

## CASE REPORT

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# Follicular Lymphoma of the Pancreas: A Case Report and Proposed New Strategies for Diagnosis and Surgery of Benign or Low-Grade Malignant Lesions of the Head of the Pancreas

Naohiro Sata, Akira Kuroguchi, Kazuhiro Endo, Kunihiko Shimura, Masaru Koizumi, Hideo Nagai

Department of Surgery, Jichi Medical University. Tochigi, Japan

### ABSTRACT

**Context** Primary pancreatic lymphoma is a rare form of extranodal lymphoma originating in the pancreas. The present report describes a case of follicular lymphoma of the pancreas with unique CT and MRI findings.

**Case report** A 58-year-old male complained of sudden abdominal pain, and routine ultrasonography detected an 8 cm hypoechoic tumor in the head of the pancreas. The 3D image generated using multi-cholangiography and virtual duodenography provided the information necessary for a laparotomy. The tumor was enucleated for diagnosis. Follicular lymphoma is quite rare in the pancreas and gastrointestinal tract. A considerable number of pancreatic lymphoma subtypes have been reported. The expression “pancreatic lymphoma” has been used to describe both primary lymphoid neoplasms originating in the pancreatic parenchyma and tumors

invading from a peri-pancreatic lymphadenopathy. The present case belongs to the latter, which might explain the unique imaging findings and histological type. These subtypes display different imaging findings and different clinical characteristics. In the future, primary pancreatic lymphoma should be discussed separately depending on the subtype.

**Conclusion** We propose a new subtype of primary pancreatic lymphoma. Multi-cholangiography and virtual duodenography provided the information necessary for a laparotomy in the present case. Enucleation is indicated for benign and low-grade malignant tumors of the pancreas, even if the tumor is located in the head of the pancreas.

**Table 1.** Behrns *et al.* criteria for primary pancreatic lymphoma [8].

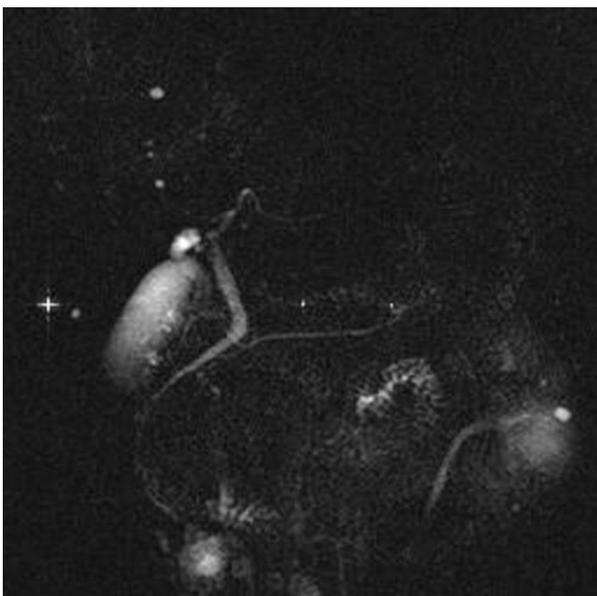
- 1.No palpable superficial lymphadenopathy
- 2.No enlargement of mediastinal nodes
- 3.Normal leukocyte count
- 4.At celiotomy, the pancreatic mass predominates, with grossly-involved nodes confined to the peri-pancreatic region
- 5.No hepatic or splenic involvement

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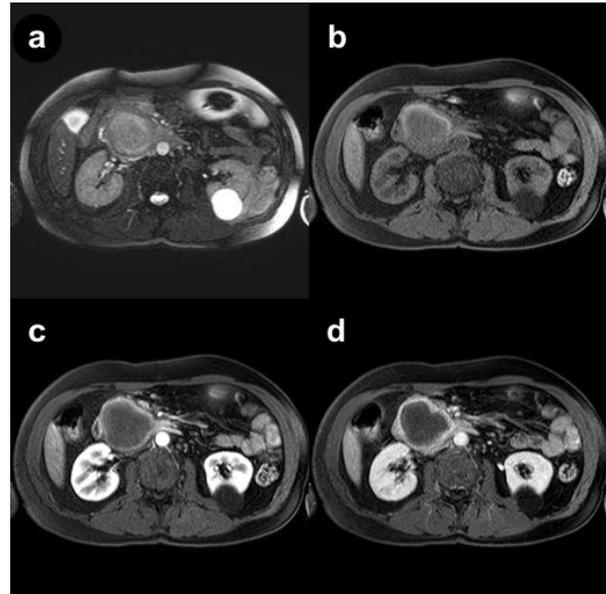
### INTRODUCTION

Primary pancreatic lymphoma (PPL) is a rare form of extranodal lymphoma originating in the pancreas, and constitutes less than 0.5% of all pancreatic malignancies [1, 2, 3, 4, 5, 6]. Some confusion surrounds the definition of PPL. Dawson *et al.* published the first criteria for the diagnosis of a primary lymphoid tumor of the intestinal tract [7]. As Dawson’s criteria did not refer to pancreatic lymphomas, Behrns *et al.* modified Dawson’s original criteria and applied it to PPL (Table 1) [8].

