

Primary Mantle Cell Lymphoma of Pancreas- Rare Case

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DOI: [10.36347/sjmcr.2022.v10i01.003](https://doi.org/10.36347/sjmcr.2022.v10i01.003)

| Received: 05.12.2021 | Accepted: 09.01.2022 | Published: 14.01.2022

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Abstract

Case Report

Primary pancreatic lymphomas (PPLs) are rare cancers that have a complicated histological classification. Sufficient sample volumes are required to adequately diagnose and decide the best treatment for PPLs. A 54-year-old man with primary pancreatic mantle cell lymphoma is described in this case study. Percutaneous biopsy was used to extract histological samples. Atypical small to medium round cells predominated in the tumor cells, which had diffuse CD20 and cyclin D1 positivity. Chemotherapy was used on the patient, and he went into complete remission as a result. Because it provides adequate samples for testing, a percutaneous biopsy could be a valuable and safe diagnostic tool for PPLs.

Keywords: Primary pancreatic lymphoma, Endoscopic ultrasound-guided fine-needle aspiration, Pancreatic tumor.

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INTRODUCTION

Only about 2% of all extranodal lymphomas and 0.5 percent of all pancreatic masses are primary pancreatic lymphomas (PPLs) [1]. A PPL is a lymphomatous mass that is localized to the pancreas and manifests as a big bulky mass in the pancreatic head [2]. In 44 individuals, the median diameter of PPLs was 7.9 cm (range 2.4–13.9 cm) according to a prior study [3]. Chemotherapy for PPLs has a high complete response rate [3], but proper pathological diagnosis necessitates a large sample size. Tumor biopsy by laparotomy obtains sufficient samples, however, it is invasive.

Despite the fact that endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) is a less invasive way for extracting tissue from pancreatic tumours [4], its effectiveness in identifying PPLs is questionable. We present a case of pancreatic mantle cell lymphoma in which the use of Ultrasound guided Percutaneous biopsy helped to make a correct diagnosis and choose an effective treatment.

CASE PRESENTATION

A 52-year-old man with abdominal pain and non-bilious vomiting presented to the outpatient

department. Over the course of two weeks, he noticed greater fatigue and an unintended weight loss of 5 kg. He also mentioned postprandial fullness and a lack of appetite. There was no recent foreign trip history, risky sexual behaviour, animal contact, or drug use history. He had not started any new prescriptions in the previous six months and was not taking any herbal remedies.

Hypertension and Type 2 Diabetes Mellitus were among his medical issues. He was on amlodipine and metformin at the time. He had an unremarkable surgical history and no medication sensitivities. He had a 5-pack-year smoking habit but did not drink alcohol.

On closer observation, he appeared pale, but there was no sign of mental illness. Temperature 36.4°C, blood pressure 120/90 mm Hg, heart rate 68 beats per minute, respiratory rate 14, and oxygen saturation 98 percent on air were among his findings. Physical examination revealed minor epigastric soreness, which was categorised as a 4/10.

Acetaminophen helped him feel better. A soft, reducible little umbilical hernia (defect 1cmx1cm) was found. The results of the respiratory and cardiovascular exams were unremarkable.

Investigations

Normal liver function tests were verified by laboratory tests. He had a negative hepatitis test.

With a lactate of 0.8 mmol/L (0.5–2.1 mmol/L) and a pH of 7.395 (7.32–7.48), his venous blood gas revealed normal physiology. Urinalysis was normal study. There was no evidence of air beneath the diaphragm on his chest radiograph.

A low attenuation mass in the body and tail of the pancreas measured 15cmx15 cm on a CT scan of the abdomen and pelvis with intravenous contrast, which was suggestive for malignancy (Figure 1). The tumour engulfed the pancreas' distal body and tail, with no dilation of the pancreatic duct. There were no visible lymph nodes in the hilar, mediastinal, or axillary regions, nor were there any lung or localised destructive bone lesions. The liver, spleen, adrenal glands, gall bladder, kidneys, bladder, and prostate gland all appeared to be in good working order. The retroperitoneal and mesenteric nodes were found to be somewhat enlarged. Pancreatic cancer (adenocarcinoma, lymphoma) with nodal metastasis was among the possibilities. His tumour markers were normal (AFP, Ca 125, CA 19-9, and CEA).



Fig 1: CT abdomen and pelvis with intravenous contrast showed large low attenuation mass in body and tail of pancreas measuring 15cm x15cm

Using fluorine-18-fluorodeoxyglucose (FDG) positron emission tomography (PET)-CT, FDG uptake was found throughout the mass in the distal body and tail of the pancreas (maximum standardised uptake value: 11.93) (Fig 2).

