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Case Report

PANCREATIC ADENOCARCINOMA IN A YOUNG WOMAN

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Abstract

Despite significant improvement in the management strategy, pancreatic cancer continues to be a great challenge for surgeons and oncologists. Length of survival largely depends upon stage at diagnosis and a completeness of resection. Distal pancreatectomy with R0 resection has been reported as a favorable method in selected pancreatic body and tail tumors. Additional organ resections are rarely required. A young woman was diagnosed with a tumor in the body and tail of the pancreas that required splenectomy in addition to distal pancreatectomy. Postoperative course was uneventful. Histopathology revealed the tumor as moderately differentiated adenocarcinoma with extensive areas of necrosis, haemorrhage and cholesterol cleft formation. Resected end of pancreas was free of tumor. She has completed a course of chemotherapy and is doing well after 8 months of surgery.

Key Words: Adenocarcinoma, Pancreatic neoplasm, carcinoma of the body and tail of the pancreas.

Introduction

Pancreatic cancer is one of the deadliest cancers, being the 4th and 6th leading cause of cancer deaths in USA and UK respectively¹. It remains a disease of the elderly, with peak incidence between 65-75 years age group². As most patients present with advanced disease, resection rate is fairly low (2.6-9%)^{3,4,5,6} compared to other digestive tract cancers. The incidence of pancreatic cancer has been rising steadily over the past 60 years throughout the world. Unlike pancreatic head tumors, distal pancreatic tumors rarely cause jaundice. Therefore left sided pancreatic tumors are often not diagnosed until they attain a considerable size.

Correspondence to: Dr. Din Muhammad, FCPS (Surgery), MRCS, Assistant Professor, Department of Surgery, Shaheed Suhrawardy Medical College, e-mail: dr.din_zip@yahoo.com Received 15 February 2014 Accepted 03 March 2014 A young woman was diagnosed with distal pancreatic tumor which was found incidentally during an abdominal imaging examination. As the tumor was encroaching the splenic hilum, splenectomy along with distal pancreatectomy was done. Histopathology revealed adenocarcinoma. Patient has completed adjuvant chemotherapy and has passed 8 months since her surgery and is still doing well. She is presented as a rare incidence of pancreatic carcinoma in the younger age group.

Case Report

A 17-year-old married lady presented with recent onset of upper abdominal discomfort. There was no history of fever, vomiting, weight loss, abdominal pain, abdominal lump or back pain. Her bowel habit was normal. She had no significant past medical or surgical history. Personal and family history were unremarkable. Her general and abdominal examinations were normal. Routine investigations revealed normal blood count (Hb% - 12 g/dl, TC – 7000/cmm, ESR – 62 mm). Her random blood sugar

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was 6.3 mmol/L. Liver and renal function tests were within normal limit. Serum amylase and lipase were also normal. Ultrasonography of abdomen revealed a well defined oval mixed echogenic mass in left hypochondriac region measuring about 6.4x4.5 cm with necrosis which could not be separated from the tail of the pancreas (figure-1). Contrast CT scan showed a 7.9x5.8x8.0 cm mixed density mass lesion

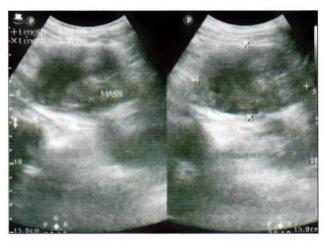


Fig.-1: Sonographic image showing a mixed echogenic mass (64×45 mm) in the left hypochondriac region suggestive of pancreatic mass.

in the left hypochondrium with mild heterogenous contrast enhancement (figure-2). The mass could not be separated from part of the body and tail of the pancreas. Serum CA 19-9 was below the reference value (< 2.5 U/mL). No preoperative tissue sampling was done. Patient was prepared for laparotomy, which was done with an upper transverse abdominal incision.

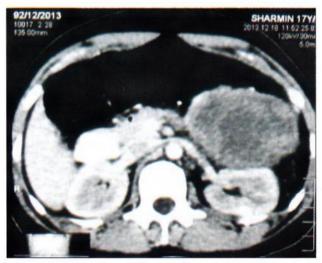


Fig.-2: Axial contrast CT showing mixed density mass lesion (79x58x80 mm) in the left hypochondriac region with mild heterogenous contrast enhancement.