Salvatore *et al.* proposed new criteria for PPL and a novel nomenclature system, including primary, secondary and tertiary pancreatic lymphoma definitions [5]. They limited primary pancreatic lymphomas to lymphoid tumors originating in the pancreatic parenchyma, and defined secondary pancreatic lymphomas as peri-pancreatic lymphadenopathy invading the pancreas. In the literature, these two types of pancreatic lymphoma have both been referred to as PPL. Pancreatic lymphomas usually appear as large pancreatic masses, and sometimes mimic pancreatic cancer or other pancreatic neoplasms. PPLs appear as low-density and heterogeneous lesions when viewed using plane CT, show poor yet homogeneous enhancement when viewed using dynamic CT, show low signal-intensity with subtle enhancement after i.v. gadolinium-contrast medium administration when viewed using T1-weighted MR imaging, and show heterogeneous low signal-intensity when viewed using T2-weighted MR imaging [9, 10]. In the latest review, Saif reported two PPL prototypes in image findings, a localized well-circumscribed tumoral form and a diffuse enlargement infiltrating or replacing most of the pancreas [6], and these appear to represent the primary and secondary pancreatic lymphomas proposed by Salvatore *et al.* [5].



**Figure 1.** MRCP showing encasement of the common bile duct and main pancreatic duct.

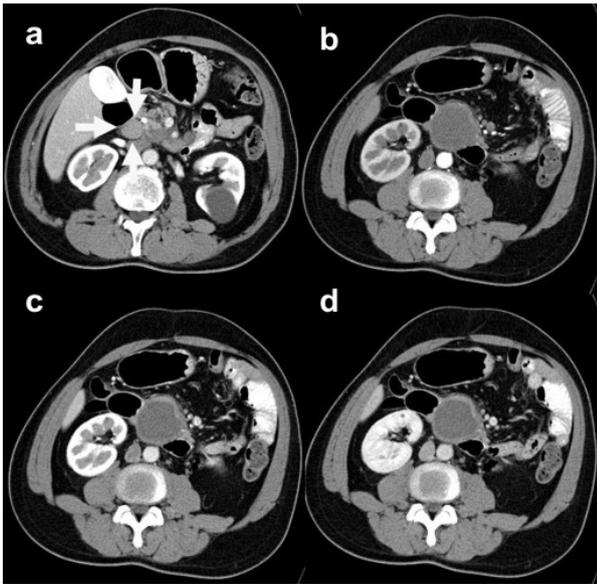


**Figure 2.** The tumor showed heterogeneous iso- or low signal-intensity on T2-weighted images, and low signal-intensity with marginal enhancement on T1-weighted images after i.v. administration of gadolinium-contrast medium. **a.** T2-weighted MR image. **b.** T1-weighted MR image. **c-d.** Postcontrast T1-weighted MR image.

The present report describes a unique case of PPL. In this case, a novel method of multi-detector raw CT (MD-CT) examination and post-processing was used in the preoperative diagnosis and enucleation of the tumor, and contributed to the histological diagnosis.

