Solid and Papillary Epithelial Neoplasm of the Pancreas in a Nine Year-old Girl

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Abstract

Solid and papillary tumor of the pancreas is also called as FRANTZ's tumor. It is a very uncommon tumor with low malignancy potential. It occurs usually in young women of second and third decades of life. Only a few sporadic cases were reported under the age of ten. Here we present the clinico-pathological findings of this rare tumor in a nine year-old girl and its management.

Key words: Pancreas, low grade, solid, cystic, papillary

Introduction

A nine year-old girl presented with abdominal pain since forty days prior to admission. She also suffered from vague symptoms such as weakness, dyspepsia, fatigue, weight loss, and mild abdominal pain for the last two months. Physical examination revealed a palpable mass in the epigastrium. The mass was hard, non-mobile and non-tender. Abdominal CT (Fig.1) and ultrasonography revealed a round mass about six cm in diameter in the head of the pancreas. FNA of the tumor revealed a benign lesion. So the patient underwent pancreaticoduodenectomy. The patient was discharged in good condition after one week.

Grossly a large well defined soft dark brown solid and cystic tumor of about 6 cm was seen in the head of pancreas (Fig.2). The histologic examination revealed cystic spaces, solid sheet of tumor cells and papillary structures lined by small round cuboidal cells. There was no pleomorphism. No mitoses identified. Prominent nucleoli were not observed. Histologically the tumor was diagnosed as benign solid and cystic papillary epithelial neoplasm, (Fig.3a, Fig.3b). Many specimens were taken from the tumor to rule out the possibility of malignancy. All sections revealed the same histologic findings, described as above. The patient was followed carefully every three months. Till now, three years after the operation, the patient is doing very well without any evidence of recurrence or metastasis.

Discussion

The solid and cystic papillary tumor of the pancreas is a very rare and uncommon tumor. It is mainly seen in young women of second and third decades of life.1,2 Review of the literature revealed only few cases at the age of nine. It is considered as a low grade malignant tumor and is usually not present with metastasis. However a few cases with metastasis were reported.3 It has a good prognosis and is curable after the complete surgical resection.

The role of FNA in the diagnosis of this tumor was described in few
articles and considered as a valuable procedure for differentiating it from more aggressive malignant tumors. CT scan, ultrasonography and FNA studies are helpful for the preoperative evaluation and management of this rare tumor.

Clinically, this case was considered as a lymphoma, but other primary pancreatic tumors were also considered in the differential diagnoses such as pseudocyst and pancreaticoblastoma. So during the first laparotomy, lymph node biopsy and fine needle aspiration was performed, which were in favor of benign lesions (solid and cystic papillary neoplasm) and ruled out the possibility of lymphoma. So, in the second stage we planned for pancreaticoduodenectomy with pylorus preservation. Although the recent advanced studies such as CT scan, ultrasonography and FNA procedures were useful for the diagnosis of this case, we suspected the diagnosis because of its rarity in this age group. As a result of that the patient was operated for the second time.

In conclusion, solid and cystic, papillary neoplasms of the pancreas is extremely rare in this age group. It is a low-grade malignant tumor and completely curable with surgical excision. FNA studies is useful procedure for the pre-operative diagnosis of this rare tumor.

Figure 1: CT Scan Revealed a 6cm Encapsulated Mass in the Head of Pancreas

Figure 2: Excised Tumor is a Well Defined Round Capsulated Tumor

Figure 3a: Papillary Tumor Without Atypia, H&E:160

Figure 3b: In Other Parts of the Tumor There is Solid and Cystic Changes Without Atypia, H&E:280
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References