



Pancreatic Head Adenocarcinoma in Preadolescence

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Abstract

Pancreatic cancers usually presents in older age group, with a median age of 66 years. Periampullary tumors may arise from ampulla, distal common bile duct, pancreas or duodenum. We present here a case of 12-year old girl with pancreatic head carcinoma.

Keywords: Pancreaticoduodenectomy; Pancreatic cancer; Whipple; Periampullary carcinoma

Introduction

Periampullary carcinomas are tumors arising within 2 cm diameter of ampulla of Vater and it comprises of carcinomas arising from ampulla, distal common bile duct, pancreas or duodenum. They share common presenting symptoms and therapeutic approaches. Here we present a case report of pancreatic head carcinoma presenting in a pre-adolescent girl and its management.

Case Presentation

A 12-year-old girl presented with complains of yellowish discoloration of eyes and skin for last 2 months. She gave history of on and off pain abdomen associated with fever, along with significant weight loss. There was no history of clay colored stools. On clinical examination, she had pallor and icterus. Per abdomen examination was within normal limits except for slight pain on deep palpation in epigastrium. MRCP done elsewhere showed significantly dilated bi-lobar intra-hepatic biliary radicals, Common Hepatic Duct (CHD) and Common Bile Duct (CBD) with abrupt narrowing of distal CBD-likely CBD stricture. Multiple discrete and confluency nodular lesions involving both hepatic lobes were present. Her hemoglobin was 8.4 g/dl and total bilirubin 5.8 g/dl at presentation. Other liver function tests were normal with slight raise of SGOT and SGPT.

An ERCP was done which revealed ulceroproliferative growth at papilla and took multiple biopsies which revealed moderately differentiated adenocarcinoma. Meanwhile, CECT abdomen done at our center revealed a periampullary mass involving 2nd and 3rd parts of duodenum, head of pancreas and distal CBD, with few ill-defined lesions in right lobe liver that could be metastases.

Hospital tumor board consultation was sought in view of suspected systemic metastasis; it was decided to administer neo-adjuvant chemotherapy after decompression of biliary system. The attempts at cannulation of CBD failed in view of bulky growth and distortion of anatomy of papilla. A Percutaneous Transhepatic Biliary Drainage (PTBD) was successful with 8 Fr plastic stent, distal end across ampulla. It helped in reduction of serum bilirubin from 11 g (raised meanwhile) to 2.9 g and neo adjuvant chemotherapy was started, in form of gemcitabine. She received two cycles of neo-adjuvant chemotherapy and then radiological assessment was done using CT scan, which showed resolution of liver lesions but static periampullary tumor without any large vessel involvement or encasement.

She underwent Pylorus-Preserving Pancreaticoduodenectomy (PPPD) with gastrojejunostomy, choledochojejunostomy and pancreaticojejunostomy. The presence of three polyps in duodenum and jejunum was noted at surgery. Patient shifted to ICU in post-operative period and received packed red cell volume and fresh frozen plasma transfusions. She developed ventilator associated pneumonia and sepsis in post-operative period and succumbed on 5th postoperative day. Histopathologic examination revealed it to be pancreatic head adenocarcinoma with R0 margins and polyps were reported as tubular adenoma with no dysplastic changes. One lymph node was positive.

Discussion

Periampullary carcinomas are increasingly recognized condition in view of increasing

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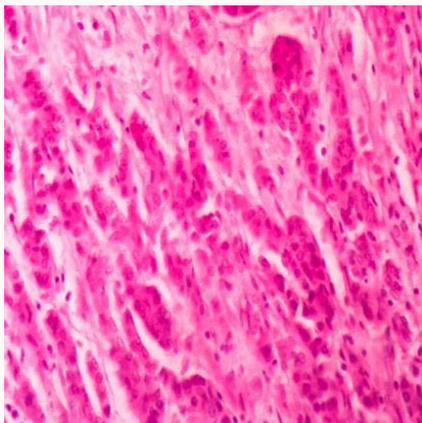
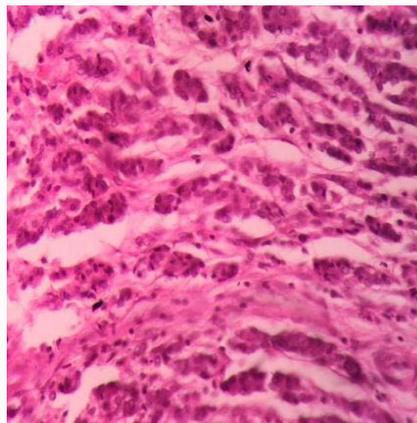


Figure 1: HPE showing duodenal mucosa along with tumor present in small nests and sheets.



Figures 3: The nests and clusters of tumor cells with high NC ratio, scant cytoplasm and prominent nucleoli.