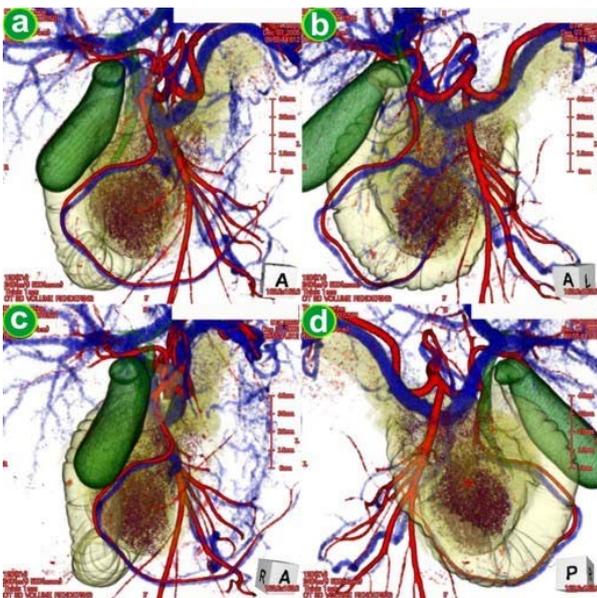
## CASE REPORT

A 58-year-old male complained of sudden abdominal pain, and routine ultrasonography detected an 8 cm hypoechoic tumor in the head of the pancreas. The tumor was carefully examined using MRI and MD-CT. MRCP showed encasement of both the common bile duct and the main pancreatic duct (Figure 1). The tumor showed heterogeneous iso- or low signal-intensity on T2-weighted images, and low signal-intensity with marginal enhancement on T1-weighted images after i.v. administration of gadolinium-contrast medium (Figure 2). Ordinary CT demonstrated a low density tumor with marginal enhancement in a dynamic study of the head of the pancreas (Figure 3). There appeared to be neither hepatic nor splenic involvement. A 2 cm lymph node was detected in the retro-



**Figure 3.** a. Enhanced CT of the retro-pancreatic lymph node (white arrows). b-c-d. Dynamic CT image study of the tumor in the head of the pancreas showing weak marginal enhancement of the tumor.

pancreatic region (Figure 3a white arrows). The patient then underwent a distinct diagnostic procedure involving MD-CT scanning and post-processing, multi-cholangiography and virtual duodenography [11]. This process allowed the tumor (which appeared as dark purple according to the window level) to be clearly observed in a 3D

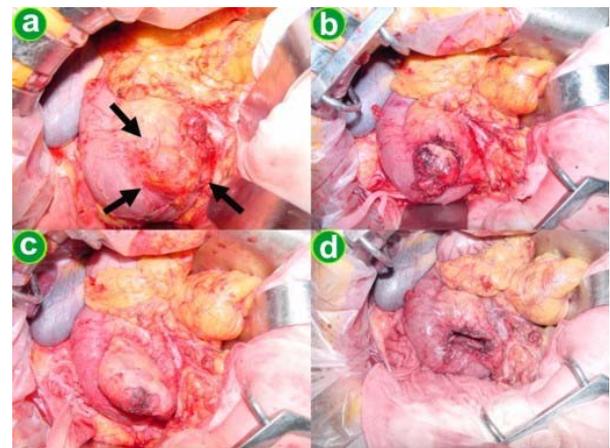


**Figure 4.** A multi-cholangiography and virtual duodenography image. The tumor appears as dark purple (according to the window level) in the 3D image. a. Anterior view. b. Left oblique view. c. Right oblique view. d. Posterior view.

manner (Figure 4), and allowed interaction with the adjacent organs and vessels to be viewed. The tumor was shown to be in the parenchyma of the head and the uncus of the pancreas, and there appeared to be no portal vein or artery involvement.

No peripheral lymph nodes were palpable and no lymphadenopathy in the pleural and abdominal cavity was detected other than the retro-pancreatic lymph node described above. Biochemical analysis and peripheral blood cell counts on entry showed normal results. Pancreatic hormones, enzymes and tumor markers (including soluble IL-2 receptor) were at normal levels.

A preoperative diagnosis of a benign or low-grade malignant lesion of the pancreas, such as an endocrine tumor or neurogenic tumor provided the information necessary for a laparotomy. The tumor, located in the head of the pancreas, was elastic, hard, covered with pancreatic tissue, and clearly bordered the pancreatic parenchyma according to intraoperative ultrasonography findings. The first procedure involved the removal of a 2 cm retro-pancreatic lymph node. While intraoperative histological examination indicated that the tumor was a lymphoid proliferative mass, a definitive diagnosis could not be made. The border between the tumor and the pancreas was clear. The tumor was then enucleated, a procedure which was



**Figure 5.** Intraoperative findings. a. The tumor was located in the head of the pancreas and was covered with pancreatic tissue (black arrows). b-c. The enucleation procedure for the tumor. d. The view after tumor enucleation.



**Figure 6.** Macroscopic findings of the tumor.

relatively easy and took approximately thirty minutes (Figures 5 and 6). The histology of the tumor was almost the same as the peripancreatic lymph node.

