

REFERENCES

1. Thull DL, Vogel VG. Recognition and management of hereditary breast cancer syndromes. *Oncologist*. 2004;9:13-24.
2. Aretz S, Stienen D, Uhlhaas S, Loff S, Back W, Pagenstecher C, et al. High proportion of large genomic STK11 deletions in Peutz-Jeghers syndrome. *Hum Mutat*. 2005;26:513-9.
3. Volikos E, Robinson J, Aittomaki K, Mecklin JP, Järvinen H, Westerman AM, et al. LKB1 exonic and whole gene deletion are a common cause of Peutz-Jeghers syndrome. *J Med Genet*. 2006;43:18-9.
4. Bruwer A, Barga JA, Kierland RR. Surface pigmentation and generalized intestinal polyposis; (Peutz-Jeghers syndrome). *Proc Staff Meet May Clin*. 1954;29:168-71.
5. Calva D, Howe JR. Síndromes de poliposis hamartomatosa. *Surg Clin N Am*. 2008;88:779-817.
6. Espin Basany E, García Aguilar J. Cáncer colorrectal: aspectos generales y lesiones precancerosas. *Manual de la Asociación Española de Cirujanos*. Madrid: Panamericana; 2010. p. 502.
7. Jansen M, de Leng WW, Baas AF, Myoshi H, Mathus-Vliegen L, Taketo MM, et al. Mucosal prolapse in the pathogenesis of Peutz-Jeghers polyposis. *Gut*. 2006;55:1-5.
8. Kinzler KW, Vogelstein B. Landscaping the cancer terrain. *Science*. 1998;280:1036-7.
9. Beggs AD, Latchford AR, Vasen HF, Moslein G, Alonso A, Aretz S, et al. Peutz-Jeghers syndrome: a systematic review and recommendations for management. *Gut*. 2010;59:975-86.

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Retroperitoneal Haemorrhage After Traumatic Rupture of a Phaechromocytoma

Hemorragia retroperitoneal tras rotura traumática de un feocromocitoma

Pheochromocytomas are neuroendocrine tumors of the adrenal medulla (originating in the chromaffin cells) or extra-adrenal chromaffin tissue that occur during involution after birth and secrete excessive quantities of catecholamines. Traumatic rupture of a pheochromocytoma and the resulting retroperitoneal hemorrhage are rare complications, and management can be difficult.

We present the case of a 45-year-old male who, in a suicide attempt, fell from the fifth floor of a building and suffered severe trauma. His medical history included arterial hypertension, insulin-dependent diabetes and the presence of a right adrenal mass that was being studied after having been incidentally detected during an abdominal ultrasound performed some weeks earlier. Meanwhile, the patient was receiving psychiatric drug treatment for anxiety and depression.

Upon arriving at our Emergency Department, the patient presented blood pressure of 180/110 mmHg, heart rate 135 bpm and hemoglobin 9.9 g/dL. Emergency abdominal computed tomography (CT) revealed a right adrenal mass measuring 10.5 cm×9 cm×8.8 cm with signs of active bleeding and a retroperitoneal hematoma (Fig. 1). The patient also presented facial trauma, bilateral pneumothorax and fractures of the right transverse processes from L1 to L5, sacrum, coccyx and right tibia and open fracture of the right astragalus.

Given the suspected active bleeding of the adrenal mass, the patient was considered a candidate for radiological embolization prior to the possible need for emergency surgery. Due to the hemodynamic stability and to the fact that hemoglobin levels were maintained above 9 g/dL for 24 h after hospitalization, we opted for a conservative approach and initiated treatment with urapidil and labetalol, which provided adequate blood pressure control 36 h after hospitalization.

Catecholamine determination in urine showed levels of metanephrine of 20 659 mcg/dL (normal, 64-302 mcg/dL) and norepinephrine 5644 mcg/dL (normal, 12-85 mcg/dL). These

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Fig. 1 – Abdominal computed tomography showing retroperitoneal hematoma with tumor rupture in the right suprarenal gland.

data, together with the radiological findings, established the diagnosis of traumatic rupture of a pheochromocytoma.

