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Malignant Pheochromocytoma with Liver Metastases: About A Case

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Abstract **Case Report**

Pheochromocytomas are rare tumors, developed at the expense of the adrenal medulla and paraganglia. The excess of catecholamines secreted leads to clinical symptomatology. The diagnosis of malignancy is made only by the presence of metastases from organs devoid of chromaffin tissue or by the appearance of neoplastic recurrences. A 20-year-old woman was reported to have hypertensive crises with cerebral and ocular repercussions. A malignant pheochromocytoma of the left adrenal gland with hepatic metastases was diagnosed and the patient underwent left adrenalectomy with hepatic metastasectomy and surgical portal embolization in preparation for a subsequent right hepatectomy.

Keywords: Malignant pheochromocytoma; liver metastases; adrenalectomy; surgical portal embolization.

INTRODUCTION

Pheochromocytoma is a rare neuroendocrine tumor, developed at the expense of chromaffin cells of the sympathetic nervous system producing an excess of catecholamines. The adrenal pheochromocytoma accounts for 80% of all pheochromocytomas, it is most often unilateral, but in 10% of cases a bilateral localization is observed.

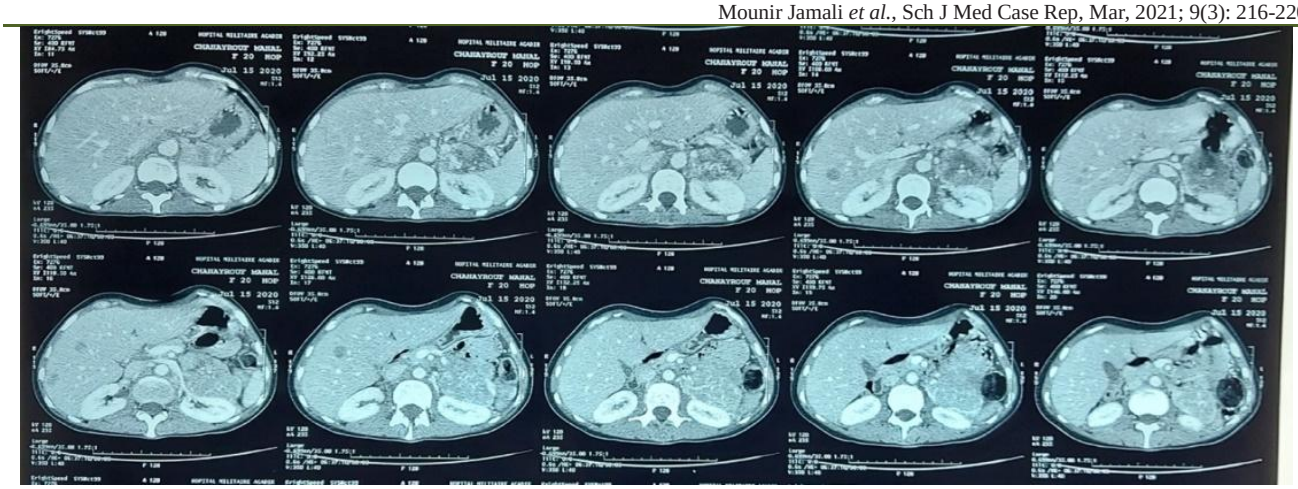
Malignancy is defined by the presence of chromaffin tissue in the vessels, neighbouring organs by contiguity, adjacent lymph nodes and distant metastatic sites and/or bone.

Symptoms are due to catecholamine overproduction or mass effect. The diagnosis is confirmed by elevated levels of methanephrine or normetanephrine in plasma or urine. Radiology helps locate the tumor and any local invasion or metastasis. All patients should have preoperative preparation with α blockers and/or other drugs to control hypertension, arrhythmia and volume expansion. Surgery is the definitive treatment. Follow-up should last a lifetime.

OBSERVATION

Mrs. M.C., 20 years old, history of neuraxitis with ophthalmologic sequelae. The patient presented episodes of headache with vertigo, jet vomiting and profuse sweating, which prompted her to consult the emergency room. Clinical examination revealed a blood pressure of 260/140 mmHg with tachycardia at 180b/min.

