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Primary pancreatic lymphoma: a rare tumour that mimics pancreatic carcinoma. Clinical case

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Primary pancreatic lymphomas are extremely rare. Clinically, primary pancreatic lymphomas usually present with symptoms of pancreatic carcinoma. A localized and well-circumscribed tumour that replaces most of the pancreatic gland and compression of the blood vessels are radiological features of lymphoma, which are similar to pancreatic adenocarcinoma. Many patients are diagnosed with lymphoma after radical resection. It's a challenging clinical task for physicians, radiologists, and pathologists.

We report a case of primary pancreatic lymphoma that was confirmed by surgical resection. A 60-year-old woman came to the clinic with non-specific upper abdominal pain that lasted 8 weeks. Computed tomography (CT) scan showed a mass in the body of the pancreas, involving the superior mesenteric artery and the celiac trunk, and regional lymphadenopathy. Endoscopic ultrasound-guided fine needle aspiration of the pancreatic mass was performed. A morphological pattern indicated ductal carcinoma. The tumour board determined the treatment plan (chemotherapy) for the patient. The patient underwent 3 courses of GEMCAP chemotherapy in our hospital. A follow-up radiological exam showed no improvement. The chemotherapy regimen was changed to FOLFIRINOX. The patient underwent 6 courses of the FOLFIRINOX regimen.

A follow-up magnetic resonance imaging of the pancreas showed tumour regression by more than 90 % in comparison with the previous study. The patient underwent distal pancreatosplenectomy with standard lymph node dissection. Postoperative period was uncomplicated. These pathological results confirm the diagnosis of diffuse B-cell lymphoma.

Conclusions. This case shows that lack of tissue can make histological examination of FNA specimens challenging and mistakes can happen due to rare occurrence of primary pancreatic lymphomas.

Keywords
pancreatic lymphoma, pancreas, non-Hodgkin lymphoma.

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Primary pancreatic lymphoma is a rare tumour that accounts for less than 0.5 % of pancreatic tumours [1], and is frequently clinically misdiagnosed as pancreatic cancer. Overwhelming majority of patients are diagnosed with lymphoma only after invasive radical resection.

Out of the two main types of lymphoma (Hodgkin's and non-Hodgkin's lymphomas), non-Hodgkin's lymphomas more often invade extra lymphatic organs. Therefore, the most common histological type of pancreatic lymphoma is non-Hodgkin's lymphoma [7].

A localized and well-circumscribed tumour that replaces most of the pancreatic gland and compression of the blood vessels are radiological features of lymphoma, which are similar to pancreatic adenocarcinoma [3].

The prognosis of lymphoma is more favourable compared to adenocarcinoma (median overall survival: 53 months compared with less than 18 months) [2, 5].

EUS-FNA (Endoscopic Ultrasound-Guided Fine-Needle Aspiration) of the pancreatic lesion accompanied with advanced immunohistochemistry

are always perfect diagnostic tools that allow making a final diagnosis and avoiding unnecessary surgical intervention in the treatment of extranodal lymphomas. But in some cases, histological examination can be quite challenging [4, 6].

Case Presentation

A 60-year-old woman without any significant medical history presented with non-specific upper abdominal pain that lasted 8 weeks. She was examined in the local clinic and underwent abdominal computed tomography (CT). A mass in the body of the pancreas, 45 mm in a greater dimension, completely involving SMA (the superior mesenteric artery) and the celiac trunk, regional lymphadenopathy and small liver cysts were found on contrast-enhanced abdominal CT. No signs of dilation of the pancreatic duct were found (Fig.1). Endoscopic ultrasound-guided fine needle aspiration of the pancreatic mass was performed. Cytological report showed signs of connective tissue with the presence of several epithelioid cells with signs of cytologicalatypia. Immunohistochemical report showed negative reaction for total cytokeratin, positive reaction for IMP3, and negative reaction for CD56. A morphological pattern and the results of immunohistochemistry indicated ductal carcinoma (Fig.2).

There were no pathological findings on EGDS (esophagogastroduodenoscopy). The patient was referred to our hospital with the diagnosis of pancreatic adenocarcinoma $cT_4N_0M_0$. The carbohydrate antigen 19—9 and CEA levels were not elevated. Magnetic resonance imaging (MRI) confirmed non-metastatic origin of liver lesions and the presence of the mass in the body of the pancreas (Fig. 3).

