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Case Report

Imaging Features of Pancreatic Dedifferentiated Liposarcoma: A Rare Case Report

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Abstract

Dedifferentiated lip sarcoma (DDLPS) is a rare subtype of lip sarcoma with poor prognosis. This current case report described a primary pancreatic DDLPS in a 51-year-old Chinese female suffering from intermittent pain in her middle and upper abdomen for one year after three years of radical gastrectomy. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) were undertaken to identify and evaluate the tumour details. The patient underwent resection of the pancreatic tumour. The spindle cell proliferation was found by pathologic HE staining. Further immunohistochemistry and genetic testing were recommended and detected the positive for minute 2 (MDM2), cyclin-dependent kinase 4 (CDK4) and signal transducer and activator of transcription 6 (STAT6) in this specimen. The case report also reviewed the literature in terms of the radiologic and pathological characteristics of DDLPS for the aim of improving the preoperative diagnosis and accurate location of tumour.

Keywords: Case Report; Computed Tomography (CT); Dedifferentiated Lip sarcoma; Magnetic Resonance Imaging (MRI); Pancreas

Introduction

Dedifferentiated lip sarcoma (DDLPS) is a rare neoplasm that exhibits various morphologies. It is a distinct subtype of lip sarcoma that defines a morphological transition from well-differentiated lip sarcoma (WDL) or atypical lipomatous tumour to high-grade sarcoma [1]. The tumour is characterized of mouse double minute 2 (MDM2) amplification by fluorescence in situ hybridization (FISH). Surgical resection is the most effective treatment for DDLPS, while radiotherapy and chemotherapy have ineffective curative effects. The earlier the diagnosis, the better the surgical effect. More effective and non-invasive diagnosis of DDLPS is needed to improve the preoperative diagnosis and accuracy location. With the development of medical imaging technology, especially the application of clinic Computed Tomography (CT) and Magnetic Resonance Imaging (MRI). Both of them play key roles to diagnose DDLPS with the typical characterization, which displayed a soft tissue mass with varied composition and closely related to the proportion and distribution of different differentiation components [2]. However, the imaging features related to pancreatic lip sarcoma were few reported previously [3]. The primary DDLPS is an extremely rare in the pancreas [3-5]. Herein, we report an unusual case of DDLPS arising from the tail of pancreas with relevant imaging features of both CT and MRI, as well as pathological evidences in order to improve the diagnosis and location of DDLPS before surgery.

Case Presentation

A 51-year-old Chinese female presented to the department of Hepatobiliary Surgery, the People's Hospital of Longhua, Shenzhen, Guangdong province, China in October 2020 due to her intermittent pain in the middle and upper abdomen for one year. The patient underwent radical gastrectomy for gastric cancer three years earlier. Before the hospitalization, she had undergone abdominal CT and MRI scans in the same hospital above. An abdominal unenhanced CT identified a round soft tissue mass measuring 5.7 X 6.7 X 6.8 cm3 in the tail of pancreas. The CT value of mass was 51 Hu in unenhanced CT images [Figure 1A], but the CT values were increased to 77 Hu, 86 Hu, and 109 Hu, respectively post-contrast at arterial [Figure 1B], venous [Figure 1C] and delayed periods [Figure 1D]. The mass was closely related to the tail of pancreas and the pancreatic tail was compressed, but the boundary of adjacent surface was not clear partly showing as yellow arrow in Figure 1A-1D. The representation of mass was relatively homogeneous, but detected a minimum CT value at -80 Hu in low-attenuation areas, which might indicate very less fat components in coronal position [Figure 1E]. The adjacent splenic artery and vein were compressed and displaced in the volume rendering (VR) imaging [Figure 1F]. The shadow of surgical clipping for radical gastrectomy was observed as long strip high density in the gastric body from multi-planar reformatting (MPR) in sagittal position [Figure 1G]. No abnormal enhanced lesions were found in CT images. A tumour of the tail of pancreas, likely neuroendocrine tumour, was considered by CT impression.

