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Malignant Pheochromocytoma - Disputable Differential Diagnostics and Follow-up Surgical Approach – Case Report

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Pheochromcytoma; Hypertension; Surgical methods

1. Introduction

Malignant pheochromocytomas are rare endocrine tumors that develop within chromaffin tissue. The diagnosis of malignancy is based on neoplastic recurrence or the presence of metastasis in organs that lack chromaffin tissues [1]. We present a case of 66-year-old patient treated 2 years for arterial hypertension with attacks of hypertension crisis after physical activity. Although with this signs that sound typical for hormonal active pheochromocytoma pre-differential diagnostics and treatment for hypertension was medically limited which let to diagnostic methods used, to make sure we wouldn't get mistaken the diagnosis for other case of disease manifested with hypertension.

2. Background

Pheochromocytomas are rare adrenal tumors arising from chromaffin cells of the adrenal medulla. The prevalence of malignant pheochromocytomas is estimated at 10%, this figure can vary between 5 and 26%, the malignancy of the tumor is judged at first diagnosis or recurrence. 40% of pheochromocytomas are of genetic origin which can be part of hereditary syndromes (multiple endocrine neoplasia type2, neurofibromatosis type1, von-Hippel-Lindau disease…). These clinical presentations have been reported in accordance with PROCESS 2020 [1,2].

The objective of the treatment of these malignant tumors is to improve the quality and survival of patients by controlling catecholamine secretions and reducing tumor volume. Adequate management requires a multidisciplinary consultation meeting [3,4,5]. Typical clinical manifestations are sustained or paroxysmal hypertension, severe headaches, palpitations and sweating resulting from hormone excess. However, their presentation is highly variable and can mimic many other diseases. If remaining unrecognized or untreated, they can be a life- threatening condition [6].

3. Pheochromocytoma and Hypertension

3.1. Clinical View

Pheochromocytoma has many clinical symptoms from asymptomatic forms up tp forms with clinical symptoms, it is because of releasing of catecholamines into the circulation either paroxysmally or continuously. They potential leans in in different factors such as blood pressure, changes in position of patient, increased physical activity and psychological activity, obstipation, anesthesia. But also impact of certain medications such as histamines, glucagons, adrenocorticoids [ACTH], Metoclopramide, Morphine, Naloxone and also tricyclic antidepressants. Among another signs belongs nervousness, anxiety, tremors, nausea, vomiting, headache, impairment of vision, paresthesia, and also abdominal pain which can imitate acute abdominal problem without defance musculaire.

Abdominal pain is localized commonly in right hypochondrium region and can be caused by hepatic stasis and sometimes lead to diagnosis of gall bladder colic [7].

3.2. Objective Signs

The basic objective sign is arterial hypertension, which can be either permanent or occur in paroxysmal forms [8]. It is not unusual rapid severely developing diastolic hypertension with tendency to malignant twist, which doesn't react to anti hypertension treatment. It occurs mostly in patients with increased secretion of noradrenaline [9].

3.3. Endocrine Forms of Hypertension

These forms belong to an important part of the secondary hypertension. Treatment of the backs disease could lead to modification of the hypertension. To this type of hypertension belongs hypertension by the cortisol over-release and mineral-corticoid overproduction, by high levels of estrogens, by acromegaly, by hyper and hypo-thyroidism, by pheochromocytoma. Endocrine based hypertension is accompanied by over-release of estrogens, diseases of thyroid gland – hyper-thyreosis, in lesser cases by hypo-thyreosis, hyper para-thyreosis, diseases of adrenal glands pheochromocytoma, hyperplasia, Conn or Cushing syndrome, congenital adreno-genital syndrome of acromegaly.

The difference of secondary hypertension from essential one is very important in practice for importance of possible specific therapy of the basic disease.

3.4. Hyper-Estrogen Conditions

The most common case of this condition is using estrogen oral contraceptives, which induces activation of renin-angiotensin-aldosterone system. The risk is in the amount on the estrogen dosage. There is 5% change in these women to develop hypertension with blood pressure over 140/90 mmHg. It depends on increased sensitivity on angiotensin II, previous kidney damage, hereditary predisposition for hypertension, age of the patient over 35 years, obesity (Table -1-3).