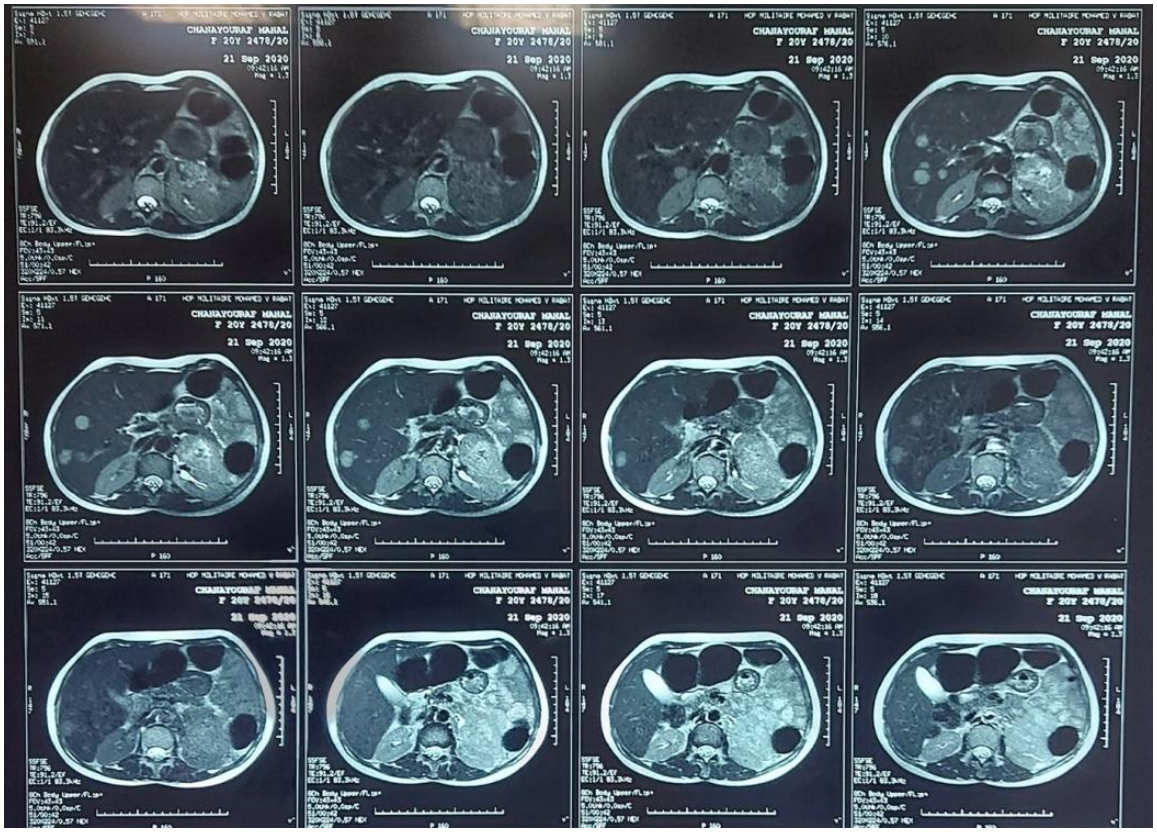
A thoraco-abdominal-pelvic CT scan was requested, which came back in favor of a malignant pheochromocytoma with multiple liver metastases.



The biological workup revealed normal cortisolemia at 506.6nmol/l, normal urinary free cortisol at 30nmol/24h, elevation of free metanephrines: Normetadrenaline >32nmol/l, metadrenaline: 1.41nmol/l, 3 OrthoMethyldopamine: 0.12nmol/l.