A globular mass with mixed consistency (partly solid, partly cystic) was found at the tail of the pancreas almost encroaching the hilum of the spleen (firuge-3). It was free from the stomach, colon and the left kidney. There was no ascitis, peritoneal seedling or any other metastatic deposit. Distal pancreatectomy with splenectomy was done. Her postoperative recovery was uneventful. Histopathological examination of the resected specimen revealed a moderately differentiated adenocarcinoma of 8.5 cm diameter with extensive areas of necrosis, haemorrhage and cholesterol cleft formation (figure-6). Resected end of pancreas was free of tumor. No lymph node metastasis was found. Pathological stage was T2N0M0 (Stage IB). She was referred to oncologist and has already completed adjuvant chemotherapy schedule. She is at the end of 8th month after the surgery with regular follow up and is doing well.

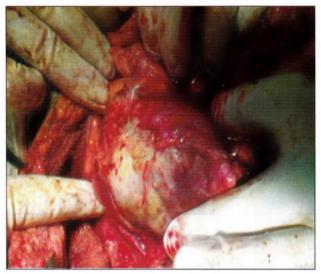


Fig.-3: Per-operative view showing the tumor arising from the tail of the pancreas close to the splenic hilum.

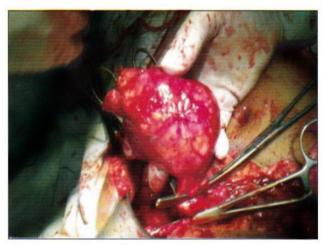


Fig.-4: The tumor is about to be resected



Fig.-5: Resected distal pancreatectomy specimen along with the spleen.

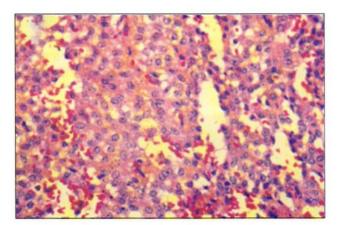


Fig.-6: Microscopic view of pancreatic adenocarcinoma.

pancreatic cancer (2002)17.			
Stage	Т	Ν	М
0	Tis	0	0
IA '	T1	0	0
IB	T2	0	0
IIA	Т3	0	0
IIB	T1-3	N1	0
Ш	T4	Any N	0
IV	Any T	Any N	M1

 Table I

 Union International Contre la Cancer staging of pancreatic cancer (2002)¹⁷.

Tis, carcinoma in situ; T1, tumour limited to the pancreas <2 cm;T2, tumour limited to the pancreas >2 cm; T3, tumour extending beyond the pancreas but without involvement of the coeliac axis or the superior mesenteric artery; T4, tumour involving coeliac axis or superior mesenteric artery; N0, no regional lymph node metastasis; N1, regional lymph node metastasis; M1, distant metastasis.

Discussion

Pancreatic adenocarcinoma is one of the most aggressive human malignancies. Owing to late presentation, this disease has very low resection rates. Overall median survival is less than 6 months and 5-year survival rate is $0.4-5\%^{4.5}$. Those who do undergo resection have a median survival of 11–20 months and a 5-year survival rate of 7–25 per cent, but virtually all patients are dead within 7 years of surgery⁷.

The incidence of pancreatic cancer varies with age, sex and race. In most countries the incidence is higher in men than in women by a ratio of about 2:17. The disease is rare in those under 25 years of age and is uncommon under the age of 45 years⁸. It is more common on Western or industrialized countries. There is strong evidence implicating smoking and dietary factors in the etiology of pancreatic cancers. Familial background is also an important risk factor which is found in approximately 5-10% of those affected. Chronic pancreatitis is also an important aetiological factor in pancreatic cancer, with an increased risk of 15-25-fold in sporadic chronic pancreatitis, increasing to 70–100-fold in hereditary pancreatitis. In our case, the patient was only 17, well below the peak age group of 65-75 years. There was no identifiable predisposing factor.

Pancreatic ductal adenocarcinoma is the most common epithelial exocrine pancreatic tumour and accounts for more than 85 per cent of all malignant pancreatic tumours. In series of surgical resections, 80–90% of tumours are located in the head of the gland⁹. Lymph node metastases are present in 20–77% of resected specimens of patients with head disease^{10,11,12,13,14,15,16}. Perineural (70–77%), vascular (45%) and lymphatic (80%) invasion is also frequently seen in resection specimens^{13,16}.

