Solid pseudopapillary tumor of the pancreas mimicking malignant adrenal pheochromocytoma

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METCALFE PD, GUPTA R, NORMAN RW. Solid pseudopapillary tumor of the pancreas mimicking malignant adrenal pheochromocytoma. The Canadian Journal of Urology. 2004;11(2):2223-2225.

We report a 75-year old woman with clinical, laboratory and imaging characteristics suggestive of a malignant left adrenal pheochromocytoma with invasion of the tail of the pancreas. The mass involving the tail of the pancreas and the left adrenal was excised Detailed histological diagnosis revealed that the lesion was a rare exocrine tumor of the pancreas.

Key Words: solid pseudopapillary tumor of pancreas, pheochromocytoma

Introduction

Solid pseudopapillary tumor (SPT) of the exocrine pancreas is rare, usually benign and highly curative.[1,2] It occurs primarily in young women. We report on a 75-year old woman with such a tumor involving the tail of the pancreas in association with clinical, laboratory and imaging characteristics suggesting a left adrenal pheochromocytoma.

Case report

A 75-year old woman presented with abdominal discomfort and urinary frequency. Past medical history included poorly controlled hypertension, occasionally associated with headaches, dizziness and syncope. Abdominal ultrasound suggested an abnormality in the right adrenal gland. An un-enhanced CT scan confirmed a 2.5 cm mass within the right adrenal gland and a 7.7 cm mass (40 Hounsfield units) arising from the left adrenal gland or tail of the pancreas; there was no evidence of metastatic disease. MRI suggested a probable left adrenal pheochromocytoma (i.e. bright fat saturated T_2-weighted image) and non-functioning right adrenal adenoma Figure 1. Adrenal I-123 MIBG scan did not show uptake in either adrenal. Radiological picture was that of a mass probably arising from the left adrenal with invasion of the pancreas. Chest x-ray showed cardiomegaly and no lung lesions.

Three 24-hour urine collections were made. Epinephrine, norepinephrine, dopamine, free cortisol and amylase were normal but VMA was elevated on two of the three collections (5.29 and 4.66 mM/Mcr; normal 0.86-4.00 mM/Mcr).

Based on the clinical history, laboratory data and imaging characteristics, it was assumed that the mass was a malignant adrenal pheochromocytoma. Following pre-operative alpha blockade, the patient underwent en bloc excision of the left adrenal gland and tail of the pancreas.

Gross pathology described a 9.5 cm, 219 g nodular mass Figure 2a. Microscopic sections showed nodular adrenal cortical hyperplasia and a small myeolipoma.

Accepted for publication February 2004

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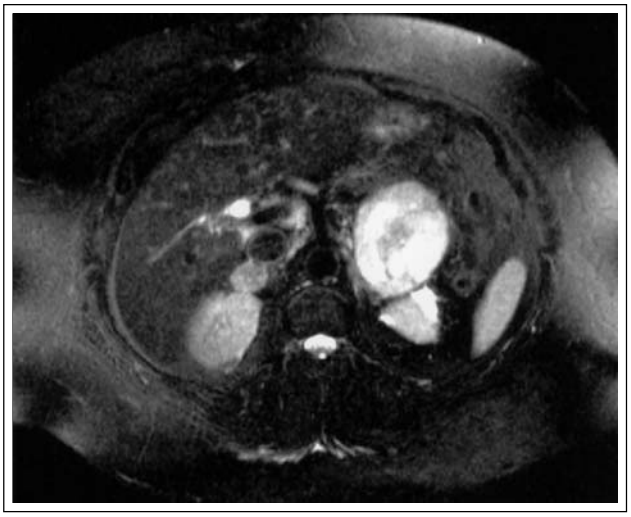

Figure 1. MRI shows a 7.8 cm mass intimate to the left adrenal gland with very bright fat saturated T2-weighted images raising the probability of a pheochromocytoma.

The extra-adrenal mass was encapsulated with focal necrosis, fibrosis and cystic changes showing tumor in a pseudo-papillary pattern Figure 2a and 2b. Vascular invasion of adjacent stroma was not identified Figure 2c. Pancreatic tissue (ducts, exocrine cells and islet cells) was seen on the surface. Positive staining with vimentin, a-1 antitrypsin and neuron specific enolase Figure 2d with negative stains for chromogranin, EMA, alpha actin, S-100 protein, monokeratin, CK-7, CK-20 and cytoplasmic mucin helped differentiate the tumor. The final diagnosis of this mass with attached pancreatic and adrenal tissue was SPT of the exocrine pancreas.