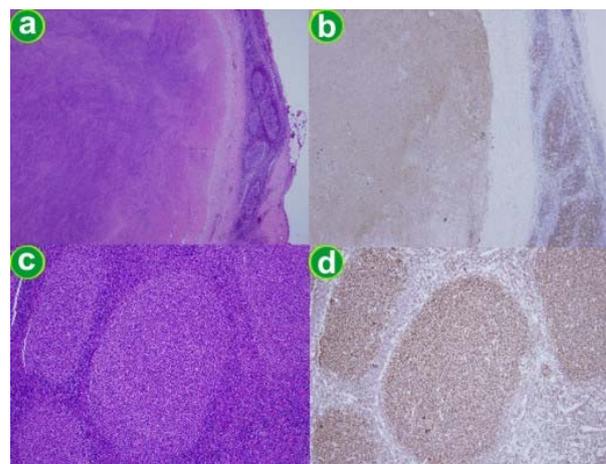
The patient was discharged on the 12<sup>th</sup> post-operative day without any significant postoperative events. The final histologic diagnosis was a B-cell type follicular lymphoma of the pancreas. The tumor had a fibrous capsule and displayed diffuse central necrosis. Most of the non-necrotic tissue in the peripheral region consisted of proliferating follicles, and immunohistochemical analysis found it to be stained positive for BCL2 gene, L26 protein and CD10 antigen (Figure 7). As the lymphadenopathy was located beside the pancreas, this case was categorized as Stage II using the Ann Arbor Staging system [12]. The patient received additional radiation and showed no signs of recurrence over the following 6 months.

## DISCUSSION

The present report describes a unique case of PPL. The tumor was located in the head of the pancreas, was a peri-pancreatic lesion as indicated by the lymphadenopathy, was compatible with all the criteria of Behrns, and was hence diagnosed as PPL [8]. Most reported cases of PPL in the English-language literature are intermediate or high-grade non-

Hodgkin lymphomas (NHL) with diffuse large cells of the B type [6]. Follicular lymphomas are quite rare in the pancreas and gastrointestinal tract. Shia *et al.* reported 26 cases of primary follicular lymphoma of the gastrointestinal tract, none of which involved the pancreas [13]. Misdraji *et al.* [14] reported a case of follicular lymphoma of the papilla of Vater, and Salvatore *et al.* [5] summarized three cases of follicular lymphoma of the pancreas out of 60 cases in the literature.

In the present case, the tumor showed unique findings according to CT and MRI. Saif summarized PPL imaging findings and concluded that neither calcifications nor necrosis within the tumor mass had been described in any case of untreated PPL [6]. However, the present tumor showed diffuse central necrosis, which explained the unique CT and MRI findings. No particular findings of follicular lymphoma have been reported in the literature. Central necrosis is possibly linked to the rare histologic findings. The 3D multi-cholangiography image provided the information necessary for the laparotomy in the present case. A safe and successful laparotomy procedure requires knowledge of the precise location of the tumor and any interactions with vessels and adjacent organs. The present method provided such information, and we believe it is applicable to



**Figure 7.** Microscopic examination following hematoxylin-eosin staining (a. x10, c. x200) and L26 immunostaining (b. x10, d. x200). The tumor showed central necrosis and L26-positive proliferating follicles in the peripheral region.

all pancreatic tumors including pancreatic cancers and neoplasm of any etiology.

The initial treatment of a PPL depends on the method by which the diagnosis is made. If the diagnosis is made using a relatively mild invasive technique, chemotherapy and/or radiation is initiated soon after diagnosis. Recently, fine needle aspiration or biopsy has become the "gold standard" for PPL diagnosis as it is highly accurate [6, 9]. Recent reports recommend US- and CT-guided biopsy as the first choice diagnostic tool for PPL [9, 15, 16, 17, 18]. The present patient underwent a laparotomy mainly because the unique CT and MRI findings did not indicate a PPL. Even if a tumor is located deep within the head of the pancreas, it is worthwhile attempting enucleation for benign and low-grade malignant pancreatic tumors. Since such tumors usually display expansive growth and low interaction with the pancreatic parenchyma, careful observation and resection can avoid bleeding and leakage of pancreatic juice during enucleation. Enucleation can also be applied to other types of pancreatic tumors, such as solid, pseudopapillary, neurogenic and endocrine tumors.

There has been considerable variation in the reporting of lymphomas involving the pancreas. The expression "pancreatic lymphoma" has been used for both primary lymphoid neoplasms originating in the pancreatic parenchyma and for tumors invading from peri-pancreatic lymphadenopathies [5]. The present case belongs to the latter category. These subtypes display different image findings and different clinical characteristics. In the future, PPL should be discussed separately depending on the subtype.

In conclusion, the present report describes a rare follicular lymphoma of the pancreas. Multi-cholangiography and virtual duodenography provided the information necessary for a laparotomy. Enucleation is indicated for benign and low-grade malignant tumors of the pancreas, even if the tumor is located in the head of the pancreas.

