

## CASE REPORT

# A Rare Occurrence of Small Cell Carcinoma in Pancreas: A Case Report

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### ABSTRACT

**Introduction** Small cell carcinoma is an aggressive tumor with poor prognosis. Lung is the predominant site in 18-20% of the patients. Overall, 2.5% of all small cell carcinoma arises in the extra-pulmonary site. Pancreatic small cell carcinoma has aggressive nature thus patients present late with advanced disease or with metastases. Survival of patient is between 2 to 9 months. There is neither any staging system nor any accepted guideline regarding the management of small cell carcinoma of pancreas. We present a rare case of pancreatic small cell carcinoma. This will enlighten us about clinic-pathological factors and prognosis of disease. **Case discussion** A 53 year old female with no past history of medical illness or addiction, married with 6 kids, presented with history of jaundice along with weight loss for 1 month. Computed Tomography Scan showed growth in peri-ampullary region. Endoscopic retrograde cholangio-pancreaticography and stenting was performed. Specimen was taken from peri-ampullary growth which showed small cell carcinoma of pancreas. **Conclusion** Small cell carcinoma is an uncommon neoplasm with only a few cases reported in literature. Rarity of disease makes it difficult to draw conclusion from this case report. However, it will be an addition in the literature for better understanding of the disease. Prospectively, meta-analyses and systemic reviews are required to understand it.

### INTRODUCTION

Small cell carcinoma is an aggressive tumor with poor prognosis. Lung is the predominant site in 18-20% of the patients [1]. Overall, 2.5% of all small cell carcinoma arises in the extra-pulmonary site [2, 3, 4]. Small cell carcinoma also affects other organs of the body [5, 6, 7, 8, 9, 10] including cervix, esophagus, stomach, urinary bladder, skin, rectum, gall bladder and pancreas [11].

About 1% of the pancreatic cancers are small cell carcinoma [12]. Most prevalent site for small cell carcinoma of pancreas is pancreatic head and it affects older age group [12, 13, 14]. Due to the rare occurrence of disease, till date fewer than 100 cases have been reported.

Round to spindle small cells with dense nuclei and sparse cytoplasm under light microscope are seen in small cell carcinoma [15]. Pancreatic small cell carcinoma

has aggressive nature thus patients present late with advanced disease or with metastases. Survival of patient is between 2 to 9 months [2, 16]. There is neither any staging system nor any accepted guideline regarding the management of small cell carcinoma of pancreas. Cisplatin and etoposide based regimen provide the response rate up to 70% [17].

We present a rare case of pancreatic small cell carcinoma. This will enlighten us about clinic-pathological factors and prognosis of disease.

### CASE DISCUSSION

A 53 year old female with no past history of medical illness or addiction, married with 6 kids, presented with history of jaundice along with weight loss for 1 month for which she consulted gastroenterologist. CT scan showed growth in peri-ampullary region involving superior mesenteric vein and superior mesenteric artery with some thickening of descending/transverse segments of duodenum. Endoscopic retrograde cholangio pancreaticography (ERCP) and stenting was performed. Specimen was taken from peri-ampullary growth which showed small cell carcinoma of pancreas. Immuno-histo-chemical stains cytokeratin and CD 56 were positive and Ki-67 proliferative index was 80% (**Figure 1**).

Patient was discussed in multi-disciplinary team meeting and the recommendation was to start induction

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**Keywords** Pancreatic Carcinoma; Pancreatic Cancer; Pancreas  
**Abbreviations** SCLC Small cell lung carcinoma; SCCP Small Cell Carcinoma of the Pancreas; CT-Scan Computed Tomography Scan; ERCP Endoscopic Retrograde Cholangio-Pancreaticography; CEA Carcinoembryonic Antigen; ICI Immune Checkpoint Inhibitor

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chemotherapy (cisplatin and etoposide) and then assessment for surgery. Six cycles were given. Post treatment computed tomographic scan (CT) was performed which showed progressive disease with multiple liver metastases and the disease was approaching right renal pelvis. Bone scan showed no metastases. She was offered 2nd line chemotherapy along with radiotherapy which she refused. She was referred to palliative team for supportive management. She died after 7 months of treatment (Figures 2 and 3).

## DISCUSSION

Small cell carcinoma is an uncommon neoplasm with



Figure 1. Endoscopy Showing Disease Involving D2 Segment of Duodenum.



