Case Report

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An unusual presentation of very early onset metastatic pancreatic adenocarcinoma in a young man: case report

Abhishek Arora*, Ashesh K. Jha, Manoj Kumar, Manoj Kumar

Department of General Surgery, All India Institute of Medical Sciences, Patna, Bihar, India

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*Correspondence: Dr. Abhishek Arora,

E-mail: aroraab95@gmail.com

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ABSTRACT

Pancreatic ductal adenocarcinoma (PDAC) is rare below 45 years of age and usually presents with constitutional symptoms, pain and jaundice. A 25 year old man, non-smoker, non-alcoholic presented to us with a lump in central upper abdomen of 1 month duration. Abdominal examination revealed a hard epigastric mass of size 7×10 cm. There were no other associated symptoms and no history of any similar illnesses in the family members. All laboratory parameters were within normal limits. Computed tomography showed a heterogeneous mass of size $7 \times 13 \times 15$ cm arising from the head and neck of pancreas abutting the anterior abdominal wall with multiple abdominal and pelvic peritoneal deposits. Ultrasound guided core needle biopsy suggested PDAC. Hence, a diagnosis of very early onset metastatic PDAC was rendered and the patient was referred for palliative chemotherapy. PDAC below 45 years of age is known as very early onset PDAC. It is usually associated with family history of PDAC and known risk factors. This case was unique because of absence of the characteristic clinical features and known risk factors, early onset of occurrence and negative family history, which led to a diagnostic dilemma.

Keywords: Pancreatic cancer, Very early onset, Metastatic pancreatic cancer, Atypical presentation, Case report

INTRODUCTION

Pancreatic cancer is the 11th most common cause of cancer in the world and the 7th most common cause of death in industrialized countries, as per GLOBOCAN 2018 estimates. Various risk factors that have been identified for pancreatic cancer including smoking, diabetes mellitus, obesity, dietary factors, alcohol abuse, age, ethnicity, family history, genetic factors and chronic pancreatitis. It is usually considered as a disease of the elderly population with most cases reported in people over 70 years of age. However, in about 20% of the cases, its occurrence is reported below 60 years of age (early onset) and in approximately 3% below 45 years of age (very early onset). There are no cases of very early onset pancreatic cancer reported from India as reviewed

in literature and few cases worldwide. Also, these young individuals customarily have either known risk factors or suffer from the familial or genetic syndromes, known to cause pancreatic cancer. We herein described a unique case of metastatic PDAC arising from the head and body of the pancreas in a 25 year gentleman who presented to us only with an abdominal lump.

CASE REPORT

A 25 year old gentleman presented to our outpatient department (OPD) with an abdominal lump of 1 month duration. Apart from abdominal lump, he was completely asymptomatic. He denied any history of smoking, alcohol intake, abdominal pain and any known malignancies in family.

The general physical examination was unremarkable. A fullness in upper abdomen was evident on inspection, upon palpation a lump of size 7×10 cm occupying the upper and central abdomen noticed. It was non-tender, firm in consistency and irregular in shape. Rest of the systemic examination was unremarkable. laboratory tests including liver function test as well as serum tumor markers were within normal limits. Abdominal computed tomography with contrast enhancement revealed a heterogeneous mass lesion of size 7×13×15 cm arising from the pancreatic head and neck abutting the anterior abdominal wall along with multiple abdominal and pelvic peritoneal deposits (Figure 1 and 2).

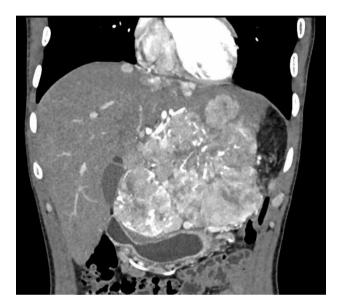


Figure 1: CECT image showing heterogeneously enhancing mass arising from head and neck of pancreas.