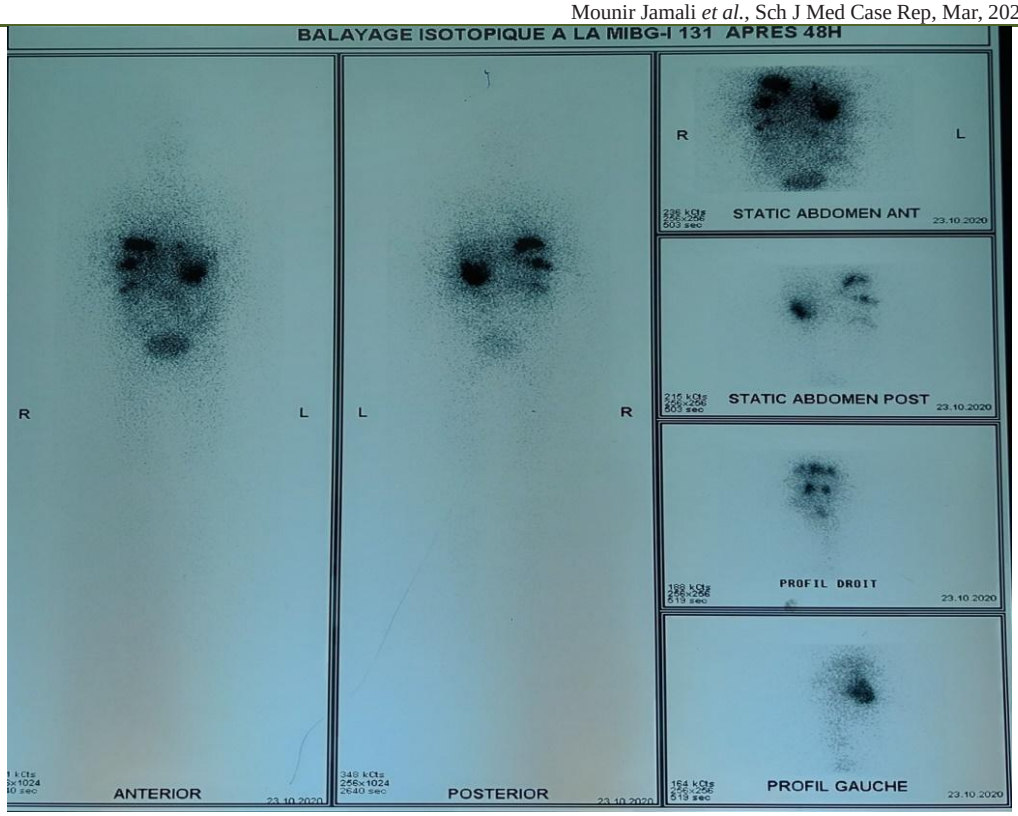
An elevation of urinary free metanephrines: Normetadrenaline > 4500nmol/24h, metadrenaline: 357nmol/24h, 3 OrthoMethyldopamine: 210nmol/24h. Aldosteronemia collected supine was elevated: 2237pmol/l with a normal aldosteronemia/renin ratio of 15.08. Active renin was also elevated at 82.30pg/ml.

A Uro-IRM was requested and concluded that there was a left adrenal lesion process invading the anterior border of the kidney and in contact with the renal pedicle, with liver metastases.

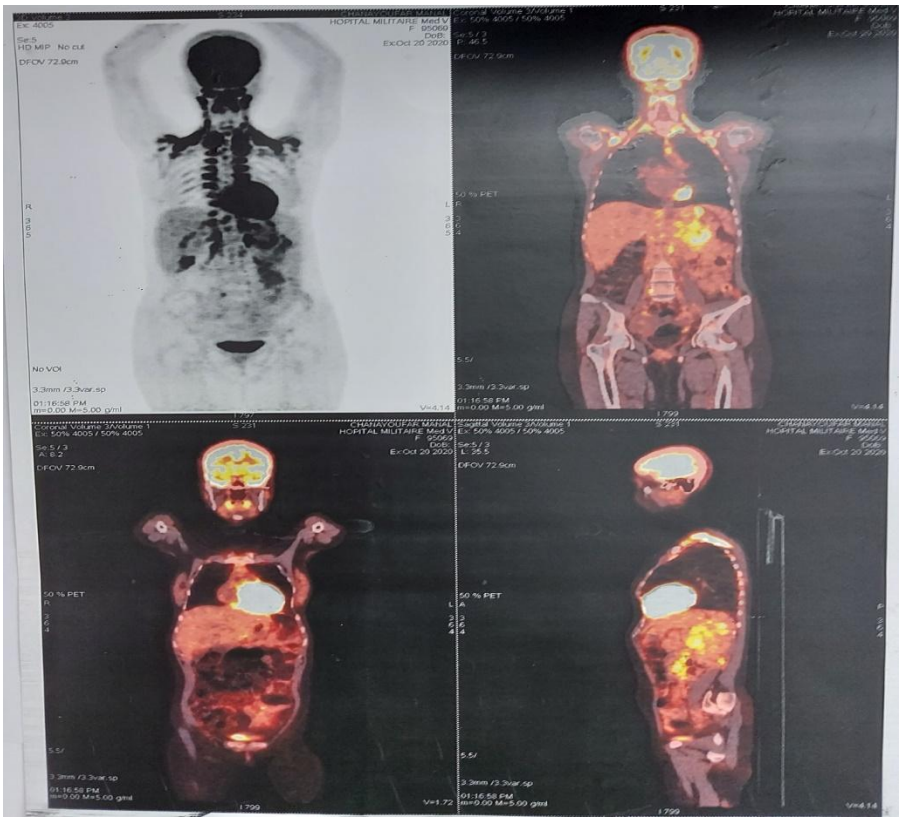


Cerebral MRI found some punctiform signal abnormalities of the supratentorial white matter with a rather vascular appearance.

