

CAS CLINIQUE

Pheochromocytoma and congenital heart diseases : about a case

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Abstract

Pheochromocytoma is a tumor developed in the deep layer of the adrenal gland from chromaffin cells responsible for hypersecretion of catecholamines.

The latter is the cause of vasoactive disorders, namely hypertensive peaks with tachycardia. These induced disorders can be difficult to manage in patients with an underlying cardiac pathology.

We report the case of an 18-year-old patient whom we had to treat for left pheochromocytoma. He has been followed since childhood for complex congenital heart disease with left ventricular single heart type.

The patient was operated on under general anesthesia with many precautions taken by conventional means without incident. It seemed appropriate to describe this case with a review of the literature for its particularity and rarity.

Introduction

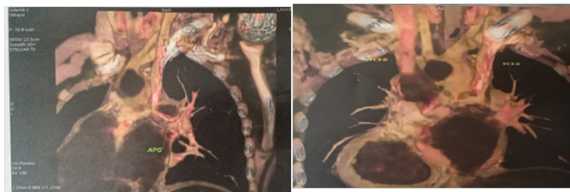
Among the etiologies of secondary hypertension, we find adrenal medulla tumor, pheochromocytoma (PCM) whose mechanism is hypersecretion of vasoactive amines (catecholamines). The discovery of a PCM often requires its surgical excision, particularly in the face of signs of radiological aggressiveness. Its occurrence in an area of complex heart disease constitutes a real challenge both for the anesthetist-resuscitator and for the surgeon.

We report the case of a PCM occurring in an 18-year-old boy with a complex congenital heart disease involving a left ventricular single heart. We suggest a possible cause and effect relationship between the background of cyanogenic congenital heart disease and the appearance of PCM.

Observation

We had to take care of a 18-year-old patient with a complex congenital heart disease involving a left univentricular heart with dextrocardia, azygocaval return, two superior vena cavae and a single atrium with minimal leakage from the atrioventricular valve (AVV) (**Figure 1**).

Figure 1. Two superior vena cavae and a single atrium



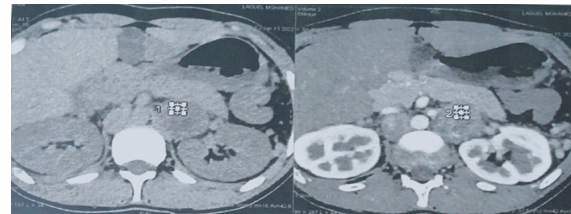
It should be noted that at the age of 4, he benefited from a bicavo-bipulmonary bypass with disconnection of the trunk of the pulmonary artery. During his follow-up, a chest CT scan was performed revealing a left adrenal incidentaloma (AI). The examination found a patient with cyanosis perioral and extremities. Presence of an incomplete Menard triad, made up of paroxysmal palpitations and sweating. With stable hemodynamic parameters.

The dosage of Methoxylated derivatives are high, with normetanephrine at 14.8 times normal, as well as high dopamine levels at 3 times normal. The standard assessment reveals polycythemia with hemoglobin at 23.2 g/dl.

We performed an abdominopelvic scan which revealed a left adrenal mass measuring 44 mm. This mass exerts compression on the tail of the pancreas and the gastric wall. (**Figure 2**).

After these arguments, the diagnosis of left PCM is made and the indication for surgery is retained. In view of his heart disease, the patient poses a therapeutic problem requiring close collaboration between surgeons, endocrinologists, intensivists and cardiologists with the aim of optimizing and securing his care.

Figure 2. abdominal CT, left adrenal mass in the arterial phase and portal phase



In order to have more details on his cardiac function, we performed an doppler ultrasound which showed a single ventricle ejection fraction of 63% and a filling fraction of 34% as well as minimal leakage of the systemic (AVV) and a patent construct with good flow.

The patient was prepared with an alpha blocker for 25 days and Osler antibiotic prophylaxis as well as an anti-flu vaccination. The patient was operated on under general anesthesia with continuous measurement of blood pressure (BP). He was approached through a left subcostal incision. This mass is firm and much vascularized. Its manipulation induced hypertensive peaks at 188/112 mm Hg; leading to the administration of a calcium channel blocker (Loxen type intravenously at a dose of 0.5 mg repeated three times).

The primary vascular control of the main adrenal vein (PSV) which was placed on the lake was facilitated by the identification of the left renal vein (**Figure 3A-3B**). Ligation of the VSP induced a drop in blood pressure to 73/50 mm Hg requiring vascular filling and the introduction of vasoactive drugs (Ephedrine 3 mg repeated 5 times).

We proceeded with the ligation of several other vessels emanating from dense neovascularization (**Figure 3C**). A lymph node dissection followed: inter-aortic-caval and latero-aortic sub- and supra-renal (**Figure 4**).

The postoperative course was marked by a normalization of blood pressure and heart rate. The patient was discharged on the 7th postoperative day.