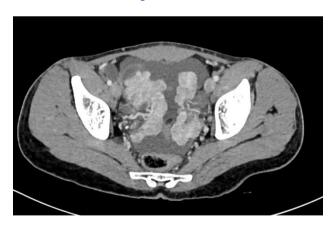


Figure 2: CECT image showing pelvic peritoneal metastasis.

As this lesion was reaching up to the anterior abdominal wall therefore, ultrasound guided percutaneous core needle biopsy was done. Final histopathological report was suggestive of moderately differentiated pancreatic adenocarcinoma. Hence, a diagnosis of very early onset

metastatic PDAC was rendered and the patient was referred for palliative chemotherapy.

DISCUSSION

Pancreatic ductal cancer is rare in young patients. PDAC before 45 years of age is known as very early onset pancreatic cancer (VEOPC).³ The age-standardized incidence rates of pancreatic cancer vary considerably in different parts of the world, from as low as 0.6/100000 persons per year in regions of Asia to as high as 12.6/100000 in the West.⁴ In India, occurrence of malignant lesions of pancreas are less in comparison to western world. In this part of the world, the frequency of pancreatic malignancy is 0.5-2.4/100,000 persons annually in women and 0.2-1.8/100,000 persons annually in men.5 Nonetheless, survivorship in individuals with pancreatic cancer is usually less with 5 year relative survival rate for all stages being only 10%.6 The reason for persistent inferior end result of this kind may be linked to the fact that this condition generally remains asymptomatic in the early phases and usually when symptoms do appear, the malignancy is either locally advanced or metastatic. Thereby, customarily only about 11% of pancreatic cancer are resectable at presentation.⁶

According to the PanC4 study, in which eight case-control studies relating to early onset (EOPC) and very early onset (VEOPC) were reviewed and analyzed, it was found that the known risk factors for pancreatic cancer like smoking, diabetes, family history of pancreatic cancer and obesity also apply to EOPC, while VEOPC has a different risk profile with heavy alcohol consumption as the single most important risk factor while smoking, diabetes and obesity has a lesser impact on risk of pancreatic cancer.³

Pancreatic cancer is generally silent in the early stages. The symptoms are often vague and unremarkable, which causes an unnecessary delay in presentation and diagnosis. This delay can have devastating effects on the life of the patient who may no longer be able to undergo a potentially lifesaving surgery. This is especially pertinent to VEOPC where years of potential life lost (YPLL) have a severe impact on the burden of disease.

Complete resection through surgery remains the only potentially curative treatment of PDAC. For unresectable cases, as per the ESMO guidelines, palliative therapies depend on the performance status, Eastern cooperative oncology group (ECOG) score of patients. In cases with good ECOG status of 0 or 1, combination chemotherapy with folfirinox or gemcitabine and nab-paclitaxel is recommended, while patients with ECOG score of 2 should receive gemcitabine with or without nab-paclitaxel. All others (ECOG 3 or 4, presence of significant co-morbidities and limited life expectancy) should receive symptomatic treatment only. Same principles may apply to EOPC and VEOPC patients, but

unfortunately no specific studies regarding the same are available in literature.

Apart from the well-known aggressive nature of this fearsome malignancy, this case also intended to highlight the occurrence of very early onset pancreatic cancer in this part of the world. As this gentleman denied any kind of substance abuse and there was no family history of malignancy in the first or second degree relatives and on top of that incidence rates of pancreatic neoplasms were significantly lower in this region. Furthermore, relatively silent nature of this malignancy in the early stages made things difficult for the treating physicians to identify it at the earlier stage. However, what made this case unique was that, in spite of reaching an enormous size and involving almost whole of the pancreas with distant metastasis, there were no systemic manifestations of this disease, in defiance of, classical clinical presentation of pancreatic adenocarcinoma involving the head of the with progressive jaundice and other manifestations, our patient presented to us only with a sizable abdominal lump.

CONCLUSION

Rarity along with poor prognosis associated with very early onset pancreatic cancer requires a high degree of suspicion while dealing with upper abdominal lumps. This case also highlights the fact that this dreaded disease can involve young individual without any risk factors or familial syndrome.

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