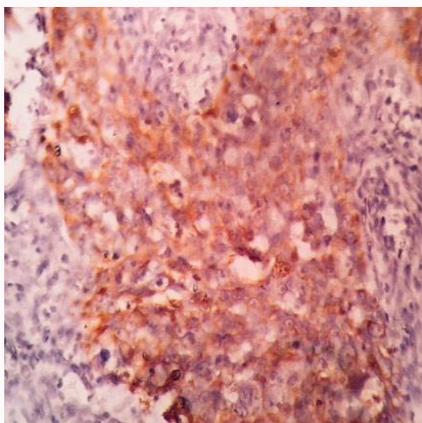


Figure 2: The nests and clusters of tumor cells with high NC ratio, scant cytoplasm and prominent nucleoli.

awareness of symptoms, education level of patients and availability of endoscopy and other radiological tools. It is a common name given to tumors arising within 2 cm of ampulla of Vater. Pancreatic cancers usually present in older age group, with a median age of 66 years [1]. Periapillary carcinomas usually present with mass effects, depending on organ of origin or secondary to metastatic complications. There may be mechanical duodenal obstruction causing gastric outlet blockage or compressing ampulla of Vater causing jaundice, yellowish discoloration of body or hemorrhage secondary to venous obstruction. Metastasis to liver or peritoneum may cause ascites and liver cell failure. This child presented with icterus and maintained liver function with increased Serum Bilirubin (SB).

MRCP and sectional MR imaging are quoted to be useful in determining the origins of periampullary carcinomas [2]. There are various signs which help in differentiating organ of origin, like pancreatic cancer usually demonstrate dilatation of side branches of pancreatic duct and enhance poorly on gadolinium enhanced images and ampullary carcinomas manifests as a small mass, periductal thickening, or bulging of the duodenal papilla. These findings could not be appreciated in our patient in view of poor quality films and non-availability of 3D reconstructed images. CECT and CT angiography with 3D reconstruction has been also quoted in literature to guide resectability of cancer [3]. Grade 4 lesions (total encasement of either

the superior mesenteric vein or artery) were considered unresectable. Resectability rates decrease as the grade of lesion increases. Our patient had grade 2 lesion on CT scan, with resectability rate of 50% as quoted in literature [3].

Endoscopy surveillance and ablative therapy has role in management of periampullary lesions especially in adenomas [4], although eradication of ampullary lesion may need multiple settings. Literature also quotes adenoma-carcinoma sequence in duodenum in familial adenomatous polyposis syndrome in adults [5], though no recognized genetic syndromes associated with pancreatic carcinoma in children or adolescents [6-9]. Although our patient had three polyps in duodenum and jejunum, which histologically came out to be tubular adenoma with no dysplastic changes, so any genetic correlation could not be sought.

Preoperative biliary drainage was introduced to decrease bilirubin load, help in symptomatic palliation in metastatic disease or render patient fit for chemotherapy and surgery. But two meta-analyses showed that patients receiving pre-operative biliary drainage have more post-operative complications [10,11]. Success of biliary drainage is documented to be decrease of >50% of SB within 2 weeks and proceeding to surgery in next 4 to 6 weeks [12], which could not be achieved in our patient in view of twice shunt blockage.

Preoperative platelet counts are also identified in literature as prognostic factor in periampullary carcinoma. Low platelet counts associated with inferior outcome results, mechanisms unclear, may be general factors or platelet specific factors [13].

The agents of metastatic pancreatic cancer include 5-FU, streptozotocin, mitomycin C, gemcitabine and doxorubicin, associated with response rates of 7% to 36% [14-16]. Our patient received 2 cycles of gemcitabine chemotherapy. Response may be quoted as we could do R0 resection and did not find any encasement of great vessels [CT showed grade 2 lesions].

Our patient underwent pylorus preserving pancreaticoduodenectomy as there was no gross infiltration of D1 or distal stomach. Literature states that pylorus preservation does neither compromise long-term survival, nor increase any operative risks [17].

In one of the largest single-institution experience with Pancreaticoduodenectomy [PD] for pancreatic cancer, median age

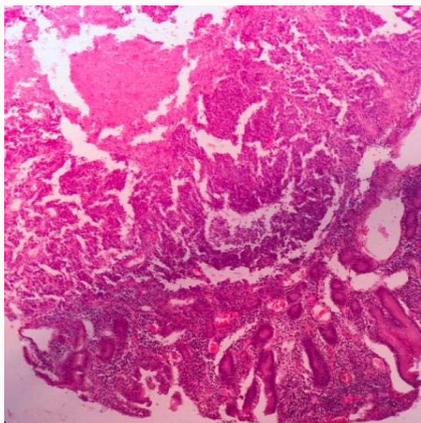


Figure 4: Tumor cells showing expression of cytokeratin on IHC.

was 66 years, median tumor diameter was 3 cm, 42% of PD specimens had positive margins, and 78% had positive Lymph Nodes (LN), with median survival of 18 months. They quoted peri operative morbidity of 38% and decrease in mortality rates of 38% in 1970s to 1% in 2000s [1]. Factors having impact on survival were tumor diameter, LN status, margins and histology grade. Pancreatic head adenocarcinoma has poor prognosis as compared to non-pancreatic peri ampullary carcinomas.

In a large UK experience of adenocarcinoma of the head of the pancreas and periampullary region, age <60 years, tumor of head of pancreas, positive LNs, R1 resection, poor differentiating tumor, portal vein invasion were studied to be independent factors for decreased survival. Mortality in study was 4.8%, with median survival of 13.4 months in pancreatic head, 35.5 months in ampullary and 16 months in distal bile duct cancers [18]. Our patient had three independent negative factors as per this study.

Simultaneous resection of peri-ampullary or pancreatic cancer, with synchronous liver metastasis has been attempted but it has proven to be associated with increase morbidity and mortality and does not increase long term survival [19]. Our patient did not have any gross lesions on liver surface and no liver procedure was done for suspected metastasis lesion as per CT scan.

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