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PANCREATIC SCHWANNOMA: A CASE REPORT

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ABSTRACT

Schwannomas may occur anywhere in the soft tissues or viscera, but do not commonly involve the pancreas and retroperitoneum. We present a case of benign pancreatic schwannoma which clinically simulated a malignant neoplastic process.

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INTRODUCTION

Most primary pancreatic neoplasms, both benign and malignant, are of epithelial origin. Non-epithelial tumors arising from indigenous structures in the pancreas do occur but are extremely rare.[11] Sporadic cases of fibroma, lipoma, lymphangioma, leiomyoma, hemangioma and fibrous histiocytoma have been described.[6,7] Schwannomas, tumors derived from the schwann cells enveloping peripheral nerves, can be benign or malignant. Solitary benign or malignant schwannomas can arise in any anatomic site in the body, however pancreatic involvement is rare. To the best of our knowledge, only 15 cases of pancreatic schwannoma have been reported in the English literature.[4,6,7,8,11]

We report a case of benign solitary pancreatic schwannoma presenting as an abdominal mass simulating a malignant neoplastic process.

CASE REPORT

A 45 year old previously healthy woman presented with abdominal discomfort, intermittent left upper quadrant pain and about 5 kg weight loss. She had no history of nausea, vomiting, gallstones, trauma, jaundice or pancreatitis.

Physical examination revealed a mildly obese woman with no acute distress. There was a large mass in the left upper quadrant. The remainder of the physical examination was unremarkable. Laboratory tests revealed normal hematocrit, amylase and liver enzymes.

Abdominal ultrasound revealed a 95 mm × 92 mm homogenous mass, located retroperitoneally, anterior to the spleen, posterior to the stomach and near the tail of the pancreas. There was no pancreatic or biliary duct dilatation and no adenopathy.

With an impression of a malignant process operation was done. At operation, there was a well-defined mass with no surrounding inflammation in the pancreatic tail and adherent to the posterior wall of the stomach. The mass was excised. The specimen was sent for pathological examination. There was an oval shaped mass measuring 11×10×8 cm with a rim of pancreatic tissue. The external surface of the mass was creamy white and smooth. It was well circumscribed. The cut surface showed a partially encapsulated mass with firm, creamy white homogenous tissue and minute hemorrhagic areas (Fig. 1). The microscopic sections revealed a schwannoma with a classic Antoni A&B pattern composed of woven spindle cells with occasional pallisading of the nuclei and formation of Verocay bodies (Figs. 2&3).

The patient followed an uncomplicated postoperative course, leaving the hospital one week after operation.

DISCUSSION

Neoplasms of the pancreas are classified according to their histologic origin. While the great majority of both benign and malignant pancreatic neoplasms arise from pancreatic epithelial cells, mesenchymal tumors, although rare, can arise from the vascular, neuronal, lymphatic and connective tissues of the pancreas.[6,11]

The schwann cell is the supporting element of the pe-

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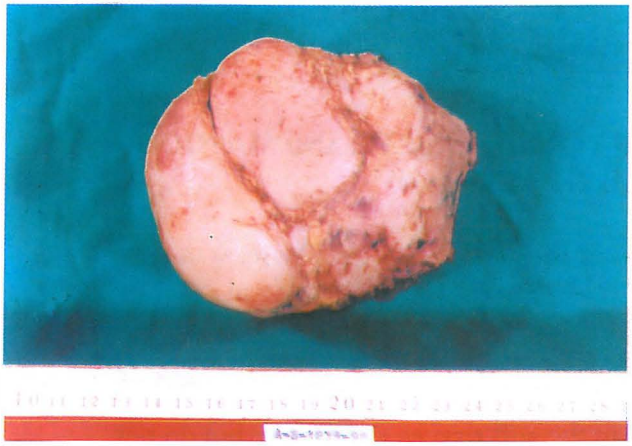

Fig. 1 (A). Well-circumscribed mass with smooth creamy white external surface.

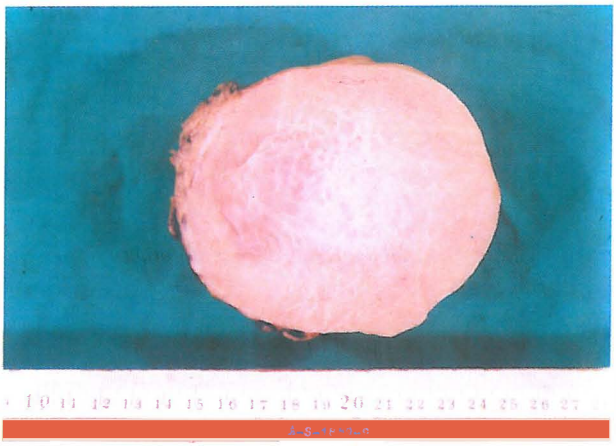

Fig. 1 (B). Cut surface of tumor with rim of pancreas.