Four weeks later, follow-up CT showed partial reabsorption of the retroperitoneal hematoma, with better tumor definition (Fig. 2).

Laparoscopic right adrenalectomy was scheduled (fourth week of hospitalization). Nonetheless, the adhesion of the tumor to the inferior vena cava and the difficulties for maintaining the patient hemodynamically stable made it necessary to convert to open surgery, and the procedure was successfully completed. The pathology report confirmed the diagnosis of hemorrhagic cystic pheochromocytoma.

Post-operative recovery was favorable. The psychiatric symptoms disappeared and catecholamine levels progressively returned to normal limits.

Approximately 5% of the adrenal tumors are pheochromocytomas.¹ In the literature, the incidence of traumatic injury

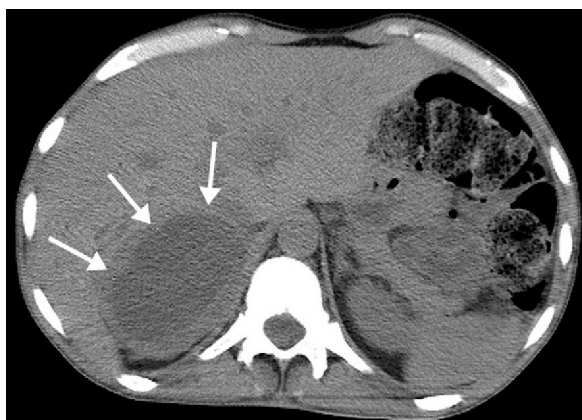


Fig. 2 – Abdominal computed tomography (4 weeks after trauma) showing partial reabsorption of the retroperitoneal hematoma with a clearer outline of the tumor.

to the adrenal gland is between 0.03% and 3%. Traumatic adrenal injury may itself present as a hyperadrenergic state simulating a pheochromocytoma, although when the hematoma is resolved and the gland recovers its normal function, both arterial pressure as well as catecholamine levels return to normal limits. However, complete bilateral destruction of the adrenal glands usually causes cardiovascular shock as a consequence of corticoadrenal insufficiency.²

Spontaneous non-traumatic hemorrhage of a pheochromocytoma is another rare form of presentation of these tumors, which is often associated with anticoagulant treatment or severe sepsis; abdominal pain and hypertensive crises are the most frequent manifestations. Only 54 cases have been described in the literature.³

Immediate surgery as treatment for the traumatic rupture of a pheochromocytoma has been associated with a high rate of mortality (44.7%), which is minimized if the surgery is performed electively after stabilizing blood pressure with the use of alpha blockers or after arterial embolization.³

If blood pressure is uncontrollable or bleeding is persistent despite pharmaceutical treatment, embolization should be the first option for controlling hemostasis before opting for emergency surgery.

In our patient's case history, the presence of high catecholamine levels in urine and the radiological images of adrenal gland rupture suggested the diagnosis of traumatic hemorrhagic pheochromocytoma. This is an extremely rare condition (5 cases reported in the literature) requiring complex management.⁴ Therefore, an adrenal mass that is discovered incidentally justifies an etiological study to rule out the presence of pheochromocytoma, which is especially relevant for trauma patients who could need surgery for other injuries. In our case, conservative management was the initial option given the patient's hemodynamic stability. The rapid response to drug treatment allowed surgical resection to be delayed, and it was performed 4 weeks later under more favorable anatomical and clinical circumstances.

Lastly, pheochromocytoma has been described as a "great imitator" due to its numerous manifestations. Anxiety is the most frequent psychiatric symptoms, with a prevalence of 22%–44% of patients with this tumor. Other less common manifestations have also been reported, such as depression, suicide attempts and delirium.⁵ The psychiatric problems that our patient presented were the cause of the fall and subsequent trauma injuries and were resolved after the tumor was resected.

