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Case Report

Solid and Cystic Epithelial Neoplasm of Pancreas with Metastasis: report of a highly unusual case

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Abstract

Solid and cystic papillary epithelial neoplasms of the pancreas are uncommon tumors occurring predominantly in young women. These tumors have excellent prognosis and after complete surgical resection, more than 95% patients are cured. Occasionally, they invade the surrounding pancreatic parenchyma. These tumors can recur even many years after resection, so long term follow up is essential. Very few cases metastasize. We present a case of a middle aged woman with metastases to liver and omentum.

Introduction

Solid and cystic papillary tumors of the pancreas

mostly occur in young women. These tumors are usually well demarcated from the surrounding parenchyma. As a result, they can be easily removed by surgery completely which is associated with an excellent prognosis. However, some tumors can invade adjacent non-neoplastic pancreatic parenchyma or produce vascular or perineural invasion.¹ Also some of these tumors can recur locally,² sometimes as long as 3 to 10 years after original surgical resection and therefore long term follow up is important.³ Although very rare, there are several reports of these tumors metastasizing to liver and omentum.⁴⁻⁸ These studies indicate that although the large majority of papillary solid and cystic tumors of the pancreas behave in a benign fashion and have an excellent prognosis, they must

still be considered at least as "borderline" tumors¹ and carefully followed up.

We report a patient with a solid and cystic papillary tumor of the pancreas with metastases to omentum and liver.

Case Report

A 42 year old woman, the wife of a doctor in Islamabad, presented to the Department of General Surgery, PIMS, Islamabad in 2001 with a history of vague upper abdominal discomfort for 6 years. She had some episodes of dyspepsia but otherwise had no other signs and symptoms. Since one year, she also felt heaviness in right upper quadrant of abdomen. On physical examination, a mass could be palpated in the right upper quadrant. It was confirmed on ultrasound and CT scan but these studies as well as FNAC and per cutaneous needle biopsy failed to determine its exact site of origin or its nature. A hepatoma in the right lobe of the liver was suspected. Laparotomy was done in March 2001. A large retroperitoneal tumor was found in the sub hepatic area. It was in intimate contact with posterior aspect of duodenum and head of pancreas. It was grayish, friable in consistency, quite vascular and had a fairly well defined capsule. It was removed along with the capsule except in the vicinity of duodenum and pancreas from which it had to be peeled off. The tumor was 17x12x9cms in size. It was markedly hemorrhagic and necrotic. Histopathology at that time was suggestive of a low grade papillary neoplasm with a possible origin from the pancreas.

On repeat laparotomy she was found to have multiple small masses along the parietal peritoneum on the undersurface of right dome of diaphragm. The omentum was also riddled with tumorous nodules. The masses which were soft and capsulated were excised and omentectomy was also done. CT scan revealed hypodense area in the body of the pancreas (Figure 1) and two hypodense lesions in the right lobe of liver (Figure 2). We received the specimen in the section of histopathology AKU in the form of multiple nodules, the largest of which measured 8x8x5 cms. On sectioning, cut surface of these nodules was light brown with hemorrhagic and necrotic areas. A piece of omentum was also received which measured 19x9.5x5.5cms. On sectioning, it showed multiple small nodular lesions. Multiple representative sections were submitted.

Microscopic examination of these nodular lesions showed fibroadipose tissue infiltrated by a neoplastic lesion (Figure 3) composed of sheets of tumor cells which in areas formed a pseudo papillary pattern). The tumor cells were uniform, medium sized, polygonal in shape with ovoid nuclei and abundant pink to foamy cytoplasm. Mitoses were

scanty. Immuno histochemically, the tumor cells showed positivity for vimentin and Neuron Specific Enolase (NSE), and focal positivity for cytokeratin AE 1/AE3.

Based on the clinical history, operative findings, radiological findings and the histological and immuno-histochemical features, a diagnosis of pancreatic solid and cystic papillary neoplasm with metastases in liver omentum and peritoneal cavity was made. The patient's treatment by systemic chemotherapy is continuing.

Figure 1. Post contrast axial CT image showing a hypodense areas in the region of body of pancreas representing pancreatic mass.

Figure 2. Axial CT image showing two hypodense lesions involving the right lobe of liver representing metastasis

ease 4 to 11 years after surgery, 3 patients above 40 years of age were alive 1 to 5 years after surgery.

Sclafani et al³ have also reported patients with liver metastases who underwent resection of metastases or were treated with tamoxifen, who are alive on long term, follow up.

These studies indicate that papillary solid and cystic neoplasms of the pancreas in young patients have a good prognosis and are compatible with long term survival even in the presence of metastatic disease. However, prognosis is less favorable for older patients with metastases, and death from metastatic disease can occur several years after initial surgery. The ultimate outcome of our patient is yet to be seen as the patient's followup is still short and treatment by systemic chemotherapy is continuing.

Figure 3. Fibroadipose tissue infiltrated by sheets of tumor cells.

Discussion

Oertel and associates have stated in their study⁸ that "several patients over 20 years of age with this neoplasm have developed peritoneal and hepatic metastases and at least one has died". Matsunou et al⁵ studied 9 cases. All their patients were females. Seven patients were 8 to 24 years of age, while the remaining patients were 47 and 60 years of age. All the 7 younger patients were alive and well on long term follow up after enucleation or distal pancreatectomy. However, the two older patients developed liver and peritoneal metastases 4 to 6 years after initial surgery and died.

Horisawa et al⁶ in their series of 174 patients found 20 patients with metastases to liver, omentum, peritoneum, mesocolon, colon lymph nodes, lung and skin. The age of these patients with metastases ranged from 11 to 62 years. Long term follow up showed that all patients under 40 years of age were alive 1 to 16 years following surgery 5 patients of 40 years of age or above died of metastatic dis-

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