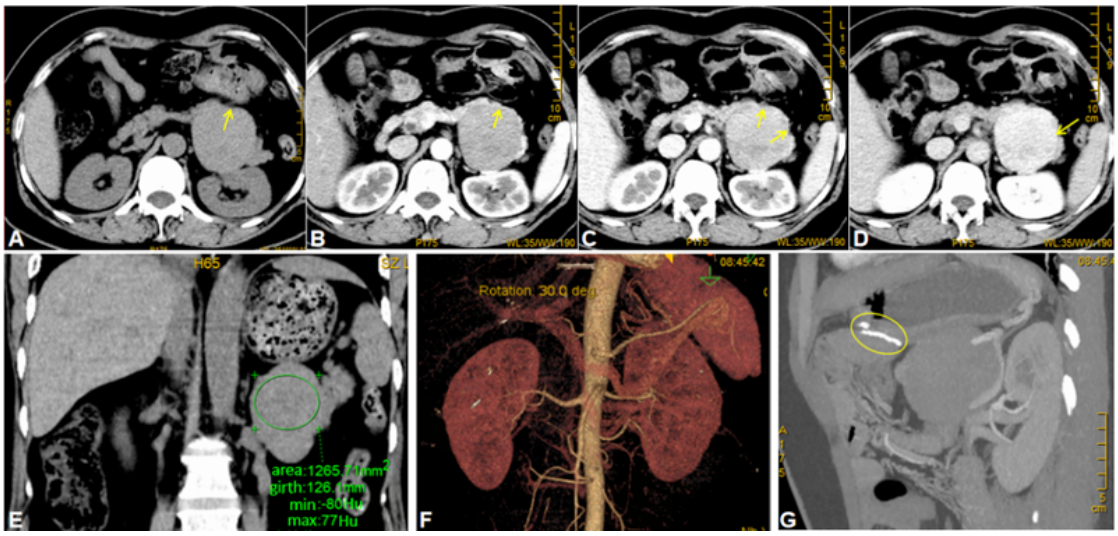

Figure 1: An abdominal CT showing a round soft tissue mass in the tail of pancreas. The representation of mass was relatively homogeneous in non-enhanced image (A), but becoming to in the arterial, venous and delayed periods in (B-D). A minimum CT value at -80 Hu in low-attenuation areas was detected in coronal position (E). The adjacent splenic artery and vein were displaced in the volume rendering (VR) imaging (F). The shadow of surgical clipping for radical gastrectomy was observed in multi-planar reformatting (MPR) in sagittal position (G). The yellow arrows indicate the unclear boundary of adjacent surface partly between the tumour and tail of pancreas.

Thereafter, MRI was implemented and demonstrated a mass in the body and tail of pancreas. Its size was approximately 5.8 X 6.7 X 6.8 cm3 with heterogeneous appearance. The most areas of the mass were hypo intensity in T2 weighted imaging (T2WI) and T2WI with fat suppression sequence, rather than hyper intensity in diffusion weighted imaging (DWI). The boundary of lesion was relatively clear as shown in Figure 2A-2C. MR enhanced images revealed the tumour in T1 weighted imaging (T1WI) from homogeneous iso-intensity in pre-contrast [Figure 2D] to heterogeneous enhancement in the arteriovenous period [Figure 2E-F] and stronger enhancement in the delay period [Figure G]. The tail of pancreas was compressed and shifted forward, while, the arteriovenous of spleen was moved backward [Figure 2H]. However, it was found that the adjacent surface between tumour and the pancreatic pancreas was not clear slightly showing as yellow arrow in Figure 2D-G. It might indicate that the tumour was originally from the tail of pancreas. There was no obvious dilation of the pancreatic duct observed in magnetic resonance cholangiopancreatography (MRCP) showing in the Figure 2I. The characteristic of metastatic lymph nodes or lesion was not found in all MR images. In addition, no abnormal enhancement was observed in the residual stomach. Therefore, MRI impressed the tumour as a ligamentous fibroma originating in the tail of pancreas.