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**Keywords** Lymphoma, Follicular; Pancreatic Diseases

**Abbreviations** BCL2: B-cell CLL/lymphoma 2 [*Homo sapiens*]; MD-CT: multi-detector raw computed tomography; PPL: primary pancreatic lymphoma

### Correspondence

Naohiro Sata  
3311-1 Yakushiji Shimotsuke  
Tochigi, 329-0498  
Japan  
Phone: +81-285.58.7371  
Fax: +81-285.44.3234  
E-mail: sata@jichi.ac.jp

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### References

1. Weingrad DN, Decosse JJ, Sherlock P, Straus D, Lieberman PH, Filippa DA. Primary gastrointestinal lymphoma: a 30-year review. *Cancer* 1982; 49:1258-65. [PMID 7059947]
2. Boyle P, Hsieh CC, Maisonneuve P, La Vecchia C, Macfarlane GJ, Walker AM, Trichopoulos D. Epidemiology of pancreas cancer (1988). *Int J Pancreatol* 1989; 5:327-46. [PMID 2691590]
3. Tanaka T, Matsugu Y, Koide K, Miura Y, Ichiba Y, Dohi K, Ito H. Malignant lymphoma of the pancreas. *Dig Dis Sci* 1996; 41:402-4. [PMID 8601389]
4. Pecorari P, Gorji N, Melato M. Primary non-Hodgkin's lymphoma of the head of the pancreas: a case report and review of literature. *Oncol Rep* 1999; 6:1111-5. [PMID 10425311]
5. Salvatore JR, Cooper B, Shah I, Kummet T. Primary pancreatic lymphoma: a case report, literature review, and proposal for nomenclature. *Med Oncol* 2000; 17:237-47. [PMID 10962538]
6. Saif MW. Primary pancreatic lymphomas. *JOP. J Pancreas (Online)* 2006; 7:262-73. [PMID 16685107]
7. Dowson IM, Cornes JS, Morson BC. Primary malignant lymphoid tumours of the intestinal tract. Report of 37 cases with a study of factors influencing prognosis. *Br J Surg* 1961; 49:80-9. [PMID 13884035]

8. Behrns KE, Sarr MG, Strickler JG. Pancreatic lymphoma: is it a surgical disease? *Pancreas* 1994; 9:662-7. [PMID 7809023]
  9. Merkle EM, Bender GN, Brambs HJ. Imaging findings in pancreatic lymphoma: differential aspects. *AJR Am J Roentgenol* 2000; 174:671-5. [PMID 10701607]
  10. Masui T, Katayama M, Kobayashi S, Shimizu S. MR imaging of primary malignant lymphoma of the pancreas. *Radiat Med* 2005; 23:213-5. [PMID 15940070]
  11. Sata N, Endo K, Shimura K, Koizumi M, Nagai H. A new 3-D diagnosis strategy for duodenal malignant lesions using multidetector row CT, CT virtual duodenoscopy, duodenography, and 3-D multicholangiography. *Abdom Imaging* 2006 Jun 27; [Epub ahead of print]. [PMID 16802199]
  12. Smithers DW. Summary of papers delivered at the Conference on Staging in Hodgkin's Disease (Ann Arbor). *Cancer Res* 1971; 11:1869-70. [PMID 5121698]
  13. Shia J, Teruya-Feldstein J, Pan D, Hegde A, Klimstra DS, Chaganti RS, et al. Primary follicular lymphoma of the gastrointestinal tract: a clinical and pathologic study of 26 cases. *Am J Surg Pathol* 2002; 26:216-24. [PMID 11812943]
  14. Misdraji J, Fernandez del Castillo C, Ferry JA. Follicle center lymphoma of the ampulla of Vater presenting with jaundice: report of a case. *Am J Surg Pathol* 1997; 21:484-8. [PMID 9130997]
  15. Tuchek JM, De Jong SA, Pickleman J. Diagnosis, surgical intervention, and prognosis of primary pancreatic lymphoma. *Am Surg* 1993; 59:513-8. [PMID 8338282]
  16. Faulkner JE, Gaba CE, Powers JD, Yam LT. Diagnosis of primary pancreatic lymphoma by fine needle aspiration. *Acta Cytol* 1998; 42:834-6. [PMID 9622727]
  17. Civardi G, Vallisa D, Berte R, Giorgio A, Filice C, Caremani M, et al. Ultrasound-guided fine needle biopsy of the spleen: high clinical efficacy and low risk in a multicenter Italian study. *Am J Hematol* 2001; 67:93-9. [PMID 11343380]
  18. Buscarini L, Cavanna L. Ultrasound and ultrasonically guided biopsy in oncohematology. *Haematologica* 1991; 76:53-64. [PMID 2055561]
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