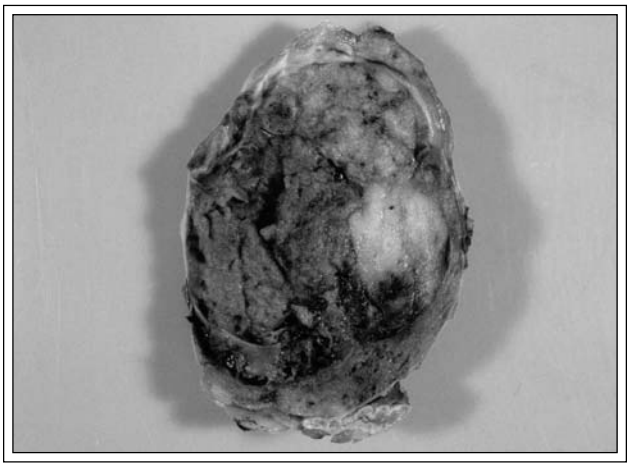

Figure 2a. Tumor is well-circumscribed, capsulated, tan to brown solid mass with hemorrhage. Adrenal gland is adherent to tumor surface.

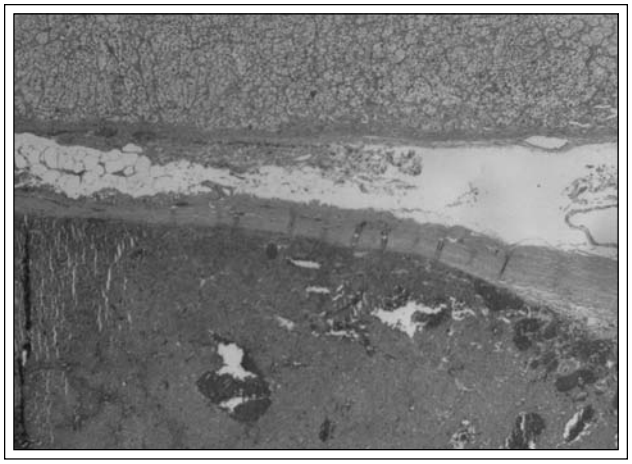

Figure 2b. Encapsulated solid extra-adrenal tumor mass (H&E X20).

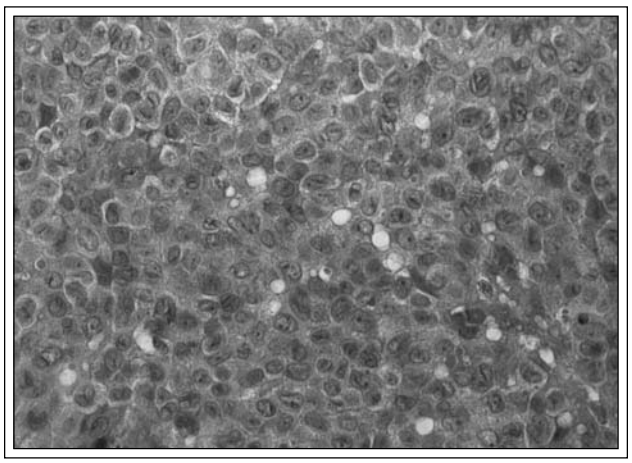

Figure 2c. Tumor with solid sheets of cells showing eosinophilic cytoplasm and prominent nuclear grooves (H&E X 400).

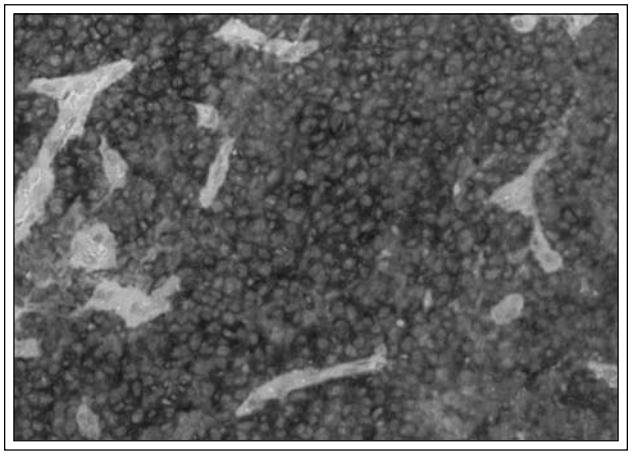

Figure 2d. Immunohistochemical reaction is positive for neuron specific enolase

Discussion

The imaging characteristics of SPT have been poorly defined and its proximity to and possible confusion with an adrenal pheochromocytoma have not been described previously. Our patient's hypertension and mildly elevated urinary VMA made us suspicious for a pheochromocytoma. There are no descriptions of the MRI features of SPT of the pancreas. This rare pancreatic tumor, when present in the tail of the pancreas, may mimic a malignant left adrenal pheochromocytoma. Our patient has demonstrated no evidence of recurrence on abdominal CT at 1 year. □

References

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