REFERENCES

1. Young Jr WF. Clinical practice. The incidentally discovered adrenal mass. *N Engl J Med.* 2007;356:601–10.
2. Khan A, Mutazindwa T, Hassan A, al-Zuhair N. Unilateral traumatic adrenal haematoma presenting as pheochromocytoma. *Eur J Radiol.* 1998;28:133–5.
3. Habib M, Tarazi I, Batta M. Arterial embolization for ruptured adrenal pheochromocytoma. *Curr Oncol.* 2010;17:65–70.
4. Hayashi T, Nin M, Yamamoto Y, Kamoto A, Ujike T, Nishimura K, et al. Pheochromocytoma with retroperitoneal

hemorrhage after abdominal trauma. *Hinyokika Kyo.* 2009;55:703-6.

5. Medvei VC, Cattell WR. Mental symptoms presenting in phaeochromocytoma: a case report and review. *J R Soc Med.* 1988;81:550-1.

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Portal Pneumatosis Secondary to Acute Appendicitis

Neumatosis portal secundaria a apendicitis aguda

Portal vein pneumatosis is a radiological finding that is classically associated with intestinal necrosis and ischemia, but it has also been described associated with other abdominal pathologies.¹⁻⁴ What is interesting about the case that we report is the unusual presence of this entity in a commonly treated disease in the emergency surgery department.

The patient is a 59-year-old male who came to our Emergency Room due to symptoms of dyspnea, oliguria, abdominal distension and fever that had been developing over the previous 3 days without other accompanying symptoms. The patient's medical history included arterial hypertension, obesity, surgery for cerebral astrocytoma 35 years earlier and ventriculoatrial shunt placement. Upon arrival, the patient presented tachypnea, hypoxemia and a trend toward hemodynamic instability that improved with serum therapy. Abdominal exploration showed a soft, depressible abdomen that was slightly distended, with diffuse pain but no clear signs of peritoneal irritation and with reduced peristalsis. The blood tests showed renal failure and dehydration (creatinine 5.3 mg/dL, hemoglobin 16.4 g/L) and parameters of sepsis (presence of 31% bands and PCR 50.2 mg/dL). After the hemodynamic stabilization of the patient, an abdominal CT showed an important portal vein pneumatosis in the left hepatic lobe (Fig. 1) and portal vein (Fig. 2), with several gas bubbles in the anterior mesenteric fat in a probable extraluminal location, an increase in the caliber of the vermiform appendix with poorly-defined edges and trabeculation of the adjacent fat. All this was compatible with acute appendicitis, and we therefore decided emergency surgery.

At surgery, we observed diffuse purulent peritonitis secondary to acute appendicitis with perforation at the base of the cecum. Appendectomy was performed with thorough abdominal cavity lavage. During the intervention, the patient presented hemodynamic instability requiring support with vasoactive drugs, tachycardia with ventricular extrasystoles (which reverted after the administration of amiodarone), severe hypoxemia and oliguria. Post-operative complications included hypoxemia requiring prolonged intubation, consumption

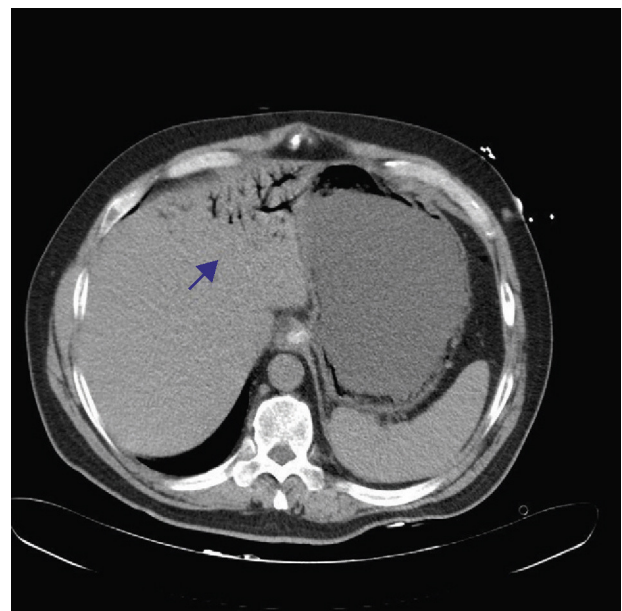


Fig. 1 – Presence of portal vein pneumatosis in the left hepatic lobe.

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