstances, to maintain hemostasis in the eventuality of spontaneous tumoral rupture.

Prabhasvat et al. published a case report where they presented patients of adrenal tumor embolization, including each of adrenal carcinoma, cavernous hemangioma and myelolipoma and this technique resulted in successful embolization with decrease in tumoral vascularity and pain in all the patients. No mortality was observed although two of the three patients had post-embolization syndrome. In one of the patients, they observed tumoral rupture and hemorrhage after embolization which was controlled with blood transfusion. But did not notice any tumor volume shrinkage in the follow-up.[3]

Adrenal gland has arterial supply from three vessels: the superior, middle and inferior adrenal arteries which are the branches of inferior phrenic, aorta and renal arteries respectively.[4] In large adrenal tumors, additional arterial supply can be seen as in this case, since a highly vascular mass can parasitize additional arterial supply from neighbouring areas.

Spontaneous rupture of a pheochromocytoma is a rare complication and tumoral rupture post-embolization is even rarer. Massive release of catecholamines as well as blood loss can be life-threatening for the patient. Pua et al, have reported one such case where they had embolized a ruptured pheochromocytoma with PVA particles to control bleeding. Their angiograms also demonstrated peripheral tumoral blush and absent central vascularity as in this case.[5] Many researchers have also embolized pheochromocytoma using Gelfoam with decreased requirements for antihypertensive drugs after the procedure.[6,7] Transarterial embolization of pheochromocytoma has been done previously to manage hypertensive crisis by Hrabovsky et al, Many researchers have shown the efficacy of transarterial embolization of both adrenal and extra-adrenal paragangliomas.[7,8] Daniele et al, had embolized a large pelvic sympathetic paraganglioma in a young female by cannulating the inferior mesenteric artery.[9]

The classic clinical triad of pheochromocytoma rupture is intense vasoconstriction, tachycardia and labile hypertension; however, this is demonstrated in less than one-third of the patient's having tumoral rupture.[10] Other common clinical features reported are loin pain, nausea and vomiting.[11]

Amongst adrenal masses, pheochromocytoma is the most common tumor to bleed. Approximately 50% of adrenal hemorrhagic masses were due to pheochromocytoma in a study by Marti et al, Risk factors for pheochromocytoma rupture are trauma, anticoagulation and initiation of alpha blocker therapy.[12,13] Another mechanism could be tumoral necrosis induced cell destruction with massive release of catecholamines causing vasoconstriction and elevated intracapsular pressure resulting in tumoral rupture and hemorrhage.[14]

Many authors have authenticated the fact of catecholamine-induced cardiogenic shock. Acute myocardial infarction can be due to hyperdynamic circulation causing increased myocardial oxygen demand or direct toxicity of catecholamines to myocardial cells. Cardiovascular complications like myocarditis, myocardial infarction and heart failure is likely due to massive dumping of catecholamines especially when vascular supply to the tumor has been blocked.[15]

CONCLUSION

In this index case, the imaging features were that of pheochromocytoma; however, unusual clinical symptoms like amenorrhea and lack of hypertensive symptoms like diaphoresis and headache along with negative urinary and serum catecholamine markers was a clinical enigma for us and after reviewing the literature of preoperative embolization of adrenal tumors and pheochromocytomas, the decision for embolization was made. Unusual complication of tumoral rupture with possible catecholamine-induced myocardial infarction and cardiogenic shock occurred which were also unusual and unexpected complications. This can be a learning experience that adrenal masses, even those without biochemical evidence of hormone production, should be handled with caution and any endovascular embolization therapy is not free of the risks and complications that can occur even after the procedure.

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