Table 1: ECG abnormalities, which occur in patients with pheochromocytoma

Sinus, nodal or arterial tachycardia
Atrial extra beats
Atrial flutter or fibrillation
Bradycardia with or without AV dissociation
P waves abnormalities
Depressions or elevations of segment ST with T wave
Prolonged interval PQ
Sick sinus syndrome [10].

Table 2: Endocrine forms of hypertension

1.Hyper-estrogene conditions
Usage of contraceptives
Gravidity
2. Adreno-cortical hyper functions
Cushing syndrome
Cushing disease
Primary hyper-aldosteronism
Congenital or hereditary adrenal-genital syndromes:
3. Pheochromocytoma
4. Hyper-thyreosis or hypo-thyreosis
5. Primary hyper-parathyreosis
6. Acromegaly [10]

Table 3: Types and Ethiology of hyper-cortisolism

Type of Hyper-Corticolism	Ethiology
Primary [with decreased production of ACTH]	
Cushing syndrome of adrenal ethiology	
CS within ectopic adrenal tissue	
Secondary [with increased production of ACTH]	
Cushing disease – over-production of cortisol is based on hypothalamo-hypophysal	basophilic or chromophobe adenomas of hypophysis malignant tumors of hypothalamus or hypophysis
Parameoplastic Cushing Syndrome	outcome failure of CRH from hypothalamus hyper-plasma of adrenal gland cortex based by peptide with ACTH similar effect, produced extra-hypophyseal tumor
Secondary [with decreased production of ACTH]	
Iatrogenic Cushing syndrome CS with relative hyper-cortisolism	after therapy with corticosteroids or ACTH of after decreased levels of transcortin [11]

3.5. Presentation of the Case

As mentioned above the patient was years treated with hypertension with occurring hypertension crisis with all the medical treatment spent. The only problem was that during all this time the patient was treated only for high blood pressure but wasn't examined by surgeon or physician for abdominal palpating, when in the first palpating there was huge tumor mass detected. In other diagnostic methods as ultrasound, CT there was confirmed tumor of right

adrenal gland and laboratory confirmation of decreased catecholamines in urine. In CT view there was significant tumor-caused decrease in lumen of vena cava inferior down to 10% of flow (Figure 1), without presence of distant metastasis and the local enlargement of lymph nodes up to 12 millimeters. Problematics of surgical treatment there is still a high-risk during operation which can lead to heart arrhythmia up to heart failure on the table. This risk is probable to decrease by specification of diagnostics of 3-D CT reconstruction in arterial-venous phase to specify artery supply of kidney tumor and adrenal gland (Figure 2). Operation technique lies in identification of arterial and venous structures and their ligation in reversed sequence as in typical nephrectomy or adrenalectomy. There is necessity to first ligate the vein system and then followed by ligation of the arterial system (Figure 3). This is physiological mechanism, that in occlusion of arteries, the organism reacts by sudden release of catecholamines, which this reversed method prevents catecholamine release to venous system and detection of flow through renal arteries. Histologically it came out as malignant pheochromocytoma without lymph node involvement.