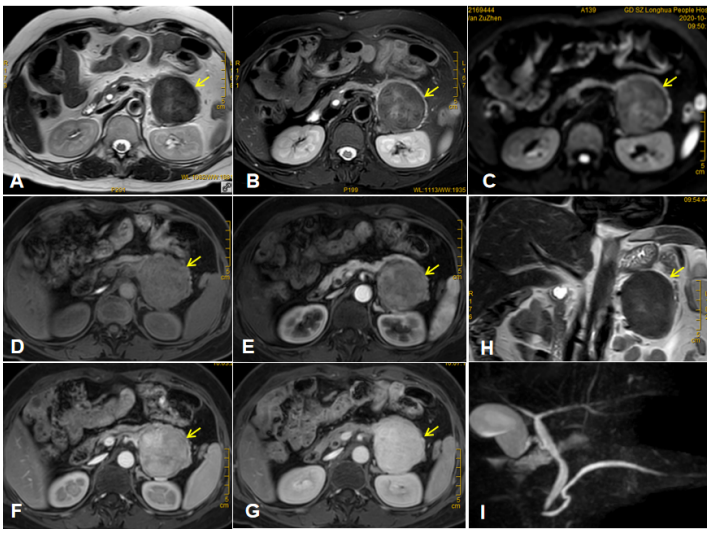

Figure 2: MR images presented the tumour in the pancreatic tail (arrow). The tumour showed the heterogeneous hypo intensity in the T2WI (A) and fat suppression T2WI (B), rather than hyper intensity in DWI (C). T1WI revealed the tumour with relative iso-intensity in precontrast (D) and increased enhancements at arteria (E), portal (F), and balance (G) period in the axial position. More details, the adjacent surface between tumour and the pancreatic pancreas was not clear slightly (arrow in D-G). The tumour in coronal position was showed in the T2-weighted with fat suppression sequence (H). The normal MRCP was presented in (I). The yellow arrows showed the location of the tumour.

In retrospective observation, a few high signal area mixed within the tumour, which showed a slight hyper-intensity in multi-Dixon T1WI [Figure 3A], but the signal intensity was suppressed to hypo-intensity in fat suppression of multi-Dixon sequence as showing in Figure 3B.

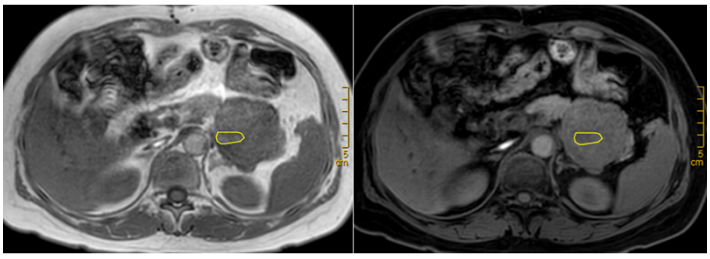

Figure 3: A slight hyper-intensity area (yellow circle) mixed within the tumour in multi-Dixon T1WI, but the signal intensity was suppressed to hypo-intensity in fat suppression of multi-Dixon sequence.

Laparoscopic surgery for complex intestinal adhesion release, pancreatic body & tail excision, abdominal cavity irrigation and drainage was performed considering the impression of CT and MRI examinations. After the tumour was completely excised, the specimen was sent for rapid pathological examination. The pathologic HE staining showed spindle cells proliferation in the tumour specimen, which supported the considering of an isolated fibroma of pancreas [Figure 4A]. Additionally, it wan found that the mature adipocytes were mixed with spindle cells in the local tumour tissue [Figure 4B]. The mild atypia, occasional mitosis, and significant interstitial collagenisation were also found in tumour specimen. However, it was finally identified as a DDLPS because a WDL component at the tumour margin by gene amplification test was detected with an over- expression of MDM2 [Figure 4C], cyclin-dependent kinase 4 (CDK4) and signal transducer and activator of transcription 6 (STAT6), while the Ki67 (nucleus related antigen) was relatively low at 3%.

