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Letter to the Editor

Unilateral aggressive pheochromocytoma revealed by a massive intraperitoneal hemorrhage five years after an initial presentation suggesting an adrenal hematoma

Phéochromocytome agressif révélé par une rupture hémorragique intrapéritonéale cinq ans après une première manifestation évoquant un hématome surrénalien

Keywords: Pheochromocytoma; Hemorrhagic rupture; Adrenal hematoma; Metabolic radiotherapy

Mots clés : Phéochromocytome ; Rupture hémorragique ; Hématome surrénalien ; Radiothérapie métabolique

Dear Editor in chief, you have recently published the unusual case of a hemorrhagic pheochromocytoma (PH) revealed by myocardial infarction in a 50-year-old man [1]. Such acute revelation due to spontaneous tumoral rupture are extremely rare. We report here the case of a "malignant" PH exhibiting a hemorrhagic and life-threatening tumoral rupture.

1. Case report

Ten years ago, a 26-year-old man, with a paternal history of primary hypertension, was referred to our department for acute hypertension (260/100 mmHg) during a physical effort, associated with headache, sweats, palpitations and paroxysmal tachycardia, strongly suggestive of PH. Initial evaluation revealed subnormal levels of urinary metanephrines (MN) (Table 1), while chromogranin A level was normal, as was calcitonin level (< 5 pg/mL). Computed tomography (CT) disclosed a 31 × 30 mm left adrenal lesion with a spontaneous density of 37 Hounsfield Units (HU) (Fig. 1A), while ^{123}I-MIBG scintigraphy showed no pathological uptake. Magnetic resonance imaging (MRI) strongly suggested a left adrenal hematoma (Fig. 1B). It was thought that the hematoma led to the release of metanephrines, masquerading for a PH. In order to explain this spontaneous hematoma, the patient was screened for a hemostasis disorder and was diagnosed for a heterozygous Factor V Leiden mutation. Patient was treated with antihypertensive drugs (urapidil 120 mg/day and enalapril 20 mg/day) during six months, until laboratory tests revealed normal levels of urinary MN. MRI showed a near complete regression of the left adrenal mass with only a small residual nodular hypertrophy, with a spontaneous T1-hypersignal, thought to be a remnant of the hematoma rather than a PH.

Unfortunately, the patient was lost to follow-up. Four years later, he developed sudden severe abdominal pain, sweats, nausea and vomiting associated with a loss of consciousness in a context of a labile hypertension, after dental care that included the use of nonsteroid anti-inflammatory drugs and local anesthetic. Initial evaluation showed a heart rate of 64 bpm, systolic BP of 74 mmHg, diffuse peritoneal signs, cold extremities associated with massive deglobulization (hemoglobin 3.7 mmol/L) and disseminated intravascular coagulopathy. This was strongly suggestive of an active hemorrhage, confirmed by a non-contrast abdominal CT revealing an intra-abdominal hemorrhage due to the rupture of a huge 106 × 236 × 147 mm heterogeneous left adrenal mass. Because of hemodynamic instability despite an aggressive resuscitation with blood products, an emergency laparotomy was undertaken, revealing extensive retro and intraperitoneal hemorrhage and allowing the removal of a left ruptured adrenal tumor of 234 grams [2] with a splenectomy because the tumor was strongly adhesive to the spleen and to the diaphragmatic cupola. Nevertheless, BP remained labile and hematocrit dropped, leading to a second lifesaving surgical procedure, with packing of the abdomen with pads that finally stopped the bleeding. Due to the massive bursting of the tumor, a precise histological analysis was difficult, but suggested the diagnosis of malignant or very aggressive PH because of blood vessel infiltration and capsular invasion (high PASS score: 9). The patient survived despite multi-organ failure, requiring intensive care monitoring and transient hemodialysis. Few days after the hemorrhagic rupture, serum MN and normetanephrines (NMN) levels were still markedly elevated. One month later, BP was controlled by bisoprolol and urapidil; levels of MN and NMN remained high. Abdominal CT and MRI were normal; ^{123}I-MIBG scintigraphy, ^{111}In-DTPA-Octreotide scintigraphy and ^{18}FDG-PET-CT showed no pathological uptake. Given the young age of the patient, the existence of a monozygotic twin later diagnosed with bifocal kidney cancer (successfully cured by surgery) and the context of aggressive PH, a genetic analysis

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Table 1
Evolution of serum and urinary metanephrines and normetanephrines levels, and serum chromogranin A levels since the hemorrhagic rupture of the pheochromocytoma until now.

Date	First Diagnosis	6 mo later	Rupture First surgery	1 mo later	3 mo later	6 mo later	13 mo later Second surgery	1 mo later	6 mo later Metabolic radiotherapy	2 mo later	6 mo later	12 mo later
Serum metanephrines pmol/L (N: 1500–5100)	–	–	8450	4530	4220	4600	13,150	7269	8208	4198	2401	2832
Serum normetanephrines pmol/L (N: 5450–10900)	–	–	34,510	19,610	14,360	15,520	30,970	10,247	19,063	15,635	6922	4771
Urinary methoxyadrenaline µmol/24 hrs (N: 0,40–1,50)	1.19	0.74	–	–	–	–	–	–	–	–	–	–
Urinary methoxynoradrenaline µmol/24 hrs (N: 0,50–2,00)	2.33	1.38	–	–	–	–	–	–	–	–	–	–
Chromogranin A µg/L (N: 20–100)	54	–	–	98	–	–	53	–	–	–	–	–

mo: months.

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