CLINICAL CASE

SURGICAL MANAGEMENT OF TYPE B AORTIC DISSECTION AND PHEOCHROMOCYTOMA

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Abstract

Arterial hypertension is the most important risk factor for aortic dissection. Arterial hypertension has been found in approximately 80% of aortic dissection patients. Typically, arterial hypertension is long-standing and poorly regulated in aortic dissection patients. A pheochromocytoma is a rare, catecholamine-secreting tumour that may precipitate life-threatening hypertension. The tumour is malignant in 10% of cases but may be cured completely by surgical removal. We present the case of a 49-year-old patient, heavy smoker, hypertensive, who addressed the hospital in the context of acute interscapulovertebral pain (November 2014). After a contrast agent CT scan of the thorax + upper abdomen + pelvis, a type B thoracic aortic dissection was observed, without subdiaphragmatic extension or extension to the supraaortic trunks, without mediastinal effraction, and a right adrenal tumour of approximately 3.6 cm was also detected. The patient was directed to the cardiovascular surgery service, where an endoprosthesis with Vaillant prosthesis of the descending aorta and subclavian carotid bypass were performed, with good postoperative evolution. The patient was then referred to The National Institute of Endocrinology "C.I. Parhon" for further investigations required prior to the adrenal surgery. The suspicion of pheochromocytoma was confirmed by urinary metanephrine and normetanephrine dosing, and adrenal surgery was recommended. The patient was placed under alpha and beta blocker treatment for the prevention of hypertensive flares and any potential arrhythmias. Surgical intervention was performed for adrenal tumour resection. Histopathological examination of the surgical sample confirmed the diagnosis of pheochromocytoma. The particularity of the case is the discovery of an adrenal tumour due to an aortic dissection, the patient having no other symptoms and presenting a recorded history of hypertension.

Keywords*: hypertension, aortic dissection, pheochromocytoma, surgery*

Introduction

Pheochromocytoma approximate incidence in the general population is 0.001% and 0.5% in hypertensive patients. The rule of "10%" is currently accepted, stating that 10% of pheochromocytomas are bilateral or multiple, 10% are ectopic (extra-adrenal), 10% are

malignant, 10% are familial (circumstance in which half are malignant) and 10% occur in children. Pheochromocytoma (Figure 1) can occur at any age, but most frequently between 30-50 years. There is no gender predilection in adults. Though they are usually sporadic tumours, familial forms are also described, with autosomal dominant transmission with variable penetrance. Sporadic pheochromocytomas are more often located on the right side. Paroxysmal hypertension is a constant manifestation. Cardiac rhythm disturbances, sweating, headache are also described. Aortic dissection is a rare event in pheochromocytomas.

Aortic dissection can be rapidly fatal, with many patients dying before presentation to the emergency department or before diagnosis is made in the emergency department. No sign or symptom can positively identify acute aortic dissection. Clinical manifestations include the following: a sudden onset of severe chest pain that often has a tearing or ripping quality (classic symptom), a chest pain that may be mild, an anterior chest pain - usually associated with anterior arch or aortic root dissection, a neck or jaw pain - with aortic arch involvement and extension into the great vessels, a tearing or ripping intrascapular pain - that may indicate dissection involving the descending aorta, no pain in about 10% of patients, syncope, cerebrovascular accident symptoms (e.g. hemianesthesia, and hemiparesis, hemiplegia), altered mental status, numbness and tingling, pain, or weakness in the extremities, Horner's syndrome (i.e. ptosis, miosis, anhidrosis), dyspnea, hemoptysis, dysphagia, flank pain (with renal artery involvement), abdominal pain (with abdominal aorta involvement), fever, anxiety and premonitions of death [1-3].

Material and Methods

We present the case of a 49-year-old patient, heavy smoker, hypertensive, who went to the hospital in the context of acute interscapulovertebral pain (November 2014). The patient had records of grade III hypertension, very high risk class, since 2012, but did not undergo antihypertensive treatment and did not regularly monitor his blood pressure in order to detect changes in value. Following his presentation to the hospital in the context of the above symptoms, a contrast agent CT scan of the thorax + upper abdomen + pelvis was performed, which highlighted type B thoracic aortic dissection (Figure 2) without subdiaphragmatic expansion or expansion in the supraaortic trunks, without mediastinal effraction, and a right adrenal tumour around 3.6 cm in diameter was also detected.

