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Pylorus-preserving Pancreaticoduodenectomy with Reconstruction of Portal Vein for Primary Pancreatic Lymphoma: One Case Report

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Abstract Primary pancreatic lymphoma is a rare disease. It is insidious, always no specific symptoms and signs, and easy to be misdiagnosed as pancreatic carcinoma. Now 1 case of primary pancreatic lymphoma is reported in our hospital for sharing.

Keywords: *primary pancreatic lymphoma, diffuse large B-cell lymphoma, pancreatic carcinoma, diagnosis, treatment, CA -199*

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1. Introduction

Primary pancreatic lymphoma is a rare disease, account for only 0.2% of cancer of the pancreas [1]. It is difficult to diagnose, and it is easy to be misdiagnosed as pancreatic carcinoma, due to it has no specific symptoms and signs. But its prognosis is better than that of pancreatic carcinoma. We report a case of PPL in an adult patient with high uptake of FDG.

2. Case Report

The patient is a 52-year-old gentleman with a good past medical history. He underwent laparoscopic cholecystectomy for adenomyomatosis of the gallbladder (ADM). After the operation, he presented with abdominal pain and jaundice 4 days later. Physical examination revealed all skin and sclera were slightly yellow stained, right lower quadrant pain. Right abdominal tenderness, no rebound pain. And peripheral lymphadenopathy was not detected. After admission to the hospital again, his lab work showed a total bilirubin 73.9, CA199 215.8; Contrast MR showed a 39×38×44mm heterogeneously enhanced mass (e.g. Figure 1) in the pancreatic head, considering malignant tumor, which was closely related to the superior mesenteric vein, does not see wrapping around SMV/PV. CBD and pancreatic duct are dilated, no focal enhanced mass was observed in the liver, but with some collection in the abdominal cavity. For further evaluation, F – 18 FDG PET/CT was done, and it (e.g. Figure 2) showed a high hyperintensity of mass with high uptake (about 22.8 standardized uptake value [SUV]) SUV max = 22.8, a malignant tumor was suspected, biopsy recommended.