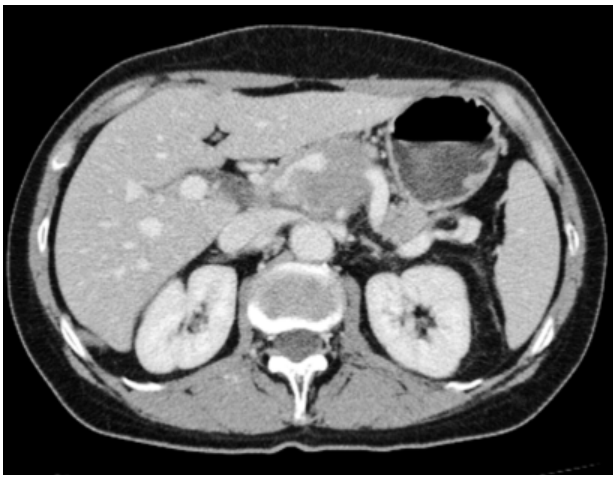

Figure 1. **CT (26/12/17). Abdominal CT findings: a 4.5-cm hypervascular mass with a rough border can be observed in the body of the pancreas. The mass semicirculary covers the celiac trunk, the superior mesenteric artery, common hepatic artery**

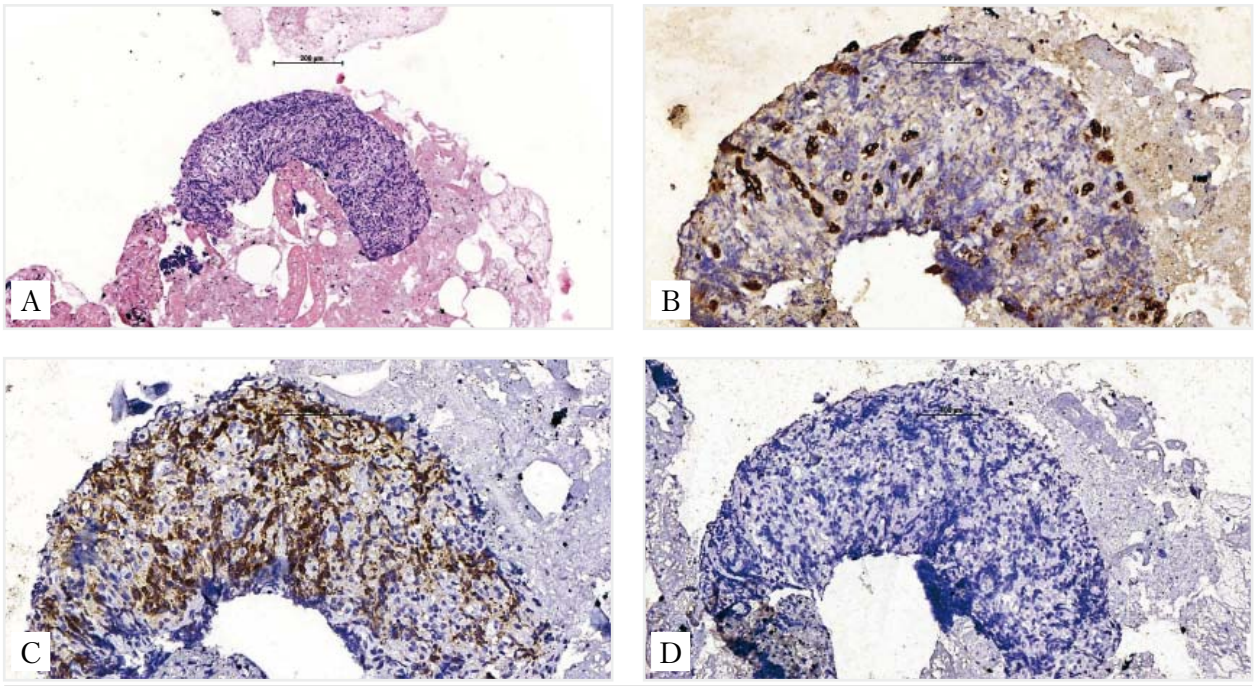

Figure 2. **Preoperative histology and immunohistochemistry:** A — FNA biopsy specimen. Some cells show a variable degree of cytological atypia. H&E (original magnification, × 70); B — pankeratin (clone CKAE1-AE3) highlights epithelial cells typical for ductal carcinoma. Differential with chronic pancreatitis is necessary (original magnification, × 150); C — IMP3 (which is positive in many malignancies) is positive in cells surrounding epithelial cells (original magnification, × 175); D — loss of expression of the CD56 which is normally positive in non-neoplastic ductal epithelium (original magnification, × 135)

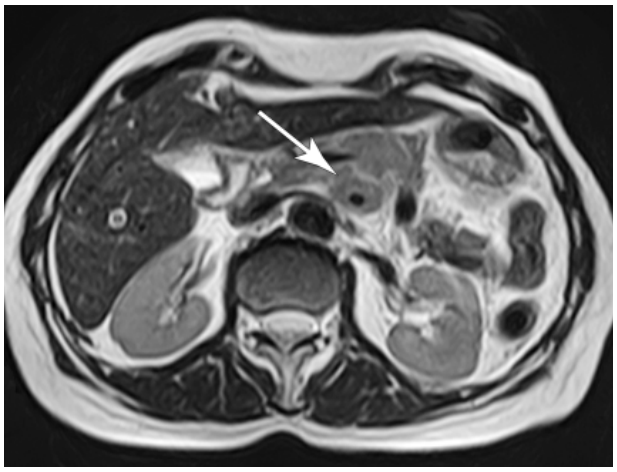

Figure 3. **MRI (20/01/18). MRI findings: The mass in the body of the pancreas that covers the celiac trunk and the common hepatic artery, liver cysts. After administration of a contrast, an increase of the MR signal in the arterial, portal, and venous phases was not visualized**

