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Severe hypokalemia revealing a pheochromocytoma

[1] Hassan Ouleghzal, [2] Soufiane Belabbes, [3] Issam En-nafaa, [4] Abdelkrim Choho, [5] Soumia Safi

[1, 5] Endocrinology department, Military Hospital Moulay Ismail, Meknes, Morocco
[2, 3] Radiology department, Military Hospital Moulay Ismail, Meknes, Morocco

Abstract
Our case study concerns an observation of a 42 years old woman with pheochromocytoma. The initial clinical presentation was characterized by digestive signs related to severe hypokalaemia. During the evolution of the situation, the diagnosis of pheochromocytoma had been evoked later on, only after discovering an arterial hypertension. The 24 hours Urinary Catecholamine essay and Adrenal Imaging (CT and MRI) had confirmed this diagnosis. After surgical excision, the hypokaliemia was corrected and hypertension disappeared.

Keywords: hypokalaemia, arterial hypertension, pheochromocytoma

Introduction
Localized to the adrenal medulla, in almost 85% of cases, Pheochromocytoma is a rare neuroendocrine tumor composed of chromaffin cells which secretes catecholamines. Especially with young subject, this diagnosis is most often evocated in the context of a paroxysmal or permanent arterial hypertension, particularly when it is associated with Menard's clinical triad (headache, sweating and palpitations). Exceptionally, atypical clinical signs may be in the foreground, such as the case reported above. The initial assessment revealed just severe hypokalemia and the diagnosis of pheochromocytoma will be considered, later on, during the course of evolution.

Observation
A 43 years old woman with no previous clinical symptoms was admitted to the emergency for epigastric pain accompanied by vomiting and nausea. The symptomatology, which had been evolved during 15 days, had not been calmed by the symptomatic treatment.
Clinical examination at admission was normal apart from epigastric sensitivity to palpation (temperature at 37.5, blood pressure at 140/90 mmHg). The biological assessment revealed severe hypokalemia at 2.45 mmol / l. There was no inflammatory syndrome (C reactive protein: normal), the complete blood count was normal and the electrocardiogram showed ventricular repolarization disorders caused by the hypokalmia. Intravenous potassium supplementation under continuous cardioscopic monitoring was advocated to our patient, the evolution was marked by the appearance of high and very labile blood pressure (190/110mmHg).
With the association of hypokalemia and arterial hypertension, the diagnosis of adrenal tumor was evoked. Then, an abdominal CT was performed. It focused on a left adrenal mass of 58x45mm tissue density (36HU) delineating areas of suspected necrosis (Figure 1). Adrenal MRI showed an oval left adrenal mass of 62x49mm in hyposignal T1, a highly heterogeneous hypersignal T2 in favor of a pheochromocytoma (Figure 2). The hormone testing confirmed the diagnosis of pheochromocytoma with high urinary metanephrines results:

- Normetanephrine at 713 nmol / 24h (NV: 44-213);
- (3) ortho methyldopamine at 538 (NV: 88-320);
- Metanephrine at 167 nmol / 24h (NV: 40-228);
- The aldosterone/PRA ratio and urinary free cortisol were normal.

The patient was under calcium inhibitor, then entrusted to the surgeons for tumor excision (Figure 3). The anatomopathological study was in favor of a pheochromocytoma. The operative follow-up was simple with normalization of serum potassium value and blood pressure.

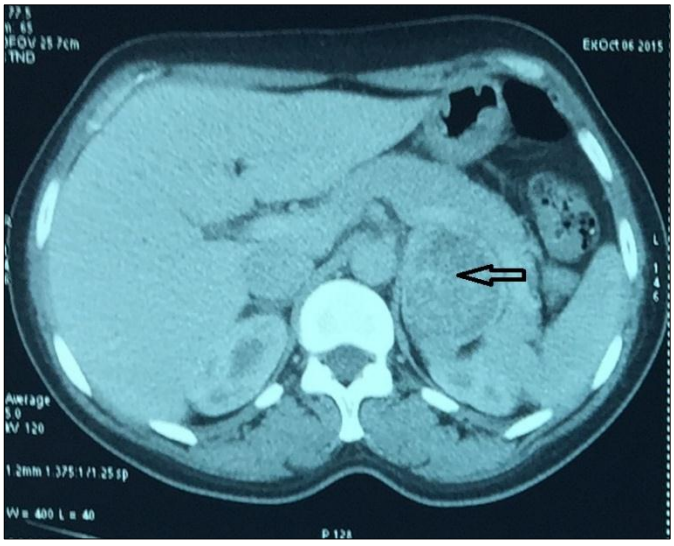

Fig 1: abdomen CT scan: axial slice showing a large heterogeneous mass of left adrenal with areas of necrosis

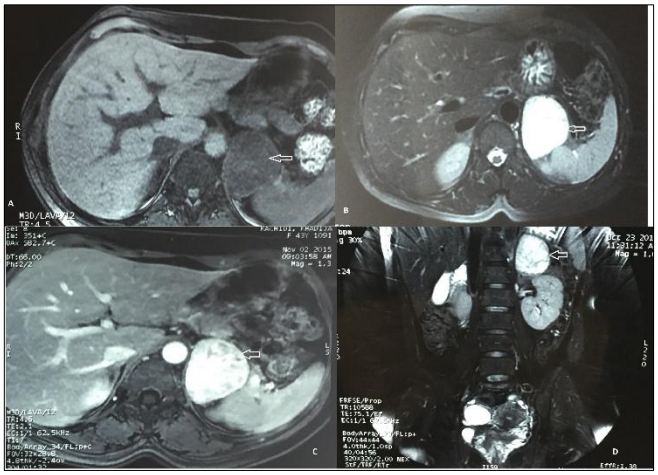

Fig 2: MRI of abdomen showing the slighlty hyposignal of the left adrenal mass on T1 weighted MR image (A),markedly hyperintense on T2 (B). Cystic and solid enhancing components are depicted in a post-contrast MR image on T1weighted with fat saturation; axial (C) and coronal (D)