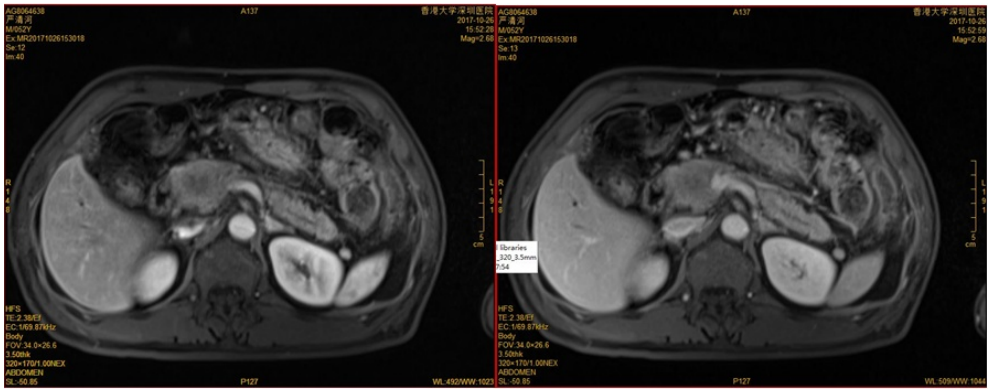

Figure 1. Radiographic findings: axial MR. Pancreatic head mass close to IVC

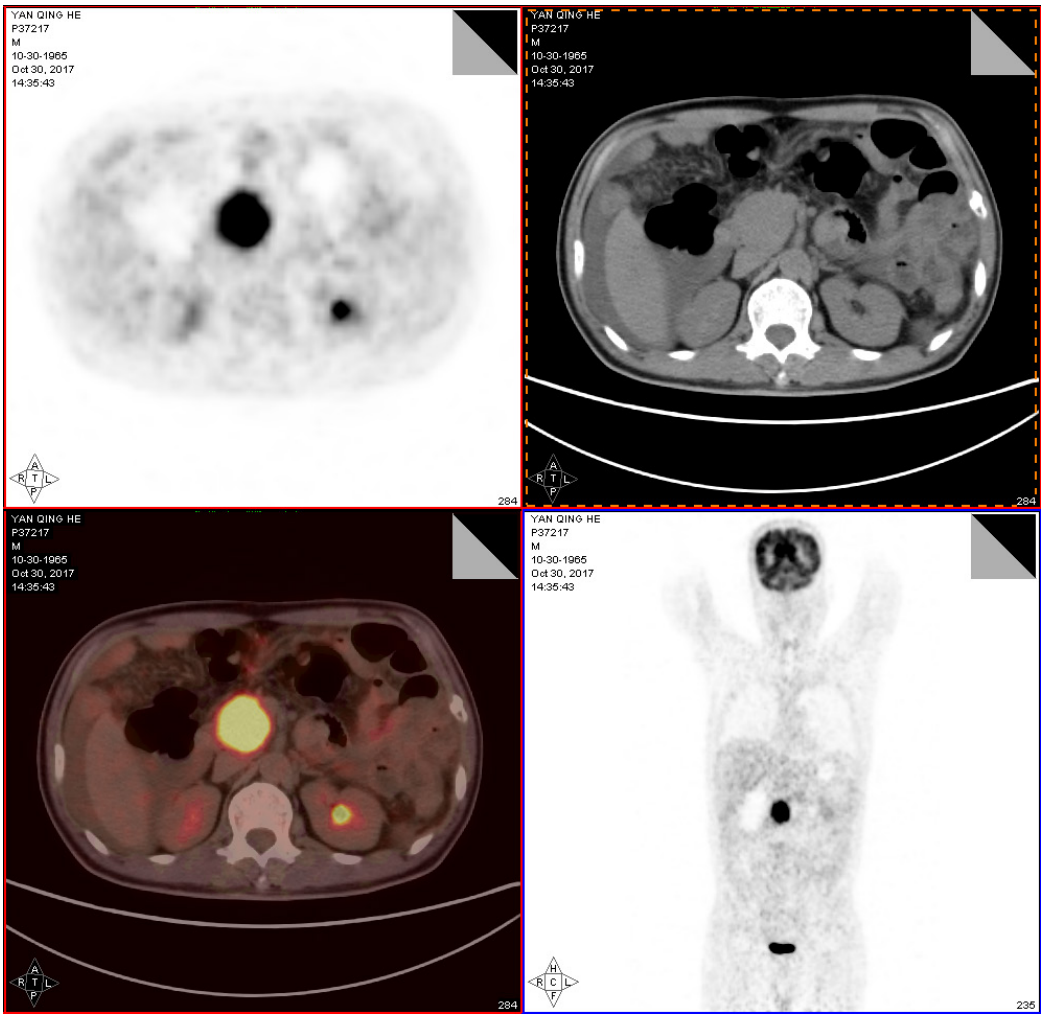

Figure 2. For further evaluation, F – 18 FDG PET/CT was done, Scintigraphy revealed a hypermetabolic lesion (arrow), which had intense FDG uptake, SUV max = 22.8. corresponding to the lesion on abdominal CT

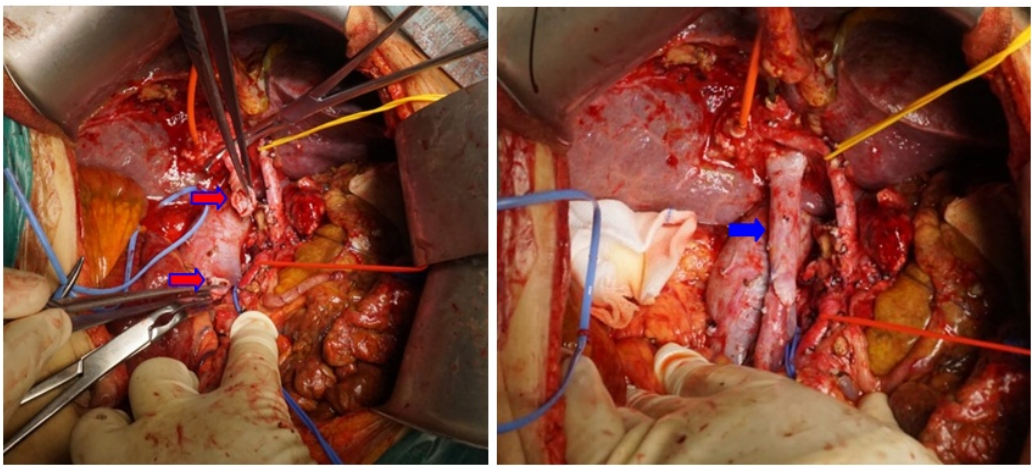

Figure 3. Intraoperative photo, portal vein end (red arrow), portal vein reconstruction(blue arrow)