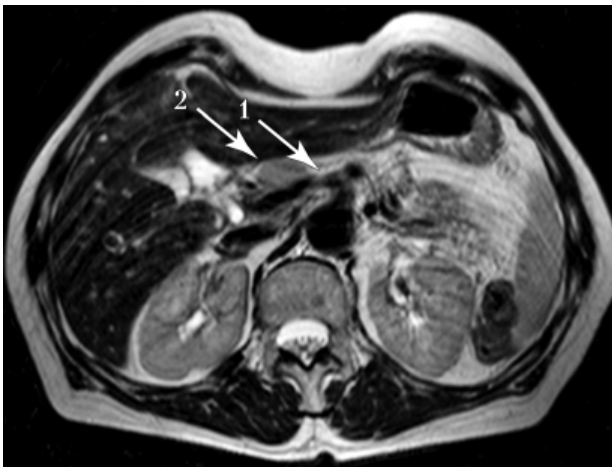

Figure 4. **MRI (31/08/18). MRI findings: In the isthmus of the pancreas, an area up to 2 mm enveloping the superior mesenteric artery (1) with a single lymph node of 12 groups (2) was determined**

The patient underwent 3 courses of GEMCAP chemotherapy in our hospital. A follow-up radiological exam showed no improvement. The chemotherapy regimen was changed to FOLFIRINOX. The patient underwent 6 courses of the FOLFIRINOX regimen.

Control

Magnetic resonance imaging of the pancreas determined tumour regression by more than 90 % in comparison with the previous study with a single lymph node of 12 groups (Fig. 4). The MRI diagnosis was adenocarcinoma of the pancreatic body $cT_1N_1M_0$.

According to the decision of the tumour board, the patient was offered an operative treatment.

Physical examination revealed good nutritional status (BMI (Body mass index) — 22.67 kg/m^2). Laboratory tests at admission demonstrated normal serum amylase 38 U/L, bilirubin 0.1471mg/dL, albumin 38 g/L. The carbohydrate antigen 19—9 level was 9.41U/mL, CEA levelwas 5.71 ng/mL.

The patient underwent distal pancreatosplenectomy with standard lymph node dissection. Postoperative period was uncomplicated. The patient was discharged home on the 9th postoperative day.