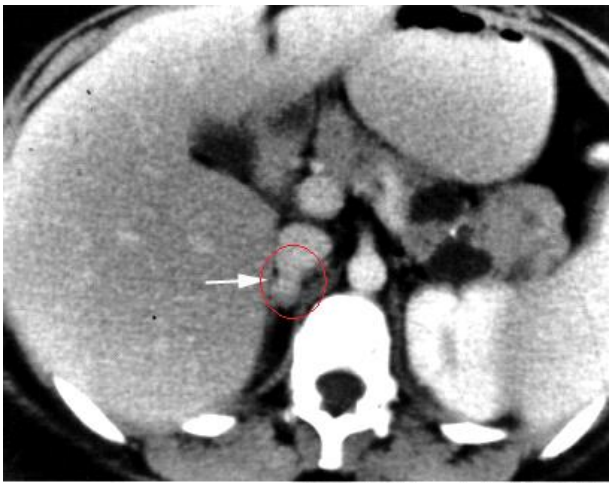

Figure 1 - Pheocromocytoma- CT scan

Anatomy and classification of aortic dissection (Acute: presentation <14 days, Sub-acute: 14 days - 2 months, Chronic: >2 months; Stanford Type A: ascending aorta (with or without descending aorta) Type B: descending aorta; DeBakey Type I: ascending aorta + descending Type II: confined to ascending aorta Type III: confined to descening aorta, beyond origin of subclavian artery) [4].

The patient was directed to the cardiovascular surgery service where the patient was hemodynamically stable upon presentation, with BP = 115/80mmHg and regular sinus rhythm (under cardiological medication), with no neurologic manifestation, afebrile, with diuresis, intestinal transit present without associated digestive pathology, but still accused pain at interscapulovertebral level. Given the presence of the dissection fold so close to the origin of the left subclavian artery, left carotidosubclavicular bypass was initially performed with No. 8 e-PTFE prosthesis. Subsequently, two vascular Vaillant endoprostheses were mounted down to the origin of the celiac trunk. The postoperative evolution was good, without neurological sequelae or angina. Transesophageal control

was performed by CT scan as well, detecting no paraprosthetic leak, no dissection present at the aortic arch or abdominal aorta level. The patient's evolution was afebrile, compensated from a cardiac point-of-view, with blood pressure levels controlled by medication (Beta blockers, Clonidine, Thiazide diuretic, Amlodipine, Aspirin). He was discharged with indication for myocardial revascularization surgery and surgical cure of the adrenal gland tumour.

The patient addresses The Emergency Hospital, Bucharest, where clinical examination upon admission revealed a slender abdomen, without any spontaneous pain or pain upon palpation, without signs of peritoneal irritation. Blood tests on admission showed leukocytosis (12,590 / mmc), anemia (Hb = 9.9 g / dl; Ht 31.1%), thrombocytosis and mixed dyslipidemia (cholesterol 220 mg / dl, triglycerides 153 mg / dl). Blood glucose was also modified (126 mg / dl).

An abdominal ultrasound revealed: normal liver size, clear shape and heterogeneous structure due to the presence of a hypoechoic structure, oval, 18 mm in size, in the VIth segment; gallbladder with a few hyperechoic images, 3 mm in size, located at the level of the fundus; right kidney with a 25 mm parapielic cyst; right adrenal gland 33 mm in size, hypoechoic, heterogeneous; all other abdominal viscera with normal ultrasonographic appearance; no free intraperitoneal fluid.

After preoperative anaesthetic-surgical evaluation a complete hormonal profile was requested and the patient was referred to The National Institute of Endocrinology "C.I. Parhon" for further investigations.

Upon admission to the department of endocrinology, the patient continued to be hemodynamically stable under treatment, with BP=120/60 mmHg and HR= 80 bpm, afebrile. Plasma and urinary metanephrines were collected, the analysis results being very elevated (urinary metanephrines 776.20ug / 24h - benchmarks 50-350 ug / 24h, urinary normetanephrines 1.161 ug / 24h- benchmarks 100-600 ug / 24h, confirming the suspicion of pheochromocytoma. Hypercholesterolemia (208 mg / dl) and high triglycerides (251 mg / dl), leukocytosis (14,000 / mmc), anemia (Hb = 10.9 g / dL) and elevated ESR (28.30) were also detected.