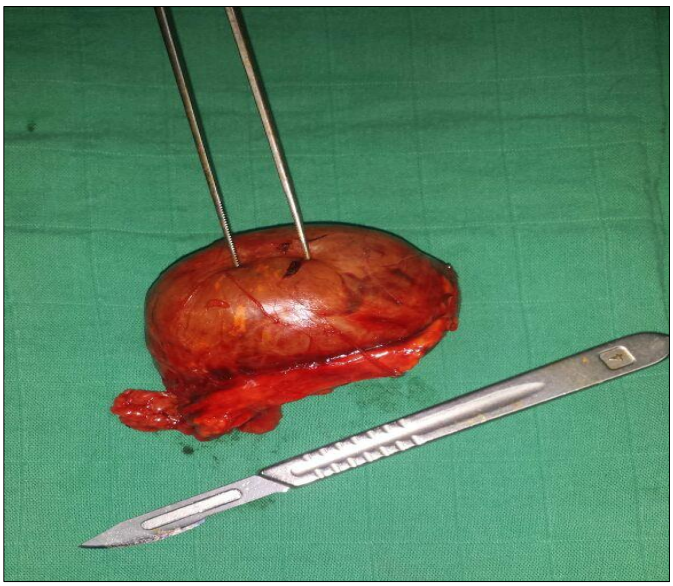

Fig 3: The tumor after resection

Discussion

Pheochromocytoma, due to the hypersecretion of catecholamines, is clinically manifested most often by an arterial hypertension associated with the classical triad (headache, palpitations, sweating) [1]. However, other signs might show in the foreground and the diagnosis of pheochromocytoma will be considered, later on, during the evolution of the situation. Indeed, other exceptional and atypical clinical manifestations can be a result of a pheochromocytoma: cardiovascular (arrythmia, cardiomyopathy, acute coronary syndrome and cardiogenic shock);neurological (isolated headache, cerebral or subarachnoid hemorrhage, and encephalopathy); pulmonary (respiratory distress, pulmonary edema); gastroenterological (abdominal pain and vomiting may indicate a hemorrhage or tumor necrosis, subocclusive syndrome, mesenteric ischemia, peritonitis, cholecystitis, pancreatitis); kidney disease (renal infarction, acute renal failure by rhabdomyolysis); metabolic (diabetes, lactic acidosis) or even pheochromocytoma multisystem crisis [PMC] [2, 3, 4, 5] In our case, the clinical symptomatology was dominated by digestive signs, initially related to hypokalaemia. During the evolution of the situation, an arterial hypertension was reported. Then the diagnosis of adrenal tumors was evoked. A hyper aldosteronism Primary by adenoma of conn seemed to be the most plausible diagnosis [6], but the tomodensitometric and MRI aspect of the tumor was much more in favor of a pheochromocytoma.

The discovery of adrenal morphological abnormalities was the instigator for hormonal determinations in order to establish their causal relationship with hypokalaemia and arterial hypertension. In this context, three diagnoses must be mentioned: a primary hyperaldosteronism, a subclinical cushing and finally a pheochromocytoma. The aldosterone/PRA ratio has the best diagnostic performance to screen for primary aldosteronism. It's considered as the pivotal test for the etiological diagnosis of hypertension with hypokalemia [7], the identification of a subclinical Cushing Is carried out by the calculation of urinary free cortisol [8] and the test of urinary metanephrines will lead to diagnosis a pheochromocytoma [9] as was the case of our patient.

On the physiopathological side, hypokalaemia is linked to a stimulation of the renin system angiotensin by the catecholamines. This secondary hyperaldosteronism is related to relative hypovolemia induced by the elevation of arterial pressure according to the pressure-natriuresis law, and by the stimulation of the beta and alpha1-adrenergic receptors of the glomerular juxtaticcells.

Among 30% of patients, pheochromocytoma may be a part of a family disease (type 1 neurofibromatosis, von Hippel Lindau disease, type 2 multiple endocrine neoplasia and hereditary paraganglioma) [6].

In the subgroup of Patients with a sporadic form of presentation (absence of family history or other lesion integrating into a family form) at the time of diagnosis, as was the case of our patient, a mutation was found in 10% to 15% of patients [10]. The genetic study was not performed in our patient.

The curative treatment of pheochromocytoma is based on surgical excision of the tumor [11]. The frequency of recurrences, especially loco regional and contra lateral, as well as the risk of delayed metastasis, can appear after some decades following the initial surgery. That's why the long-term monitoring of patients is required [12]. Follow-up one year postoperatively, no recurrence to be reported in the case of our patient.

Conclusion

Even in the absence of evocative clinical symptomatology, a diagnosis of pheochromocytoma should required in the case of patient who presented hypokalaemia and arterial hypertension.

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