Pathological findings: a tumour tissue consists of solid layers of atypical lymphoid cells with a moderate amount of eosinophilic cytoplasm and atypical nuclei containing granular chromatin. There is a large number of tumour cells mitosis figures. Such tumour structure most closely corresponds to a large cell lymphoma (Fig. 5). Tumour cells are positive for CD20 and bcl-2, negative for CD3, CD5, SOX-11, tdt, CD30, c-myc. Approximately 80 % of tumour cells were positive for the Ki-67 proliferation marker. These results confirm the diagnosis of diffuse B-cell lymphoma. Moreover, tumour cells were positive for FoxP1 and bcl-6, and negative for CD10. According to the Visco-Young algorithm, this phenotype is common for a lymphoma originating from activated lymphocytes (ABC subtype) (Fig. 6). The diagnosis of diffuse large B-cell non-Hodgkin's lymphoma (ABC subtype), stage IIEA, was made.

Multidisciplinary tumour board recommended a dynamic observation.

4 months after the operation, the patient underwent PET-CT (22/01/2019). There were no signs of disease recurrence.

Primary pancreatic lymphoma is quite challenging for diagnostics. There are no specific clinical features of primary pancreatic lymphoma. They are similar to those that appear in pancreatic carcinoma.

Similar radiological findings do not facilitate the diagnostic process. In this clinical case, there was more radiological data for pancreatic carcinoma such as invasive growth of the tumour, involving surrounding blood vessels.

Ca19—9 is the most useful tumour marker in pancreatic carcinoma [8], but can be misleading as it may also be elevated in other malignancies, particularly of the upper gastrointestinal tract, including primary pancreatic lymphomas, especially when biliary obstruction is present [4], and decreased in pancreatic adenocarcinoma. Without definitive pathology diagnosis, potentially curable conditions such as primary pancreatic lymphoma as well as

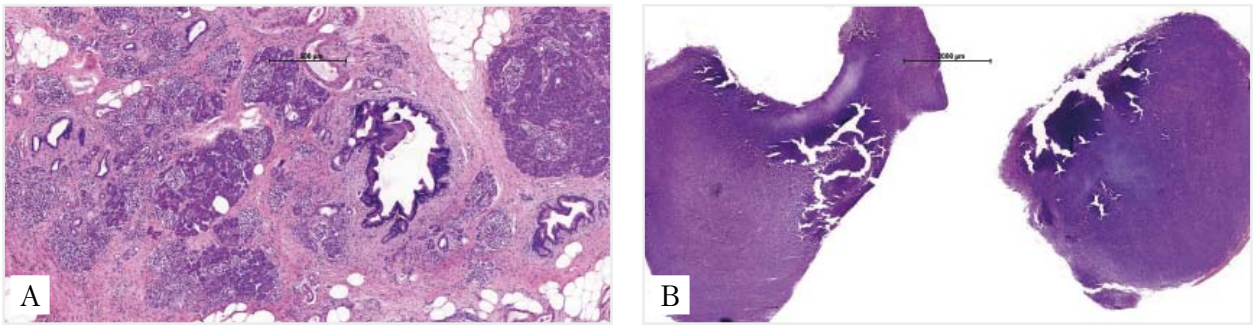

Figure 5. **Postoperative histology:** A — postoperative specimen shows no ductal adenocarcinoma H&E (original magnification, × 30); B — regional lymph nodes with lymphoma features H&E (original magnification, × 10)