Being under alpha and beta blocker therapy for the prevention of hypertensive flares and potential arrhythmias for about a month, the patient was immediately directed to the general surgery service of the Emergency Hospital, Bucharest for surgical cure. Surgery was performed via laparoscopic approach and a moderately increased liver volume was present. A right adrenal tumour formation of 4/4/3 cm, well-delimited, with its own capsule, without extension into the adjacent structures, without locoregional adenopathy was found (Figures 3,4).

Laparoscopic right adrenalectomy was performed (Figure 5). Postoperative evolution was favorable from a surgical point-of-view with resumption of bowel movement and of oral nutrition, as well as per primam healing of the surgical wounds. Postoperatively, the patient received 200mg Hydrocortisone Hemisuccinate at 10 a.m. and 200mg at 16 pm until the 30th of January 2015, 10 a.m. included. The patient was discharged with the recommendations to avoid physical efforts, to continue chronic treatment in an outpatient unit, and to present to regular medical follow-up.

Histopathological examination of the operative sample confirmed the diagnosis of pheochromocytoma (Figures 6,7). The particularity of the case is the discovery of an adrenal tumour due to an aortic dissection, the patient having no other symptoms and presenting recorded history of hypertension.

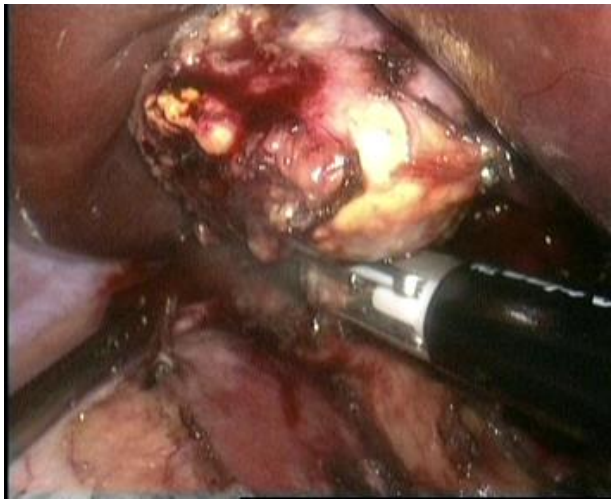

Figure 3 - Intraoperatory tumoral aspect

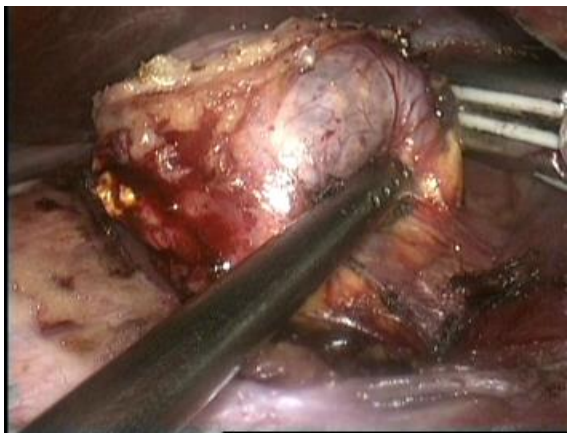
Figure 4 - Intraoperatory tumoral aspect

