Sclerotic thoracic vertebral compression with metastatic pheochromocytoma.

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Letters

Sclerotic Thoracic Vertebral Compression with Metastatic Pheochromocytoma

Symptoms of pheochromocytoma are frequently due to elevation of peripheral catecholamines and hypertension, but some pheochromocytomas metastasize and cause unusual clinical presentations [1]. Metastases may produce hormones, and venous sampling [2] has been increasingly useful in localization of multiple primaries and metastases.

A 20-year-old man with numerous café au lait spots and a family history of neurofibromatosis was found to have variable hypertension. He had a history of pathologic fracture of the femur due to multiple medullary cystic lesions. Elevated catecholamines and a left suprarenal mass were demonstrated. At surgery a left suprarenal pheochromocytoma with a neuroganglioblastoma element and a paraaortic paraganglioneuroma were found.

Back pain developed 1 year after surgery. A compression fracture of T11 was found. Visceral angiography and venous sampling were unsuccessful in defining the source of the catecholamines. Three years later he developed progressive spastic paraplegia with sensory and bowel changes consistent with a lower thoracic lesion. Plain radiography demonstrated a sclerotic compression fracture of T11 and a right paraspinal mass that had developed since the initial evaluation. The vertebral body was smoothly scalloped and the pedicles were poorly defined. Pantopaque lumbar myelography and metrizamide from above showed a complete extradural obstruction at T11–12 interspace (fig. 1). Radionuclide scans of bone, spleen, and liver failed to demonstrate other peripheral lesions.

The differential diagnosis included a neurofibroma, osteomyelitis, and metastatic pheochromocytoma. A metastatic pheochromocytoma was considered relatively unlikely because nearly all metastatic pheochromocytomas are osteolytic.

At surgery a fibrous tumor was found surrounding T11, permeating the centrum, and filling half the spinal canal. Surgical manipulation caused fluctuation in blood pressure. Pathologic examination showed metastatic pheochromocytoma with invasion of blood vessels and bone. Following surgery the strength in his legs improved but his hypertension was changed little. A venogram showed irregular involvement of the inferior vena cava.

Most pheochromocytomas arise within the abdomen. Many are multiple and 10% metastasize [3, 4]. Metastatic pheochromocytoma is defined as the presence of chromaffin tissue where it is not normally present, with evidence of hormones within or secreted by the tumor [5]. The most common metastatic site is bone [6], followed by liver, spleen, and lymph nodes. Benign appearing tumors may metastasize [7]. There is increased incidence of pheochromocytoma and medullary bone cysts [8] with neurofibromatosis. Increased catecholamines from metastases may persist after the primary tumor has been removed [9]. Venous sampling may locate functioning metastases not apparent clinically or radiologically. Both lytic and sclerotic metastases have been described in a single patient, though the destructive lesions predominated [10]. Only rarely is there an osteoblastic response. The unusual feature in this case is the progressive sclerosis of this solitary metastasis. The course of metastatic pheochromocytoma is variable, and long survivals are, as in this case, occasionally seen.

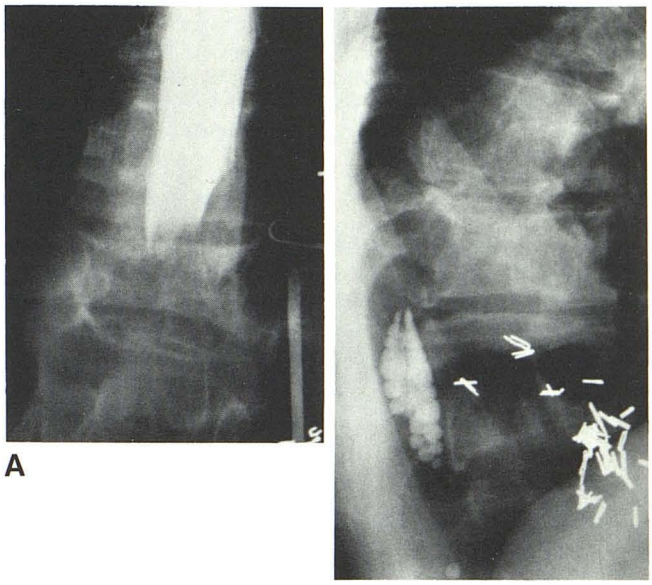

Fig. 1.—**A**, Lower thoracic spine after introduction of metrizamide by C1–2 puncture. Obstruction of subarachnoid space at T10–11. Compression and sclerosis of T11 thoracic vertebral body and paraspinal mass. Poorly defined pedicles and deviation of subarachnoid space from left to right. **B**, Lateral film. Compression, sclerosis, and smoothly scalloped margins of T11 vertebral body. Obstruction at T11–12 interspace demonstrated by lumbar injection of Pantopaque. Surgical clips related to prior adrenalectomy.

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