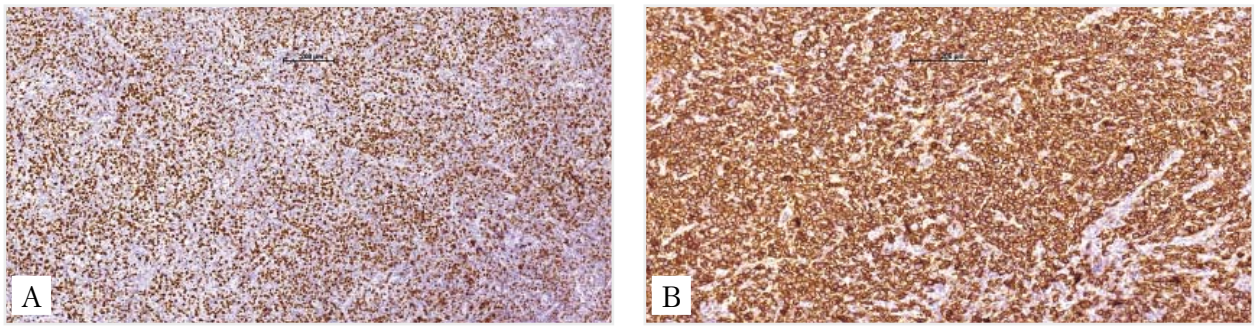

Figure 6. **Postoperative immunohistochemistry:** A — Ki-67 in lymphoma (original magnification, × 55); B — CD20 immunostain highlights B-cell lymphoma (original magnification, × 80)

other malignancies with more favourable prognosis may be misdiagnosed.

EUS-FNA of the pancreatic lesion with immunohistochemistry are great diagnostic tools when there is enough material for an advanced immunohistochemical panel [9]. This case shows that lack of tissue can lead to misinterpretation of the results as all necessary investigations can't be performed. The combination of factors — loss of expression of CD56 and strong expression of IPM3 — manifested ductal adenocarcinoma. Re-examination of the preoperative material after receiving the postoperative pathological report made it possible to suspect the presence of signs of lymphoma.

This case shows that histological examination of FNA specimens is challenging in case of lack of tissue and mistakes can happen.

Declaration of interests

The authors of this case report declare that they have no competing interests. The authors declare that they have no affiliations with or involvement in any organization or entity with any financial interest or nonfinancial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter discussed in the manuscript. The authors of this case report declare that no financial support nor grant support has been received for the preparation of the manuscript.

Ethics approval

The authors have no ethical conflicts to disclose.

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Первинна лімфома підшлункової залози: рідкісна пухлина, яка імітує карциному підшлункової залози. Клінічний випадок

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Первинні лімфоми підшлункової залози зустрічаються вкрай рідко. Клінічно первинні лімфоми підшлункової залози зазвичай репрезентуються симптомами раку підшлункової залози. Рентгенологічні ознаки лімфоми, такі як локалізована, з чіткими краями пухлина, яка заміщує більшу частину підшлункової залози, компресія судин, подібні до аденокарциноми підшлункової залози. У багатьох пацієнтів з первинними панкреатичними лімфомами встановлюють діагноз після радикальної резекції. Це складне клінічне завдання як для лікарів і рентгенологів, так і для патогістологів.

Ми презентуємо випадок первинної лімфоми підшлункової залози, що виникла в тілі підшлункової залози, що було підтверджено після хірургічної резекції. Жінка 60 років звернулася в клініку з 8-тижневим неспецифічним болем у верхній частині живота. Комп'ютерна томографія підтвердила наявність утворення в тілі підшлункової залози, що залучає верхню брижову артерію та черевний стовбур з регіонарною лімфаденопатією. Виконано ендоскопічну аспіраційну біопсію пухлини тіла підшлункової залози під ультразвуковим контролем. Морфологічна картина була на користь протокової карциноми. Мультидисциплінарна комісія рекомендувала хворій пройти хіміотерапію. У нашій лікарні пацієнтці було проведено 3 курси хіміотерапії GEMCAP з негативною рентгенологічною динамікою. Схема хіміотерапії була змінена на FOLFIRINOX. Пацієнтка пройшла 6 курсів хіміотерапії за схемою FOLFIRINOX.

Контрольна магнітно-резонансна томографія підшлункової залози показала регрес пухлини більш ніж на 90 % порівняно з попереднім дослідженням. Пацієнтці було виконано дистальну панкреатоспленектомію зі стандартною лімфодисекцією. Післяопераційний період протікав неускладнено. Післяопераційні патогістологічні результати підтвердили діагноз дифузної В-клітинної лімфоми.

Висновки. Цей випадок показує, що гістологічне дослідження зразка, одержаного за допомогою тонкогольної аспіраційної біопсії, є складним у разі нестачі матеріалу і можуть статися помилки у діагностиці та лікуванні через рідкість первинних лімфом підшлункової залози.

Ключові слова: лімфома підшлункової залози, підшлункова залоза, неходжкінська лімфома.

FOR CITATION

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