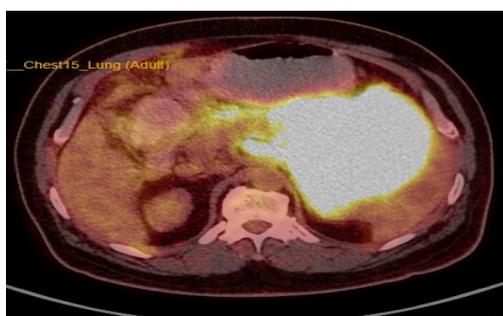


Fig 2: FDG PET-CT images. FDG PET-CT showed FDG uptake in the whole pancreatic lesion (SUV max of 11.93). FDG, fluorine-18-fluorodeoxyglucose; PET-CT, positron emission tomography/computed tomography; SUVmax, maximum standard uptake value

Ultrasound guided Percutaneous biopsy with a 18-gauge needle was performed to assess the pancreatic tail mass.

The tumor cells were mostly made up of atypical small to medium round cells with a high nuclear/cytoplasmic ratio, with no Hodgkin or Reed-Sternberg cells (Fig 3 & 4). Immunohistochemically (Table 1), the lesional cells are positive for CD20 (Fig 11), Cyclin D1 (Fig 6), CD45 (Fig 5), CD5 (Fig 7) and Bcl-2(Fig 5). They are negative for CD3 (Fig 8), INSM1, Panck, CD10, MUM1, Bcl-6. Ki- 67 (Fig 10) displays a proliferation index >60%. These morphologic and immunohistochemical features are diagnostic for Mantle cell lymphoma.

Table 1

Antibody	Result
C20	Positive in lesional cells
CD45	Positive in lesional cells
CD3	Positive in Background T cells
CD5	Positive in lesional cells
BCL2	Positive in lesional cells
CYCLIN D1	Positive in lesional cells
INSM1	Negative in lesional cells
Panck	Negative in lesional cells
CD10	Negative in lesional cells
MUM1	Negative in lesional cells
BCL6	Negative in lesional cells
Ki 67%	>60% Positive

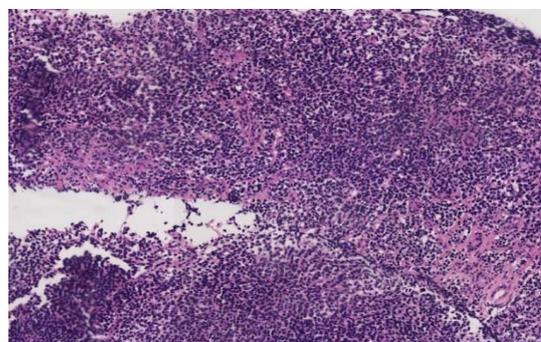


Fig 3: Large lymphoid cells with frequent mitotic figures and apoptotic bodies. Low power, x10

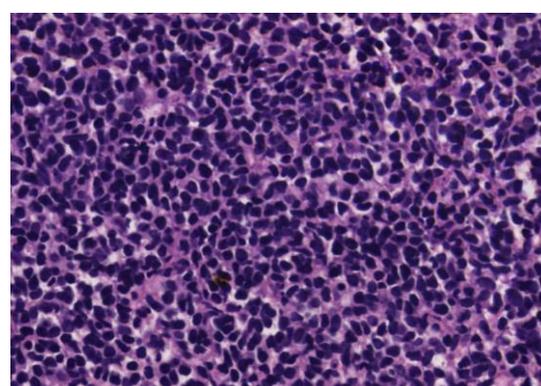


Fig 4: Large lymphoid cells with frequent mitotic figures and apoptotic bodies. High power, x 40