Figure 2. CT Scan Abdomen with Disease with Head of Pancreas.



Figure 3. CT Scan: Progressive Disease of Pancreas with Hepatic Metastases.

only a few cases reported in literature. In Mayo clinic, 202 patients with pancreatic carcinoma autopsied, only 2 small cell carcinoma of pancreas were found [18]. Cubilla and Fitzgerald found seven small cell carcinomas at Sloan Kettering Cancer Center from 1949 to 1972 out of 508 patients with pancreatic malignancies [19]. Over a period of 29 years, out of 16,585 autopsies, Reyes and Wang found 5 small cell carcinoma [13].

In our study, patient presented with history of jaundice secondary to peri-ampullary lesion causing obstructive symptoms. Small cell carcinoma of pancreas is infrequently associated with para-neoplastic syndrome. Some published case reports highlighted this presentation among patients with this cancer [20]. Since small cell carcinoma arises in lung, imaging is essential in determining the location of disease. Imaging technique, such as CT scan is helpful in diagnosing and staging of the disease. Pre-operatively, it is difficult to distinguish between pancreatic cancer and small cell carcinoma of pancreas on the bases of imaging scans.

Due to the aggressive nature of disease, the presentation of disease is late. As a result, patient presents with advanced disease or with metastases. Thus, in most of the patients, surgical resection is not possible. Immuno-histo-chemical markers play role in diagnosing this distinct cancer. The specimen was positive for CD 56 and cytokeratin in our study. Neuron specific enolase concentration was seen increased in some cases hence it can be used for the diagnoses and assessment of treatment in patients with small cell carcinoma of pancreas [21, 22, 23]. CEA levels can be used for the assessment of treatment response but its role in defining the diagnosis of disease is not clear as it is not specific [22].

Small cell carcinoma of pancreas is a systemic disease which needs multi-modality treatment. There is no standard treatment for the management. Chemotherapy is the preferred choice of treatment. Cisplatin and etoposide was the commonest regimen used in managing the disease. In 1989, Marrant et al reported complete remission after cisplatin and etoposide [24]. Surgery alone is not sufficient for the management of disease.

In our patient, survival was 12 months after chemotherapy. The longest survival is 173 months after diagnosis [25]. The median survival was 20 months which is comparable with ductal adenocarcinoma of pancreas [26]. The most frequent metastatic site is peri-pancreatic lymph nodes (62%), liver (38%), lungs (14%), bone marrow (14%), bone (10%), colon (10%), and the adrenal gland (10%); rarer sites included the spleen, gallbladder, kidney, skin and brain [27]. In our report, patient presented with multiple hepatic metastases after 7 months.

Another new development in the treatment of small cell lung cancers is Immune checkpoint inhibitors which are being investigated for the treatment of metastatic small-cell lung cancer with positive outcome. Recent clinical trials have demonstrated some favourable response of

SCLC to treatment with the immune checkpoint inhibitor (ICI) nivolumab. Emerging data suggest that ICIs may have meaningful clinical activity in SCLC. The initial result of the CheckMate 32 trial showed that patients with advanced-stage SCLC who progressed after one or more platinum-based chemotherapy experienced durable objective response with either single-agent immunotherapy with nivolumab or combination of nivolumab and ipilimumab [28]. A case report has previously reported an excellent result for a trial of nivolumab on a patient with recurrent metastatic PD-L1-negative small-cell neuroendocrine carcinoma of the cervix, another rare type of extra-pulmonary small-cell cancer [29]. In that case, treatment was limited due to side effects. Although SCLC is not yet an approved indication for ICIs, a search on trials.gov indicates that studies are moving ahead to explore the numerous ways ICIs can be used in the treatment of SCLC including their simultaneous combination with other treatment modalities. Information derived from these studies will likely place ICIs in the centre stage of advanced SCLC treatment and without doubt advance the treatment of closely related extra-pulmonary small-cell neuroendocrine tumours such as SCCP.

## CONCLUSION

Rarity of disease makes it difficult to draw conclusion from this case report. However, it will be an addition in the literature for better understanding of the disease. Prospectively, meta-analyses and systemic reviews are required to understand it.

## CONSENT

IRB (Institutional review board) approval was taken for publication of this case report and any accompanying images.

## Conflicts of Interest

All named authors hereby declare that they have no conflicts of interest to disclose.

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