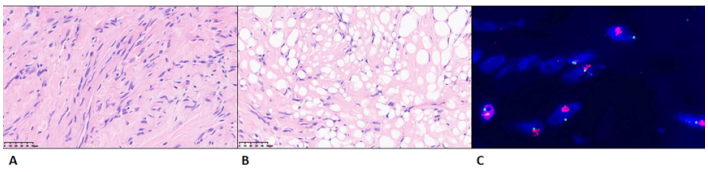

Figure 4: Representative photomicrograph images of tumour specimens. The pathologic HE staining showed spindle cells proliferation (A) and mixed fat cells (B). The immunohistochemistry indicated positive for MDM2 (C).

The patient in this case report provided verbal informed consent for publication of this report. The reporting of this case conforms to CARE guidelines [6].

Discussion and Implications

Dedifferentiation can occur in any area of the body [7]. However, DDLPS arises mostly in the retro peritoneum and deep soft tissue of proximal extremities, while other anatomical areas occasionally reported, such as the small bowel mesentery, the colon, the sigmoid mesocolon and rectum [8-10]. In this current case, the tumour with DDLPS was located in the tail of pancreas. The pancreas is an rare site of primary lip sarcoma [2,3,11,12]. In the present case, an extremely rare pancreatic DDLPS appeared during follow-up of radical gastrectomy without any signs of recurrence or metastasis in clinic. In terms of symptoms, the DDLPS usually presented abdominal pain or the syndromes generated from pressure on the surrounding tissues [8]. However, DDLPS in the tail of pancreas may grow for a long period without causing any symptoms. In this current case, due to the impact on the tail of pancreas, the patient had experienced intermittent pain in the middle and upper abdomen for one year. Such symptom is quite common and unspecific. As a result, several diagnostic methods are needed to identify the tumour. A variety of clinical examinations, such as CT and MRI, should be undertaken to diagnose DDLPS. The histology of DDLPS is typically characterized as the conversion of atypical lipomatous tumour and WDL with non-fatty sarcoma components [8]. Under a microscope, the two components are generally clearly separated, with a sharp transition between the two. Immunohistochemistry was positive for harbour MDM2 amplification via fluorescence in situ hybridization (FISH), which can be used to confirm the diagnosis of DDLPS [8,13]. In this case, report, fatty sarcoma components were not found in specimen histopathological, while MDM2 was positive in genetic amplification testing. In addition to histopathology, CT and MRI can differentiate fat from other soft tissue components with higher precision [14]. CT imaging is helpful for the diagnosis of LPS in the location and components of tumour, while MRI gives more accurate information about the heterogeneity and position of the tumour, but it cannot completely identify the type of the tumour. The details about imaging features of DDLPS were not reported in the previous publications. This current case had manifestations with heterogeneous, soft tissue and hypo-intenty CT and T2WI appearance, which consisted with other report [15]. DDLPS or LPS should be suspected when the tumour has a heterogeneous structure and a relatively low blood. In this case report, the tumour with a very few fat tissue, but fatty sarcoma imaging features were hard to be found in both CT and MR images, which supported with its typical characterization of non-fatty sarcoma components histopathological. Imaging examinations can be used to determine the extent of the tumour and the infiltration of surrounding organs, but the final diagnosis depends on pathological examination as of now [14,15]. The tumour was quite complex and lack of specificity in this current case. The preoperative MRI findings suggested that tumour was closely related to the tail of pancreas with the compressed pancreatic tail and indistinct of the adjacent surface between tumour and the tail of pancreas partly. These findings might indicate the tumour was originally from the pancreas, the tail of pancreas. In conclusion, this current case report revealed a unique case of the DDLPS originating from the tail of pancreas, one of the rarest locations for digestive sarcomas. DDLPS should be thoroughly distinguished from its morphological mimickers as the tumour may be more highly aggressive and more extensive than clinically and radiologically expected. The pancreatic tumour was confirmed finally as DDLPS by MDM2 proliferation. However, the radiologists should keep close to DDLPS in the location, size, homogeneity, component, and enhancement in order to improve the preoperative diagnosis and curative effect.

Patient consent: Obtained.

Conflicts of interest: The authors declare that there are no conflicts of interest regarding the publication of this article.

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