The initial diagnosis was pancreatic head cancer. Whipple operation was arranged for the patient. And we found SMV was involved about 5cm intraoperatively, and it was too long, so we performed Pylorus-preserving pancreaticoduodenectomy (e.g. Figure 3) with the reconstruction of the portal vein for the patient. Postoperative pathology report confirmed diffuse large B-cell lymphoma (e.g. Figure 4) and histological report showed A7:MNF-166 (-), CD3 (+;small lymphocytes), CD20 (+;large neoplastic cells); All: CD20 (+;large neoplastic cells), CD3(+;small lymphocytes), Bcl-2(+), Bcl-6(-), CD10(-), Ki-67(index 80%), CD79α(+), PAX-5(+), MNF-166(-); A5: IgG(+), IgG4(+); rare plasma cells).

Then the patient was transferred to hematology department and received five cycles of chemotherapy with doxorubicin, cyclophosphamide, vincristine, and prednisolone (CHOP regimen), Seven months after the operation, he underwent autologous peripheral blood stem cell transplantation. After that, he is still alive and asymptomatic after followed with 10 months.

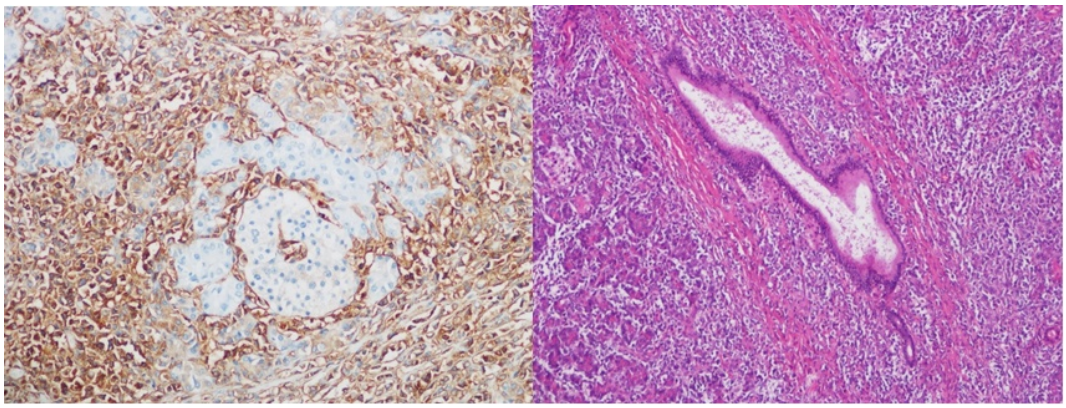

Figure 4. Pathology of PPL: Diffuse lager B cell lymphoma, non-germinal center type

3. Discussion

Primary pancreatic lymphoma (PPL) account for 0.2 % [1] of all primary tumors of the pancreas and approximately 0.1 % of all malignant lymphomas. Of these, 58% were males, and the median age was 65-69 years, significantly higher than the average age pancreatic carcinoma. Among them, diffuse large B-cell lymphoma is the most common type, accounting for about 56% [2,3]. PPL originates more commonly in the head of the pancreas [4], which has the greatest amount of lymphoid tissue.

Clinical symptoms of primary pancreatic lymphoma are not specific [5], which mimic with pancreatic carcinoma, The most common symptoms for PPL are pain 73%, weight loss 45%, and jaundice 18%, while patients with adenocarcinoma presented with pain 52% (P = 0.3), weight loss 30% (P = 0.47) and jaundice 76% (P = 0.001).