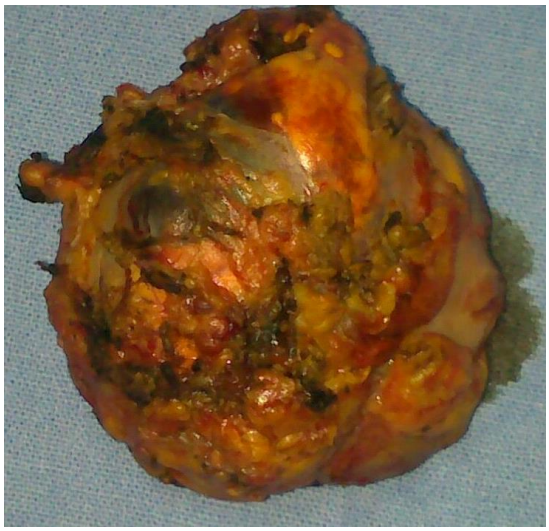
Figure 5 - Macroscopic tumoral aspect

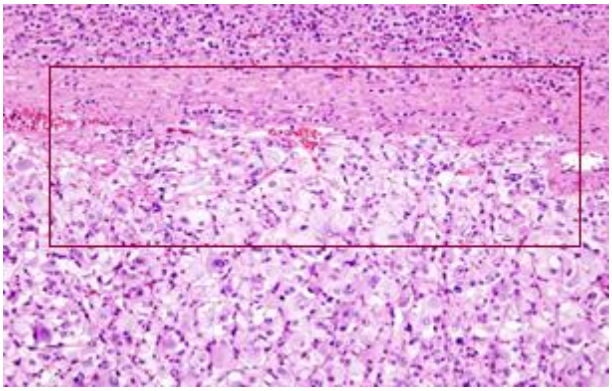
Figure 6 - Microscopic details of pheochromocytoma

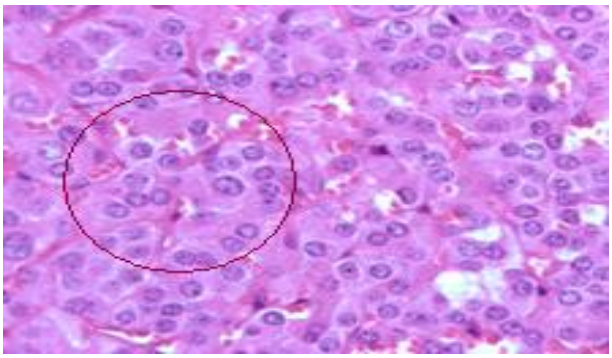
Figure 7 - Microscopic details of pheochromocytoma

Discussions

Both pheochromocytoma and aortic dissection are rare pathologies, their association being all the more special. The scarcity of symptoms present in our patient is also a notable element, as well as the fact that an acute event, an aortic dissection, led to the diagnosis of the adrenal tumour. The only effective treatment in pheochromocytoma is exclusion of the source of catecholamines by surgical ablation of the tumour formation or formations, preceded, accompanied, and followed by medication preventing or fighting paroxysmal hypertensive and/or hypotensive flares.

Similar cases in literature: Triplett and Atuk [5] reported the first case of a patient with dissecting aortic aneurysm and pheochromocytoma in 1975. Azizi and colleagues [6] reported a case of concomitant type A dissection and pheochromocytoma in 1994. There have been other reports of pheochromocytoma associated with dissecting abdominal aortic aneurysm [7,8] but for our knowledge there have been no reported cases of type B aortic dissection and pheocromocitoma in Romania.

In a patient who has a pheochromocytoma, an acute hypertensive crisis may develop at the induction of anesthesia or intraoperatively, and this can be lethal. Manipulation of a pheochromocytoma can precipitate the release of massive amounts of catecholamines. In one study, 36 of 143 patients undergoing pheochromocytoma resection had sustained intraoperative hypertension develop [8].

Ascending dissections require emergency surgical repair, whereas medical therapy is usually the initial strategy for acute type B dissections. Ascending aortic dissection is most common in the 50- to 60-year age range, whereas descending dissections occur more commonly in older individuals. Because acute aortic dissection is much less common than other conditions associated with chest or back pain, a high index of suspicion is important in making this diagnosis.

Although most patients with type B dissections are hypertensive, many patients with type A dissections are normotensive or hypotensive on presentation.

To avoid a hypertensive crisis at the induction of anesthesia, complete alpha blockade and beta blockade are necessary. A 10-day to 14-day course of catecholamine blockade is optimal. Alternatively, metyrosine (a tyrosine hydroxylase inhibitor that blocks the synthesis of norepinephrine) is very effective but typically takes as much as 3 weeks to eliminate all norepinephrine synthesis [9].

Conclusions

Pheochromocytoma is a rare adrenal tumour, which presents as a dominant symptom the development of secondary hypertension, which in turn can lead to major cardiovascular complications such as aortic dissection. Aortic dissection is a severe, potentially fatal situation, which can determine a range of clinical manifestations. In the case of our patient, the only symptom prompting him to present to the hospital was an acute interscapulovertebral pain.

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