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Rare periampullary carcinoma: A case study

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Abstract
Periampullary carcinoma is usually used to define a heterogeneous group of neoplasms raised on the head of pancreas, duodenum and distal common bile duct. Most of the periampullary growths are adenocarcinomas. Timely diagnosis and successful surgical treatment is totally dependent on first physician. A 60 years old male patient was presented to medical outpatients of Bahawal victoria Hospital Bahawalpur in October 2019 with rare etiology of unexpected vomiting, nausea, fatigue, weight loss and abdominal cramps from 12 days continuously. Abdominal ultrasound revealed hypo-echoic mass with measurement of 2.6x2.7cm on the head of pancrease causing obstruction of distal common bile duct with mild intrahepatic cholestasis. On the basis of physical examination sign & symptoms and lab investigations patient was considered of having periampullary cancer and prompt wipple plan was prepared. Vigilance of physician and support of patient's family helped to make an early decision of pan creato duo denecto my of the patient before the multi nodulation of tumor hence, increased the life expectancy.

Keywords: periampullary, wipple plan, pancreatoduodenectomy, pancreatic cancer

Introduction

Periampullary carcinoma is usually used to define a heterogeneous group of neoplasms raised on the head of pancreas, duodenum and distal common bile duct. This is a different from ampullary carcinoma as topography of the tumor is centered in this position and anatomically twisted among three organs (Braasch, *et al*, 1975) [1]. Holzheimer *et al*, in 2001 [6] reported that periampullay carcinoma is responsible for >30000 annual deaths in United States. It is different from ampullary carcinoma on basis of its origin and transformation in prognosis and respectability (Holzheimer, *et al*, 2001) [6]. Incidence of carcinomas involving pancreas has been reported to be 12.4/100000 annually and only around 1.5% are diagnosed during their life time (Ries, *et al*, 2003) [7]. Although the incidence of these type of cancers is low however it is associated with low survival rates and ranked fourth or fifth most common cause of cancer mortality. Incidence of this type of cancer is not known in Pakistan though in India it is reported to be 0.2-1.8/100000 among women and higher rate of 0.5-2.4/100000 of men (Dhir, *et al*, 1999) [7]. Calhoun et.al, in 2008 reported decreasing trends of mortality and incidence of cancers related to pancreas during 1992 to 200 and showed static mortality due to pancreatic cancer among standardized age groups (Chalhoun, *et al*, 2008). Most of the periampullary growths are adenocarcinomas as reported in Lahey clinic that 95.2% cases among 348 patients underwent pan creato duodenecto my were diagnosed as adenocarcinoma in last 30 years. Timely diagnosis and successful surgical treatment is totally dependent on first physician, who is suspicious of periampullary carcinoma (Braasch, *et al*, 1975) [1]. Aim of present case report is to show the vigilance of physician in timely diagnosis of periampullary cancer patient presented with complaint of vomiting from number of days with nausea, fatigue and weight loss.

Case Report

Patient Profile

Presenting Complaints

A 60 years old male patient was presented to medical outpatients of Bahawal victoria Hospital Bahawalpur in October 2019 with rare etiology of unexpected vomiting, nausea, fatigue, weight loss and abdominal cramps from 12 days continuously.

Past Medical History

A case of an elderly lady diagnosed with metastatic periampullary cancer who received suboptimal chemotherapy and managed to survive for 17 months after diagnosis. The case was chosen due to its unusual course and unexpectedly prolonged survival with minimal chemotherapy in metastatic setting

Family History

The women belong to a lower socio economic class had 7 children three daughters and four sons.

Physical Examination

Vital Signs: Temperature, 98.6F, heart rate, 88/min; respiratory rate, 22/min; pulse, 104/min,

General Appearance: The patient was appearing drowsy, dehydrated, and appeared malnourished lying in his mother lap

having height 147cm and weight 50kg

Respiratory System: He has bilateral crepitations that were present more on right side of the chest.

Cardiovascular System: He had a regular rate and rhythm with no murmurs, rubs, or gallops.

Reflexes and Power: Reflexes and power is good

Laboratory Evaluation: Patient was advised laboratory testing for complete blood counts (CBC), renal function test, liver function test, serum electrolytes, random blood sugar, Hepatitis B Virus Antigen, Hepatitis C Virus Antibody, abdominal ultrasound, echo cardiogram, Echo cardiograph, Chest X-ray P A view, prothrombin time and activated partial thromboplastin time. Abdominal ultrasound revealed hypo-echoic mass with measurement of 2.6x2.7cm on the head of pancrease causing obstruction of distal common bile duct with mild intrahepatic cholestasis. Further patient was diabetic and on oral hypoglycemic from one year or so. Liver function tests were raised with high bilirubin, transaminases and alkaline phosphatase.

Differential Diagnosis: Endoscopic images of periampullary tumor prior and after endoscopic mucosal resection. A: A 1 cm × 1.5 cm isoechoic, submucosal nodule near the major ampulla (arrows); B: Periampullary submucosal nodule with a normal overlying mucosa; C: Lesion post endoscopic mucosal resection.