PPL is often misdiagnosed as pancreatic cancer [6], Because of its clinical manifestations, there is no obvious difference between the examination and pancreatic cancer. It is reported that about 25% of PPL patients have an abnormal elevation of CA 19-9, but at the same time there is an increase in LDH [2]. In terms of CT performance, PPL is also difficult to distinguish from pancreatic cancer, but Teefey, S.A et al. and Boninsegna, E et al. [7,8] considered that the size of the tumor may provide some diagnostic significance. The general volume of PPL is more larger than pancreatic cancer. It is reported that the diameter is even more than 10cm, while the diameter of pancreatic cancer is less than 10cm mostly.

At present, there are few reports on the diagnostic value of PET/CT for PPL. Only two articles describe the role of PET/CT in the diagnosis of PPL. Yoon, S.N.et al. [9] considered that routine examination methods, such as MR and CT, are difficult to identify rare tumors such as PPL from pancreatic carcinoma. They reported a 67-year old male patient with PPL whose PET/CT presented a round, high-density F-18 FDG uptake. In addition, Abe, Y et al. [10] also report A case of a 56-year old patient with atypical CT and MR findings, but his PET/CT showed a high metabolic intake and SUV max reached 10.3. one case report from Bai Y et al. [11] considered, such as the diffuse large B cell lymphoma of the lung, which is not in the pancreas, also has a higher uptake on PET/CT, with a SUV max value of 14.7. PPL have tended of markedly FDG avid, Awareness of the feature of PPL will enable the radiologist to give the correct diagnosis [12]. In our case, we found nodular and radioactive abnormal concentration foci, which SUV max value was 22.8.

Because PPL is different from pancreatic cancer in prognosis and treatment, preoperative diagnosis is very important. Ackerman, N.B.et al. [13] analyzed PPL and similar tumors, and It is considered that biopsy should be paid attention to. Tissue biopsy or exploratory laparotomy under the guidance of CT have used in the past time [14]. But In recent years, with the development of EUS, the method of diagnosis has been gradually popular through obtaining tissue specimens from EUS under FNA. A retrospective study in a single center showed that FNA had a higher diagnostic accuracy, a sensitivity of 88% and a false negative rate of 4%, which published by Zhou J et al. [15]. Khashab, M et al. [16,17], considered that the sensitivity of FNA examination combined with cytologic examination under EUS for PPL is significantly higher than that of FNA alone. Grimison, P. S et al. [15] considered that the biopsy of the pancreatic tumor is very necessary, so that the laparotomy can be avoided effectively.

The treatment of PPL is still controversial. PPL is currently considered to be a potentially curable malignant tumor of the pancreas. Tuchek, J.M et al. [18] study showed that patients who received postoperative chemotherapy survived for an average of 6.3 years, and the treatment of untreated PPL remains controversial, and PPL is currently considered a possible cure for pancreatic malignancies with an average survival of 5 months. Battula, N et al. [19] believed that the combination of pancreatoduodenectomy and chemotherapy provides a radical cure for the patients. Lin, H et al. [17] analyzed the prognosis of 6 patients with PPL. And the results showed that surgery and radiotherapy or chemotherapy were effective. But Grimison, P. S et al. [15] and Yoon, W.J. et al. [20] considered not to have surgery but to combine multiple treatments, such as radiotherapy and chemotherapy, which have better clinical outcomes.

4. Conclusion

PPL is a malignant tumor with cure potential. Because its clinical manifestation is not specific, it is often misdiagnosed as pancreatic carcinoma. But we can still get a definite diagnosis through CT, PET/CT, even FNA and FCM examination under EUS. If CT and MR are difficult to identify pancreatic tumors, but PET/CT

indicates that SUV max is too high, it is likely to be PPL, and then biopsy under EUS may be necessary. It is controversial whether this type of patient needs surgery, but whether or not it is operated, it is recommended that the patient should be treated with appropriate chemotherapy to achieve a good long-term effect.

Abbreviations

PPL: primary pancreatic lymphoma; ADM: adenomyomatosis of gallbladder; SMV: superior mesentric vein; PV: portal vein; CBD; common bile duct; PET: positron emission tomography; SUV: standard uptake value. FDG: fluorodeoxyglucose.

Authors' Contributions

CH Liu, analyzed and interpreted the patient data. All authors contributed to writing the manuscript. All authors read and approved the final manuscript.

Ethics Approval and Consent to Participate

Not applicable.

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing Interests

The authors declare that they have no competing interests.

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