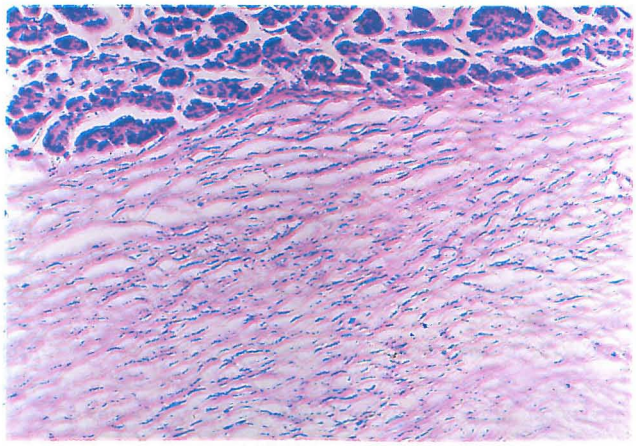

Fig. 2. Tumor with woven spindle cells, surrounded by normal pancreatic rim (H & E stain, ×250).

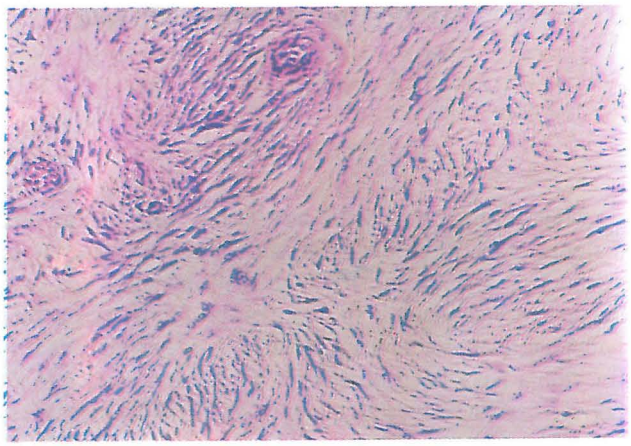

Fig. 3. Antoni A area with Verocay body (H & E stain, ×250).

ripheral nerve. These cells produce and maintain the myelin sheath, and enclose and protect the axons of myelinated neurons and help the regeneration of nerve fibers.[6] The schwann cells give rise to two types of tumors: the neurofibroma and the schwannoma or neurilemmoma. Malignant schwannoma is, in contrast to neurofibroma, an exceptionally rare event.[12]

Schwannoma is one of the few truly encapsulated neoplasms of the human body and is almost always solitary. Its most common locations are the flexor surfaces of the extremities, neck, mediastinum, retroperitoneum, posterior spinal roots and cerebello-pontine angle.[12,13] The tumor can vary in size from a few millimeters to >20 cm but usually measures <5 cm. The deeply situated tumors of the retroperitoneum and mediastinum can grow much larger and may manifest secondary degenerative changes such as cyst formation, hemorrhage, hyalinization and calcification.[9,10,12-14]

Microscopically, the schwannoma consists of two alternating components: an organized cellular component consisting of long bipolar cells that often form a pallisading arrangement known as Verocay bodies (Antoni A area) and a loose hypocellular component (Antoni B area).[6] Immunohistochemically, the tumor cells exhibit intense and uniform staining for S-100 protein, which helps distinguish it from the neurofibroma.[6,12]

The distinction between schwannoma and neurofibroma is important because neurofibromas have a significant association with Von Recklinghausen's disease, in which there is a risk of malignant transformation.[6,12] Benign schwannomas are most common between 20-50 years of age with a near equal male: female ratio. Although presenting signs and symptoms of solitary benign schwannomas vary according to anatomical site, most patients present with a painless slowly growing mass.[6]

Pancreatic schwannomas can arise from the parasympathetic or sympathetic division of the autonomic nervous system present in the pancreas.[6]

Pancreatic schwannomas are rare. Only fifteen cases have

been reported previously in the English literature, of which eleven cases were benign schwannoma.[1,2,16] Two cases of solitary malignant pancreatic schwannoma have been reported.[5,10]

In addition, two cases of pancreatic schwannomas in patients with Von Recklinghausen's disease have appeared in the literature.[6,15]

Including the present case, there have been eight men and eight women. The patients ranged in age from 40 to 87 and often presented with a history of left-sided abdominal pain. The tumors were distributed throughout the gland but were more common in the proximal pancreas than in the tail.[6]

Eight of these tumors had some cystic components on preoperative radiologic imaging.[9,10,13,14] On CT scan, the tumors are usually well defined and encapsulated. Tumors with high Antoni A areas usually look inhomogenous due to high lipid content. In tumors with high Antoni B components, the poor cellularity and loose stroma can cause a cystic or multiseptated appearance on CT scan. The presence of both components in the same tumor also leads to inhomogeneities on CT scan.[7] While the final diagnosis of any pancreatic mass is made after operative excision, preoperative imaging employing multiple modalities can be helpful in narrowing the broad differential diagnoses.[3,6]

The treatment of nonpancreatic benign schwannoma consists of surgical excision from the underlying nerve trunk.

Adequate treatment of solitary benign pancreatic schwannoma consists of simple enucleation of the encapsulated tumor from the surrounding pancreatic parenchyma.

Intraoperative consultation with a pathologist may help establish the diagnosis of benign schwannoma on frozen sectioning. This makes a significant contribution to the selection of the operative intervention, limiting it to enucleation rather than radical resection.[7]

CONCLUSION

Although rare, the nonendocrine nonepithelial tumors of the pancreas must not be neglected in the approach to the patient with a pancreatic mass, since their identification has biologic and prognostic significance. While the definite diagnosis is often made following surgical excision, preoperative evaluation including multiple radiologic techniques as well as percutaneous aspiration cytology, may help in defining the origin of the tumor and its location in the pancreas.

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