MIBG scintigraphy performed revealed hyperfixation over the left renal area persisting on images 24 and 48 hours after injection with hyperfixation of the dome and segments IV, VI, VII.

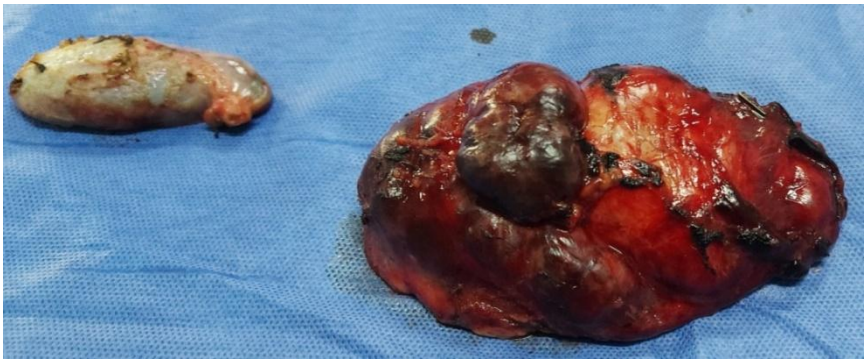


The PET Scan found a heterogeneous pathological hypermetabolic mass of the left adrenal area, poorly limited, pushing back the left kidney with discrete hypermetabolisms involving the dome and several hepatic segments and a bilateral and symmetrical hypermetabolism of the cervical, axillary, paravertebral and peri-renal fat along the sympathetic territories, linked to an activation of brown fat secondary to adrenergic stress.



During her hospitalization, the patient presented a decrease in visual acuity and visual fog, the ophthalmologic examination concluded hypertensive retinopathy.

After cardiologic, endocrinologic and pre-anesthetic consultation, the patient was placed on alpha- and beta-adrenergic blockade in preparation for surgery. The patient benefited after extensive exploration from an open left adrenalectomy with resection of liver metastases, cholecystectomy and ligation of the right dividing branch of the portal vein to prepare the patient for a subsequent right hepatectomy. The procedure was completed without incident and blood pressure remained within desirable limits.



The postoperative period was marked by hypertensive peaks despite the treatment α and β blocking and normalization of the blood pressure figures after a few days.

The result of the anatomopathological and immunohistochemical study came back in favor of a PASS pheochromocytoma score 4 for the adrenalectomy piece but also for the resected liver metastases.

DISCUSSION

Pheochromocytoma is a rare endocrine tumor derived from adrenal medullary chromaffin cells. It is generally accepted that 90% of cases occur sporadically, while 10% have a familial incidence, fitting within the framework of NEM types 2A and 2B, or associated with neurofibromatosis type 1 or Von Hippel-Lindau disease [1].

Malignancy occurs in about 10% of cases, with a height greater than 5 cm, local invasion, internal necrosis, pleomorphism and nuclear hyperchromism being suspect features of malignancy, however, the only criterion used is the existence of secondary localizations devoid of chromaffin tissue [2].

Biological diagnosis is essentially based on the determination of plasma and urinary methoxylated derivatives, while the determination of plasma and urinary catecholamines has been abandoned as a first-line screening method. Plasma methoxytyramine is a predictive marker of pheochromocytoma malignancy [3].

A genetic study is recommended in cases of suspected genetic disease or in cases of bilateral pheochromocytomas and in patients under 45 years of age [4].

The radiological diagnosis is based on CT scan with a spontaneous density of malignant tumors greater than that for benign tumors, scintigraphy with MIBG labeled with iodine 123 has been supplanted by PET with 18F-DOPA especially for the detection of secondary localizations with a sensitivity close to 100% [5].

Pre-operative preparation is a crucial time, in order to avoid arrhythmia and hypertensive crisis on the operating table secondary to a mass release of catecholysis. mines during the induction of anesthesia or during the manipulation of the tumor by the surgeon. Blocking of α and β-adrenergic receptors is necessary.

The surgical management of a malignant pheochromocytoma with metastases that can be cured by total eradication requires adrenal surgery with focal treatment of metastases or metastasectomies. Chemotherapy, on the other hand, is aimed at non-operable or non-resectable forms. In the case of liver metastases, preoperative transepatic portal embolization appears to be an excellent option for postoperative safety, and may prolong survival or the symptom-free period [6].

CONCLUSION

The malignant pheochromocytoma is a rare entity whose management requires multidisciplinary collaboration. The surgical risk is important even with adrenergic blockade but remains the pillar of therapeutic management.

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