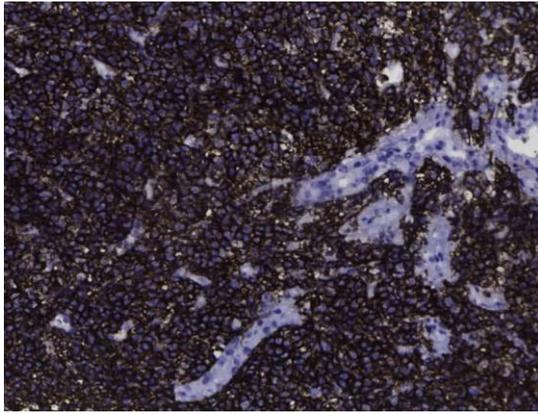


Fig 5: CD45 Positive

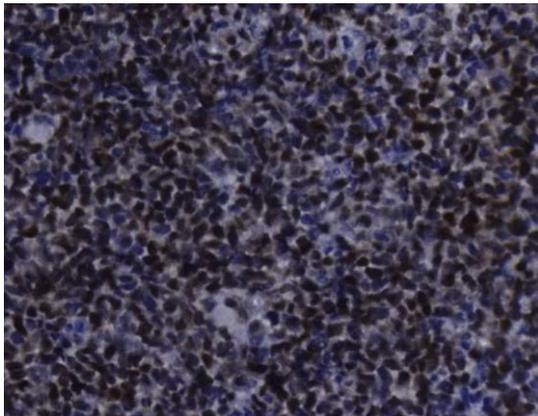


Fig 6: CYCLIN D1 Positive

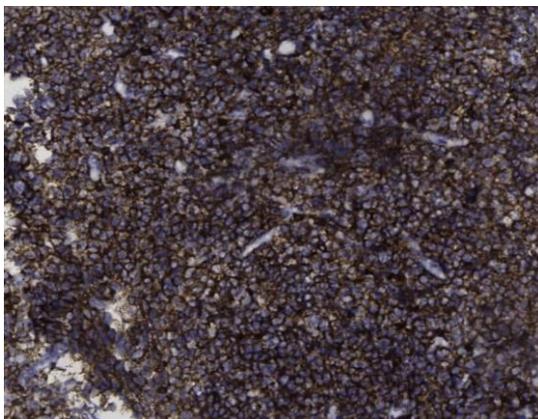


Fig 7: CD5 Positive

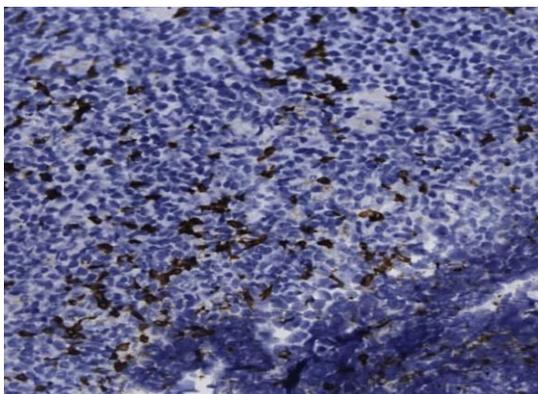


Fig 8: CD3 Positive Iin background T cells

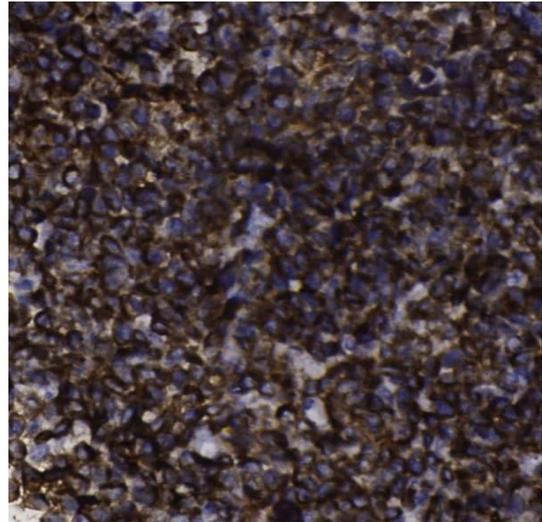


Fig 9: BCL2 Positive CD5

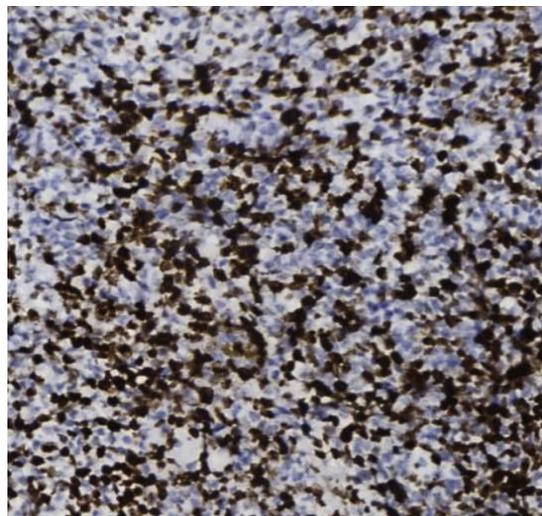


Fig 10: KI67 >60% Positive

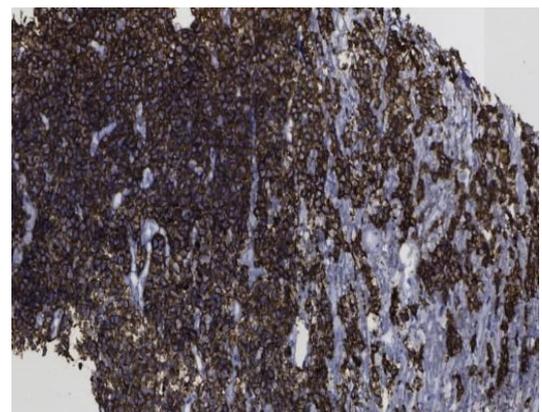


Fig 11: CD 20 immunostain showing B cells

According to the International Prognostic Index, these findings led to a final diagnosis of mantle cell lymphoma. He was in complete remission after 9 months of treatment with rituximab, pirarubicin, cyclophosphamide, vincristine, and prednisolone (R-THP-COP). PET-CT demonstrated FDG uptake in the pancreas and para-aortic lymph nodes one year and nine months after first-line treatment was stopped, indicating

recurrence. He attained complete remission again after receiving the rituximab-bendamustine combo as a second-line treatment. The patient was doing well two years after starting first-line treatment and had no recurrence.

DISCUSSION

We provide a case of a patient who achieved long-term survival after chemotherapy after receiving an accurate pathological diagnosis of mantle cell lymphoma using ultrasound guided percutaneous biopsy. Because the head of the pancreas has more lymphoid tissues [7], a PPL is typically observed as a bulky mass in the head of the pancreas [6]. This is not the case in our situation.

Berhns *et al.*, [8] proposed the following criteria for PPLs:

1. No palpable superficial lymphadenopathy.
2. No enlargement of mediastinal lymph nodes.
3. Normal leukocyte count.
4. At laparotomy, the pancreatic mass predominates with grossly involved lymph nodes confined to the peripancreatic region.
5. No hepatic or splenic involvement.

In combination with the MRI data, the patient allegedly met the criteria outlined above. A modestly contrast-enhanced tumor localized to the distal body and tail of the pancreas was visible on the contrast-enhanced CT imaging. PET-CT scans, on the other hand, showed FDG buildup throughout the pancreatic tail lesion.