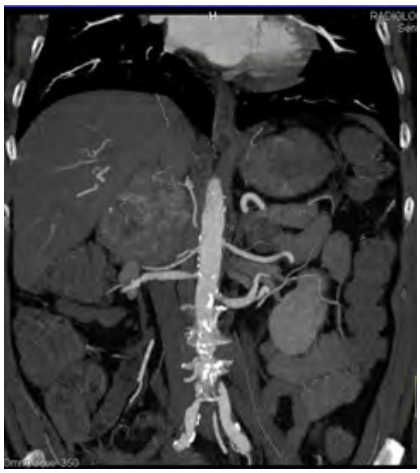

Figure 1: site of right tumor of adrenal gland

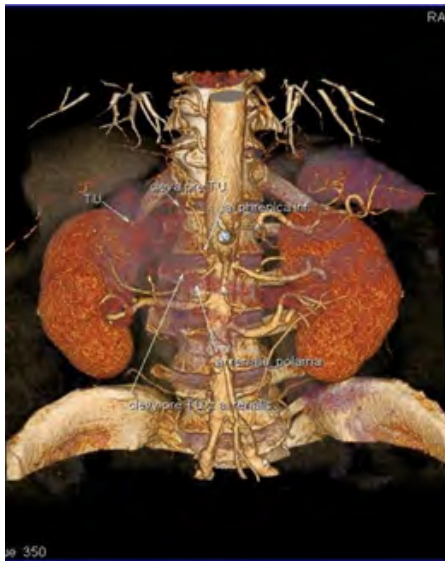

Figure 2: 3-D reconstruction

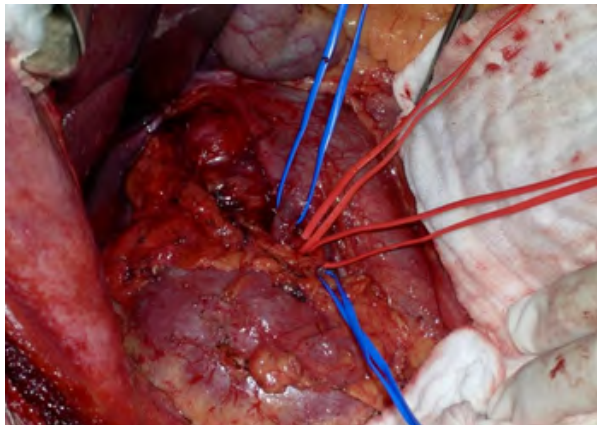

Figure 3: preparation of arterial and venous system

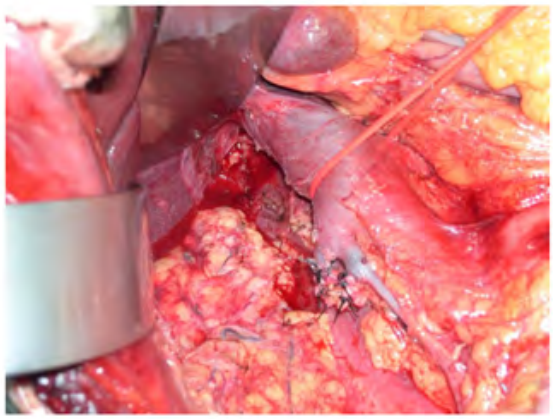

Figure 4: Status post nephrectomy and adrenalectomy

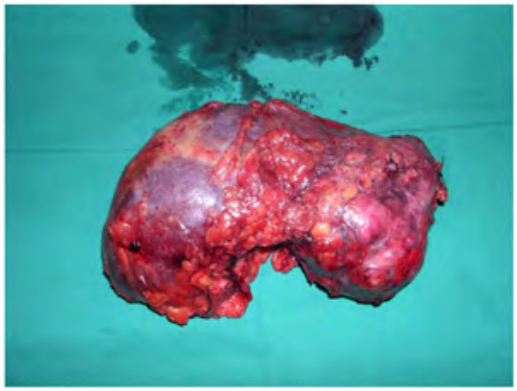

Figure 5: Resected right adrenal gland with kidney

4. Conclusion

The main prognostic factors of the malignant pheochromocytomas are a large tumor volume, the existence or number of visceral metastases, and the presence of a mutation in the SDHB (Succinate dehydrogenase B) gene.

Pheochromocytoma is a rare neuroendocrine tumor, its annual incidence is 2 to 8 per million adults. A peak frequency is observed between 30 and 40 years of age. Approximately 10% of pheochromocytomas are malignant and in 10% of cases, bilateral localization is observed [1]. Surgical resection is the definitive therapy for

benign and malignant pheochromocytoma. Adrenalectomy morbidity is as high as 40% and is attributed to pulmonary embolism, sepsis, cardiac arrhythmia and myocardial dysfunction. Mortality for adrenalectomy had dropped significantly to 2% [12].

5. Discussion

In treatment of arterial hypertension mostly, in cases of hypertension crisis after physical activity, there must be excluded hypertension as a result of other organ damage, failure or disease, renal artery stenosis and pheochromocytoma as the most common reason of secondary hypertension. In surgical treatment there is need to respect physiological renin-angiotensin- aldosterone mechanism renal function. There is need of gentle manipulation with the tumor and first of all the ligature of venous system to avoid releasing the enormous amount of catecholamines into the blood stream and that way, to avoid the hypertension crisis during the surgery. This kind of technique was used on 8 patient and even though this number is not significant in general, but in all the cases there wasn't any per-operative or post-operative complication and we had zero morbidity and mortality rate.

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