The 2002 Union International contre la Cancer (UICC) tumour node metastasis (TNM) classification¹⁷ is shown in Table 1. Staging classifications that identify 'resectable' (stage I or II), 'locally advanced' (stage III) or 'metastatic' (stage IV) pancreatic cancer have become increasingly important for stratifying patients in prospective clinical trials and in the development of treatments for particular stages of the disease.

In most patients the clinical diagnosis is fairly straightforward. Patients with tumors in the pancreatic head region present with painless progressive obstructive jaundice while weight loss with intractable back pain may be the only feature in case of tumors of body and tail of the pancreas. There are associated conditions, such as late-onset diabetes mellitus or an unexplained attack of acute pancreatitis, which may point to an underlying cancer. Clinical features, such as persistent back pain, marked and rapid weight loss, abdominal mass, ascites and supraclavicular lymphadenopathy, usually indicate an irresectable tumour.

Initial diagnostic workup includes abdominal ultrasonography which might reveal the tumor mass, dilated extrahepatic biliary tree and/or main pancreatic duct (MPD), ascitis, lymphadenopathy or liver secondaries. In this patient, there were no other abnormalities other than the pancreatic mass itself. Contrast-enhanced computed tomography (CT), using multislice scanners with arterial and portal venous phases of contrast enhancement, is the 'gold standard' method for clinical staging; it accurately predicts resectability in 80-90% of patients^{18,19,20}. Factors contraindicating resection include liver, peritoneal or other metastases, distant lymph node metastases, major venous encasement (over 2 cm in length, more than 50% circumferential involvement), superior mesenteric, coeliac or hepatic artery encasement, as well as major co-morbidities. Factors that do not contraindicate resection include contiguous invasion of the duodenum, stomach or colon, lymph node metastasis within the operative field, venous impingement or minimal invasion of the superior mesenteric vein-splenic vein-hepatic portal vein trifurcation, gastroduodenal artery encasement, and the age of the patient. Endoluminal ultrasonography is highly sensitive in the detection of small tumours and invasion of major vascular structures²¹. Laparoscopy, including laparoscopic ultrasonography, can detect occult metastatic lesions in the liver and peritoneal cavity in 10-35 percent of instances not identified by other imaging modalities^{22,23,24}. Positron emission tomography is an evolving technique that measures the metabolism in tumor cells; at the present time there is overlap with chronic pancreatitis but it does seem to be able to detect metastases not visible by other imaging modalities²⁵. Preoperative tissue diagnosis may be obtained by brushings at ERCP or by endoluminal ultrasonography guided fine needle aspiration for cytology. Linder S et al found in their series of 270 patients with pancreatic carcinoma that the sensitivity, specificity and accuracy of percutaneous fine-needle biopsy were 69, 100 and 75 per cent respectively²⁶

Pancreatoduodenectomy, either standard (Kausch-Whipple procedure) or its pylorus preserving variant, is the surgical option for resectable tumors of the pancreatic head. A left resection (distal pancreatectomy) is indicated for lesions in the body and tail of the pancreas, but pancreatic ductal carcinoma is seldom resectable in this location. But resectable tumours in the body and tail of the pancreas often carry a favourable prognosis (such as mucinous cystic tumours)⁷. Studies of the effects of adjuvant therapy following resection have produced mixed results and debate on this issue continues. The most recent and largest trial till now is the European Study Group for Pancreatic Cancer (ESPAC-1) trial. It concluded that although adjuvant chemoradiotherapy showed no survival benefit, there was evidence of improved survival with adjuvant chemotherapy⁷.

Conclusion

Pancreatic cancer is not so uncommon in surgical practice in our country. But adenocarcinoma in a 17 year old lady is rare. Despite having a poor prognosis, there has been a substantial sharpening of the standard approaches to the treatment of pancreatic cancer. Currently, resection provides the only meaningful chance of cure or at least increased survival. But other adjuvant modalities including gene therapy are being explored and improved. Till date the standard of care is now clearly defined as resection followed by adjuvant chemotherapy. This patient has been managed likewise and is doing well so far. She is kept under lifelong regular follow up.

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