Confirmatory Evaluation

Histologic characteristics of the gangliocytic paragangliomas. A mucosal location of the periampullary tumor (H and E, original magnification × 2); Epitheliod cells (black arrow) with surrounding spindle cells (white arrow). Ganglion-like cells present (arrow heads) (H and E, original magnification × 20); B: Immunohistochemistry of tumor showing positivity for cytokeratin in the epitheliod cells (original magnification × 4); C: Immunohistochemistry of tumor showing S-100 positivity of the spindle cell component (original magnification × 10).

Principal Diagnosis

- Gangliocytic paragangliomas.
- Epitheliod cells
- B: Immunohistochemistry of tumor showing positivity for cytokeratin in the epitheliod cells

Based on clinical presentation, patient family history and diagnosis of Rare Periampullary Carcinoma

Pathophysiology

The periampullary region is anatomically complex, representing the junction of 3 different epithelia, pancreatic ducts, bile ducts, and duodenal mucosa. Grossly, carcinomas originating in the ampulla of Vater can arise from 1 of 4 epithelial types:

- Terminal common bile duct
- Duodenal mucosa
- Pancreatic duct
- Ampulla of Vater

Distinguishing between true ampullary cancers and periampullary tumors is critical to understanding the biology of these lesions. Each type of mucosa produces a different pattern of mucus secretion. In a complete histochemical study, Dawson and Connolly divided acid mucins into sulphomucins and sialomucins; in general, ampullary cancers produce sialomucins, whereas periampullary tumors secrete sulfated mucins. These researchers demonstrated that ampullary tumors secreting sialomucins had a better prognosis (100% vs 27% 5-y survival rate). [2] Other investigators have confirmed the prognostic power of the pattern of mucin secretion.

Carter *et al* suggest that, histologically, ampullary tumors can be classified as either pancreaticobiliary or intestinal, and that the clinical behavior of these tumors reflects this classification; the course of intestinal ampullary adenocarcinomas is similar to that of their duodenal counterparts, whereas pancreaticobiliary tumors follow a more aggressive course, similar to that of pancreatic adenocarcinomas.

However, the former classification has been challenged by lines of evidence showing a significant interobserver variability upon the interpretation of these patterns, with a mixed subtype being the predominant subgroup, representing up to 40% of cases. In addition, poorly differentiated tumors can further confound the histological classification. The prognostic significance of this histological classification has been subjected to investigation, with inconsistent results.

Immunohistochemical stains for expressions of carcinoembryonic antigen (CEA), carbohydrate antigen (CA) 19-9, Ki-67, and p53 have been studied for prognostic power. In a series of 45 patients, expression of CA 19-9 labeling intensity and apical localization both were statistically significant predictors of poor prognosis. The 5-year survival rates were markedly different between tumors that expressed CA 19-9 and those that did not (36% vs 100%). [6] CEA expression also might be a marker for prognosis, but it is much weaker. Ki-67 and p53 were not demonstrated to have an effect on outcome. Research along these avenues ultimately might provide the rationale for discriminative administration of adjuvant therapy.

Management

Discussion

Prompt diagnosis and early management of the patients with periampullary carcinoma and other cancers related to pancreas is the only key of success for batter recovery and life expectancy of patients (Braasch, *et al*, 1975) [1]. Since the etiology is very rare and one may take much time sometimes to reach final conclusion results in delayed diagnosis and treatment as well. Urgent decisions of surgery some times are not acceptable for patients or their attendants due to number of reasons in present settings but their cooperation is of great importance at this stage.

National Institute of Health, United States, has National Cancer Institute which generate data on surveillance, epidemiology and end result program (SEEP) among cancer patients which estimated 56770 new cancer cases till date in 2019, of which pancreas related cancers were 3.2%, similarly total deaths due to cancers remained 45750 till date in 2019, of which pancreatic cancer related deaths are 7.5% clearly showing more than double of the proportion of occurrence of cancer. Further the chances of survival for 5 years, once diagnosed with pancreatic related

cancers is also reported to be only 9.3% (SEEP, 2019). We lack such information in Pakistan as no public surveillance and cancer registry is present at public as well as private hospitals.

Presently classical palpable gallbladder was observed by the physician as study reported that the classic palpable gallbladder in the presence of painless jaundice has been seen in about one-fourth of patients with resectable disease. The gallbladder may not be palpable because of hepatomegaly or because of scarring from previous inflammation. Therefore periampullary cancer may not be omitted if gallbladder is not palpable (Braasch, *et al*, 1975) [1]. Brown et.al, concluded that around 80% of the patients who undergo pancreaticoduodenectomy among node negative periampullary cancer further reported that long term survival is expected once the patients survives for 3 years after surgery (Brown, *et al*, 2005) [2] completely support the decisions taken in present study.

Present study is also in agreement with a recent study which determined by He, et.al, also supported timely prediction and in time pancreatoduodenectomy of periampullary carcinomas (He, *et al*, 2018). On the other hand prognosis has been reported to be poor usually and multi-model approaches comprising endoscopy, surgery and oncology are recommended (Sarocchi *et al*, 2018) [8] are in concordance to the present study.

Conclusion

Literature support the timely prediction and prompt surgery of periampullary cancer on the basis of available physical, clinical, biochemical and imaging evidences as is done in this case. Vigilance of physician and support of patient's family helped to make an early decision of pancreatoduodenectomy of the patient before the multi nodulation of tumor hence, increased the life expectancy.

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