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Primary pleomorphic liposarcoma of the pancreas: Our first encounter

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Abstract

Liposarcomas are malignant soft tissue tumours of adipocyte origin with or without lipoblasts, typically found in the limbs or retroperitoneum. Like most sarcomas, liposarcomas are rare with even fewer instances of visceral occurrences reported. The pancreas is an exceptionally rare site of primary liposarcoma, accounting for less than 0.1% of all pancreatic malignancies with only 8 cases reported thus far since 1979. We present a patient with primary pleomorphic liposarcoma of the pancreas.

A 49 year-old lady presented with obstructive jaundice and was found to have a pancreatic mass on imaging. A Whipple's procedure was performed and revealed a bulky pancreatic head mass. A large 92 mm tumour was seen within the pancreatic head and microscopic examination revealed a rare primary pleomorphic liposarcoma of the pancreas.She developed complications post-surgery but recovered over time. She did not receive any adjuvant therapies after consultation and discussion with oncologist and remains under close follow-up.

Pleomorphic liposarcomas of the pancreas are rare entities and this case report illustrates imaging work-up, surgery, possible complications, management and histopathology examination of the tumour.

Keywords

pancreatic tumour; liposarcoma; pleomorphic liposarcoma; Whipple's procedure; chyle leak.

Abbreviations

STS: Soft tissue sarcoma; MDM2: Mouse double minute 2 homolog; H&E stain: Haematoxylin and Eosin stain; PLS: Pleomorphic liposarcoma.

Introduction

Soft tissue sarcomas (STS) are rare malignancies of mesenchymal origin with an incidence of 2.4/10 million to 5/10 million, accounting for approximately 1% of adult malignancies and 15% of children [3]. With distinct heterogeneity, STS frequently occurs in adolescents and middle-aged adults [3].

The 5-year survival rate is 40-60%, varies with prognostic factors such as age, tumour location, size, histological grade, and tumour metastasis. As visceral soft tissue sarcomas mostly occur in a concealed position and progresses slowly without any symptoms, it causes delayed detection of the disease until the tumour compresses the adjacent tissue and causes clinical symptoms [4].

Mesenchymal tumours of the pancreas account for less than 1% of pancreatic tumours and liposarcomas are an even more rare entity [1].

Surgical resection of the primary tumour remains the main stay of treatment. Unfortunately, considerable numbers of patients develop local recurrences or distant metastases. Effective systemic therapies for this broad range of rare disease have been lacking, consequently leading to poor prognosis for patients with advanced disease, especially for dedifferentiated liposarcomas.

Case Report

On the 16th of April 2020, the Sultanah Bahiyah Hospital in Kedah, Malaysia, a government tertiary specialist hospital, received a referral from a hospital in a neighbouring state. Our patient is a 49-year-old lady with underlying diabetes mellitus, hypertension, dyslipidaemia and plaque psoriasis referred for symptoms of obstructive jaundice; yellowish discoloration of her skin, pruritus, passing tea-coloured urine with pale stools, with loss of appetite and unintentional weight loss of approximately 5 to 6 kg, over a course of 1 to 2 months. She claimed to have had no known family history of cancer. Upon initial examination, she was noted to be clinically jaundiced with a palpable mass over the right hypochondrium of her abdomen.

Blood investigations were in keeping with obstructive jaundice while her CA 19.9 tumour marker was raised at 471.04 IU/mL. The abdominal ultrasound done revealed features of obstructive jaundice secondary to a hypoechoic lesion at the head of the pancreas, measuring approximately 8.1 cm X 6.8 cm X 6.8 cm, with increased vascularity.

This was subsequently followed-up by a CT Pancreatic Protocol and Thorax which further supported the diagnosis of a borderline resectable pancreatic head malignancy causing biliary tree obstruction. No evidence of distant metastasis was noted (Figure 1 and 2).

A Whipple's procedure (pancreaticoduodenectomy) was performed on the 19th of April 2020. During the operation, there was a bulky head of pancreas tumour noted, extending to the uncinate process and abutting the superior mesenteric artery, involving the first 2 jejunal branches of the SMV. A Sugiyama intestinal derotation procedure was used in combination with an artery first approach to resect the tumour and complete the Whipple's procedure. Multiple portal, common hepatic and aortocaval nodes were noted and dissected. An on-table frozen section of the pancreatic resection margin was sent which yielded no tumour. To prevent a potentially difficult appendicectomy in the future, an appendicectomy was performed as well.

Five days following the operation, pus discharge was noted at the periumbilical aspect of the wound with erythema of the surrounding skin and tenderness over the area of erythema. A CT-scan was done which found evidence of an abdominal wall abscess, for which 2 peritoneal washouts were done on the 25th of April and 8th of May respectively. During the first reoperation, chyle leak was noted adjacent to the hepaticojejunostomy site, it was addressed by application of ligaclips. Other anastomotic sites were intact. A follow-up CT 2 week later subsequently showed a resolving collection. Unfortunately, the chyle leak still persisted from the wound which required a second washout which found slough covering the whole abdominal and small bowel, mainly at the subhepatic region covering the area of the pancreaticojejunostomy and gastrojejunostomy anastomoses – the anastomotic sites could not be assessed due overlying dense adhesions. Cultures taken from the wound yielded ESBL Klebsiella Pneumoniae which required the escalation of anitibiotics of IV Tazocin (Piperacillin/Tazobactam) and clinical input from the infectious disease team.

