

Malignant Pheochromocytoma and neurofibromatosis type 1

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Introduction: Patients with neurofibromatosis (NF) are at higher risk for tumors' development . Pheochromocytomas are rare in NF type 1 (NF1), and malignant ones are even rarer. We aimed to report a malignant pheochromocytoma in a patient with NF1.

Case report: A 24 year-old female, whose father had colonic cancer, consulted in 2013 for abdominal pain and vomiting. Clinical examination pleaded for NF1 (multiple "café-au-lait" spots, and freckles). Blood pressure and the rest of clinical examination were normal. Echosonography discovered a right heterogeneous adrenal tumor measuring 6cm. Biological exploration showed normal urinary catecholamines (0.87ng/ml n<1) . Abdomen CT scan confirmed the solid and cystic tumor (Fig1). MIBG scintigraphy pleaded for right adrenal uptake. Work up for metastases was negative.



Fig1:CT scan showing the 6cm right adrenal tumor.

Treatment and follow up. After surgery, histological examination confirmed the pheochromocytoma with a Pass score of 4%. Post operative work up was negative.

9 months later, although both adrenals were free of tumors, urinary normetanephrines increased : 3750nmol/24H (n=44-231), and diffuse bone (fig2) and medullary metastases with MIBG uptake were observed (Fig 3).

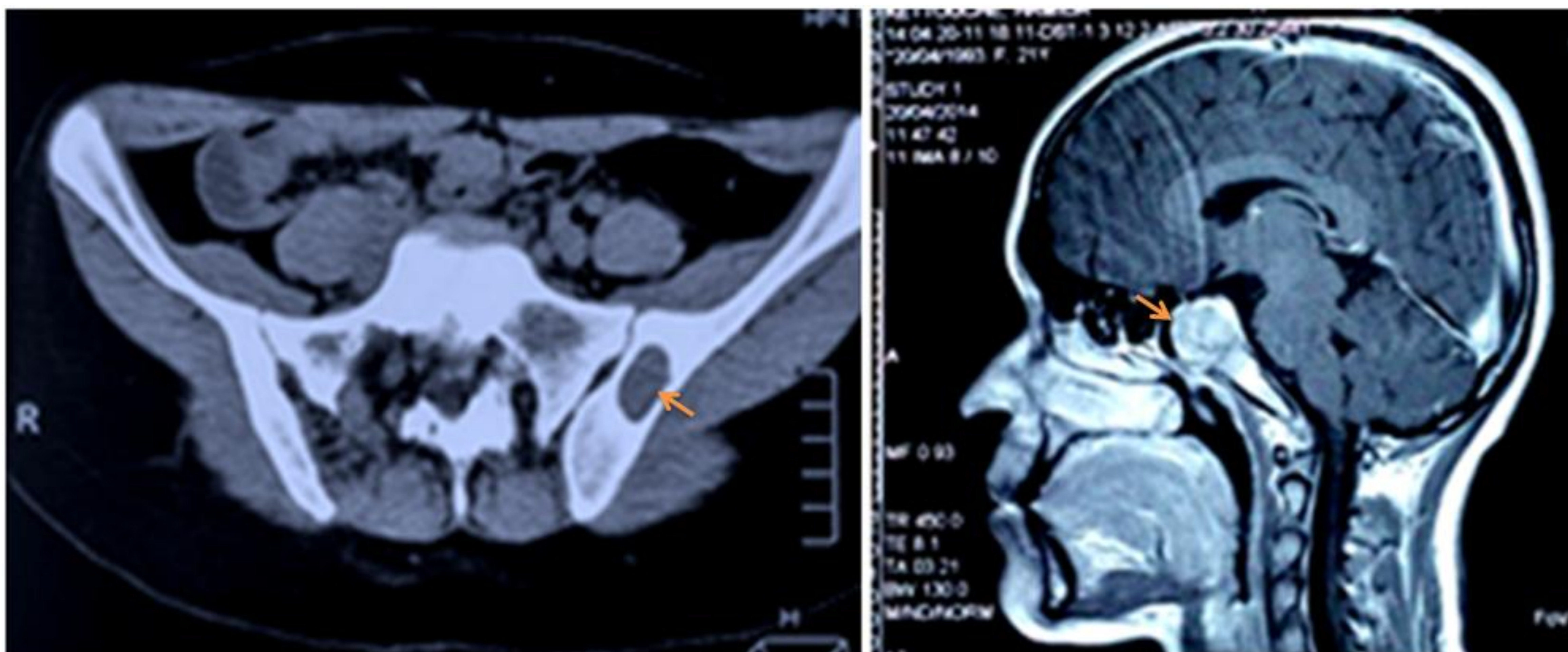


Fig 2 showing metastases in iliac and sphenoid bones

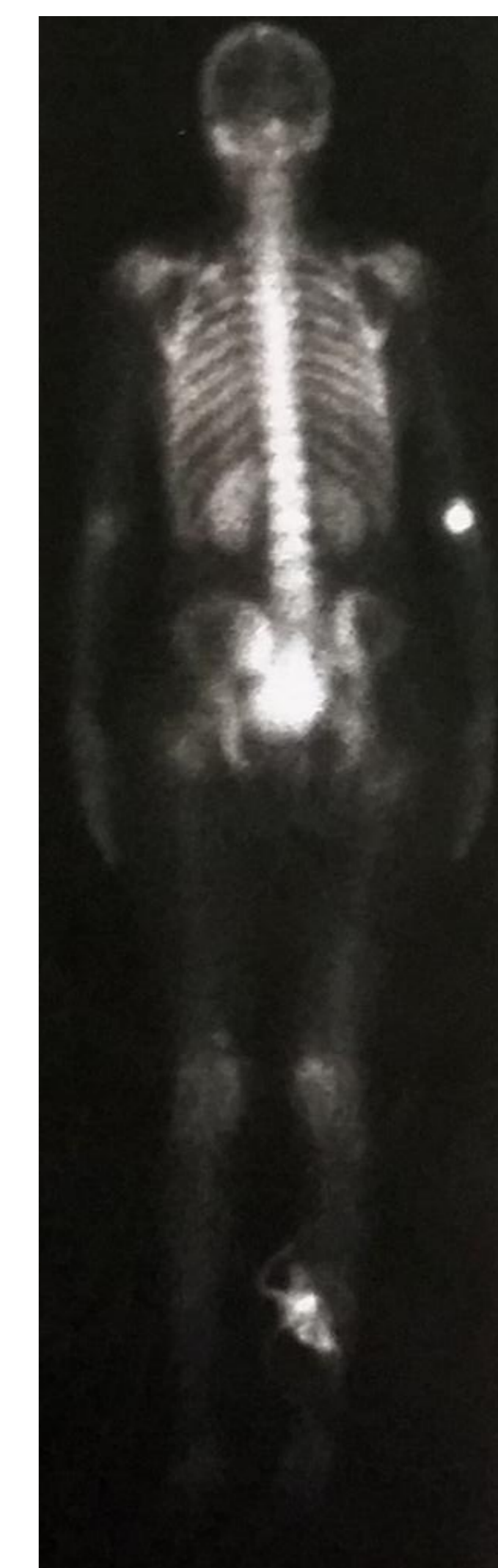


Fig.3 MIBG scan showing diffuse uptake

Conclusion: The association of malignant pheochromocytoma with NF1, although very rare, should be known as a pheochromocytoma and its metastases may be silent as in the presented case.