Figure 3. dissection and intraoperative appearance of the adrenal mass-placed on lac of the main adrenal vein (3A, 3B, 3C)

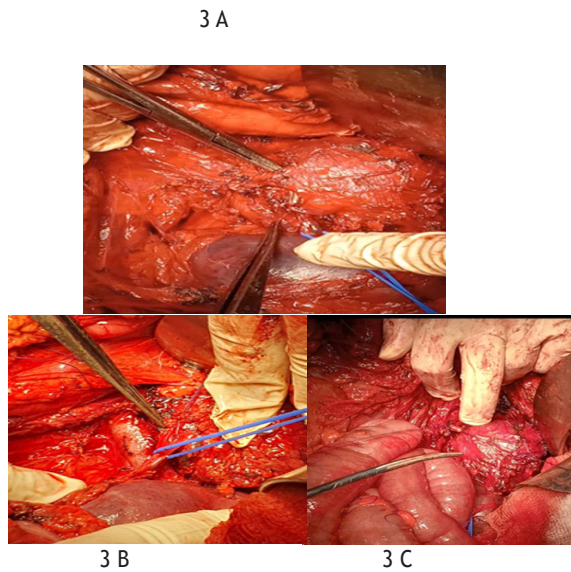
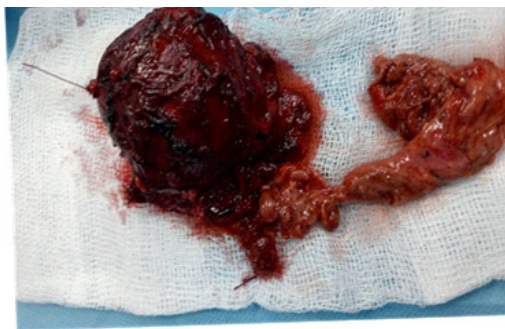


Figure 4. Adrenal mass with lymph node dissection



The pathological study confirms the neuroendocrine nature of the tumor by diffuse positivity of chromogranin A. The diagnosis of PCM is retained with a PASS score estimated at 08 (aggressive and potentially metastatic PCM). The first check-up was done at 3 months postoperatively, it is noted that the symptoms have disappeared. The methoxylated derivatives have normalized and imaging does not find any signs of recurrence or tumor remnant. He is currently at 16 months of follow-up and still without recurrence.

Discussion

Pheochromocytoma is a rare neuroendocrine tumor. Its prevalence is between 0.2% and 0.6% in hypertensive patients [1]. It represents 10% of AI as is the case in our patient [2], the attack is often unilateral. We looked for a syndromic context; our patient has a sporadic form as is the case in 75% [1].

Hypertension is present in 80-90% of patients. Blood pressure abnormalities can frequently be associated with orthostatic hypotension and the classic MENARD triad: headache - palpitation - sweating [3].

At CT scan the tumor is often larger than 5 cm with a spontaneous density (SD) greater than 10 HU found in 95% of PCM, which suggests a high negative predictive value if the (SD) is less than 10 HU [4]. Our patient presents a 54 mm tumor with a DS of 30 HU and an absolute wash-out of 20%.

Excision of a secreting and symptomatic PCM is the rule; the latter is all the more essential in a patient with congenital heart disease in view of the possible acute cardiovascular complications linked to hypersecretion.

In our patient, the immediate complication to fear is the decompensation of his heart disease due to heart failure and hypoxia.

The univentricular heart is a cardiac malformation characterized by the presence of a single ventricular cavity which, as in our patient, is a left ventricle in 80% of cases. This anomaly is two atrioventricular valves or a single valve by atresia of the other. It represents approximately 5% of congenital heart diseases. It is associated with chromosomal anomalies in 16% of cases and with major extra-cardiac anomalies in 20% of cases.

Cardiac disease was reported in 25 cases of PCM including only four cases of single ventricular heart. Patients with congenital heart disease are at high risk of developing complications related to chronic hypoxia. Does PCM represent one of these complications? [5]. There are two theories held in the literature. The first incriminates chronic hypoxia which would promote erythropoiesis and the proliferation of neural crest cells (embryological origin of the adrenal medulla) [6]. The second theory is genetic. PCM is a tumor that occurs in a context of autosomal dominant familial disease in approximately 25 to 30% of cases [7].

Around twenty predisposition genes have been reported, including SDHB, SDHD, VHL, RET and NF1 [7]. The genetic theory is suggested given the existence of analogies between chromosomal regions common to the two diseases [7].

The patient's preparation aims to prevent and limit the duration and severity of possible hemodynamic instability linked to excess hormonal secretions and their sudden correction by surgical excision [8]. According to the criteria of Roizen (1982) [8] reported by Ramakrishna [9], alpha blockers remain the treatment of choice.

Intraoperative hemodynamic changes can be major [10]. The main anesthetic challenge is maintaining hemodynamic stability [11].

Ligation of the last vascular pedicle preceding tumor excision leads to a sudden decrease in plasma concentrations of catecholamines. A decrease in blood pressure is a common occurrence. This can lead to collapse [12].

Postoperative hypotension is linked to the addition of residual effects of the preoperative preparation and the acute decrease in catecholamines. Most often blood pressure normalizes during the first 24 hours.

In 2002 Thompson introduced the PASS score (Pheochromocytoma of the Adrenal gland Scaled Score), in an attempt to develop a numerical scale to predict the metastatic potential of PCM [13]. Thompson defines a threshold at 4, beyond which a tumor is considered aggressive and therefore metastatic [14].

In addition to histo-prognostic scores, other risk factors for malignancy were established by the recommendations of the COMETE network for adrenal cancers in 2014.

These factors are: the size larger than 5 cm, dopaminergic and noradrenergic phenotypes, mutated SDHA and SDHB status, tumor multiplicity and diagnosis before the age of 20 [15]. Three of these factors are present in our patient: the size, dopaminergic phenotype and age of diagnosis at 18 years.

Conclusion

Decompensation of congenital heart disease should raise suspicion for the diagnosis of pheochromocytoma. The existence of this heart disease should not be a barrier to surgical excision of the PCM. Management must include perfect knowledge of heart disease and its intraoperative hemodynamic repercussions.

Competing interests

The authors declare no conflict of interest.

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