Clinical signs associated with a PPL were not observed in this case. Epigastric discomfort, nausea, exhaustion, and anorexia are recorded as clinical subjective symptoms of a PPL [3]. Severe fever, night sweats, and weight loss, which are common in other lymphomas (i.e., B signs), are uncommon in PPLs [9].

The current instance was mildly symptomatic and discovered during abdominal ultrasonography. The use of EUS-FNA in the differential diagnosis of pancreatic disorders has been proven to be effective [10]. Although a meta-analysis found that EUS-FNA had a high sensitivity (86.8%) and specificity (95.8%) for solid pancreatic tumors [11], it has been reported that histological examination with EUS-FNA is not suitable for the diagnosis of PPLs due to the difficulty in obtaining an adequate sample amount [12]. When compared to other pancreatic tumors, such as pancreatic ductal adenocarcinoma, a considerable volume of sample is required for the definite diagnosis of PPLs [13]. Flow cytometry and immunohistochemistry improve the diagnostic efficiency of PPLs when compared to cytology alone.

Although laparoscopic biopsies for the pathological analysis of PPLs have been undertaken, the

laparoscopic examination is an intrusive technique since it requires general anesthesia and takes a long time (about 1–4 h) [14]. In our case, we were able to successfully perform immunostaining on specimens obtained through Ultrasound guided percutaneous biopsy, implying that it could be a useful and safe method for determining the definitive diagnosis of PPLs.

CONCLUSION

We report a rare case of a primary pancreatic mantle cell lymphoma diagnosed via Ultrasound guided percutaneous biopsy, which provided a sufficient amount of sample for histological examination. Accurate histological findings helped to avoid unnecessary surgery and contribute to the selection of appropriate treatments. Ultrasound guided percutaneous biopsy could be a useful and safe diagnostic modality in pancreatic tail lesions where the differential diagnosis of a PPL is considered.

Statement of Ethics: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement: The authors disclose no conflicts of interest.

Funding Sources: No specific funding has been received for this study.

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Final approval of the manuscript: all authors

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