Shortly after the resumption of her normal diet, she developed feeding intolerance. As a result of this, throughout the remainder of her hospital stay, her diet was tuned and adjusted with the aid of the resident dietician to have a low fat content due to the evidence of chyle leak while meeting her caloric, protein and other nutritional requirements. Despite this, she still suffered from multiple bouts of loose stools daily despite multiple dietary alterations and the addition of pancrelipase (Pancretin), ultimately requiring parenteral nutrition support.

The prolonged hospital stays, multiple operations, feeding intolerance and separation from family (local Movement Control Order during Covid – 19 pandemic) ultimately resulted in patient developing adjustment disorder with depressive mood, for which she was referred to the psychiatry team and started on antidepressants.

On Day 38 of hospital stay, the patient was transferred back to her referring hospital for continuation of care upon her strong request despite poorly established feeding, reliance on parenteral nutrition and persistent chyle leak from wound.

Her follow up care was encouraging once she returned closer to home and family support. Her wound healed gradually leaving only a small discharging sinus of low volume chyle. Her TPN was weaned off over a period of 2 weeks and full enteral diet resumed. A multidisciplinary team of physicians, surgeons and therapists worked to get her fit and optimised. She was discharged home and remains on close follow-up. She has opted not to go for adjuvant radiotherapy nor chemotherapy but remains on follow-up with the surgical and oncology teams.

Histopathology

On gross examination, a lobulated well-circumscribed tumour with solid greenish-yellowish and beige myxoid/gelatinous surface was seen within the pancreatic head measuring 92 X 76 X 62 mm (Figure 3). On histology, the tumour is seen arising from within the pancreatic parenchyma (Figure 6). It displays a heterogenous histology composed of lipoblast and non-lipogenic areas with neoplastic spindled to pleomorphic cells in a predominantly loose myxoid background with incomplete fibrous septa. The spindled cells are arranged in short fascicles with intervening curvilinear thin-walled blood vessels suggestive of an

intermediate to high grade myxofibrosarcoma component (Figure 4). Univacuolated and multivacuolated lipoblasts, some appearing pleomorphic are identified within non-lipogenic areas (Figure 5). Other areas with undifferentiated pleomorphic sarcoma appearance are also seen. A histological interpretation of pleomorphic liposarcoma (PLS) was made.

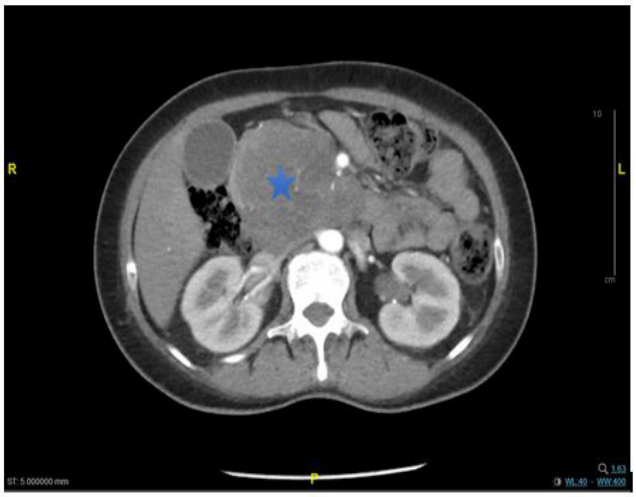

Figure 1: CT scan in the arterial phase showing a pancreatic tumour(star) abutment with the superior mesenteric artery.

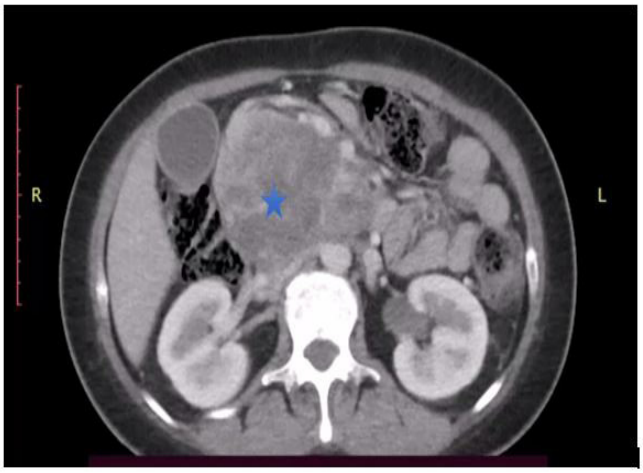

Figure 2: CT scan in the venous phase showing a pancreatic tumour (star).

Figure 3: A well-circumscribed tumour is seen with a rim of residual pancreatic tissue (arrows).

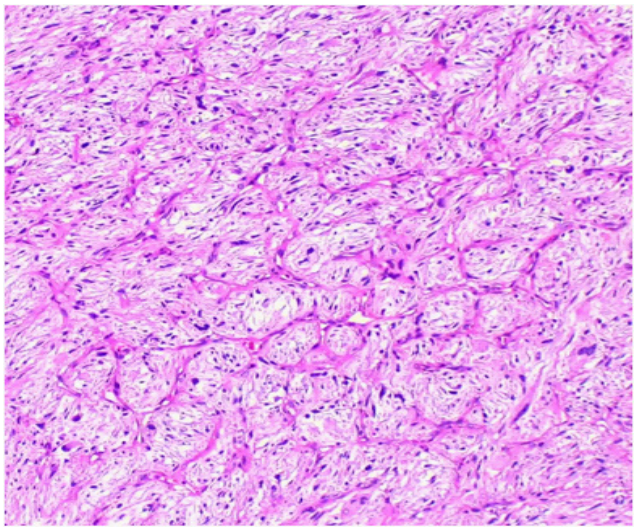

Figure 4: Fibrosarcomatous areas with curvilinear vessels. H&E stain. X40 magnification.

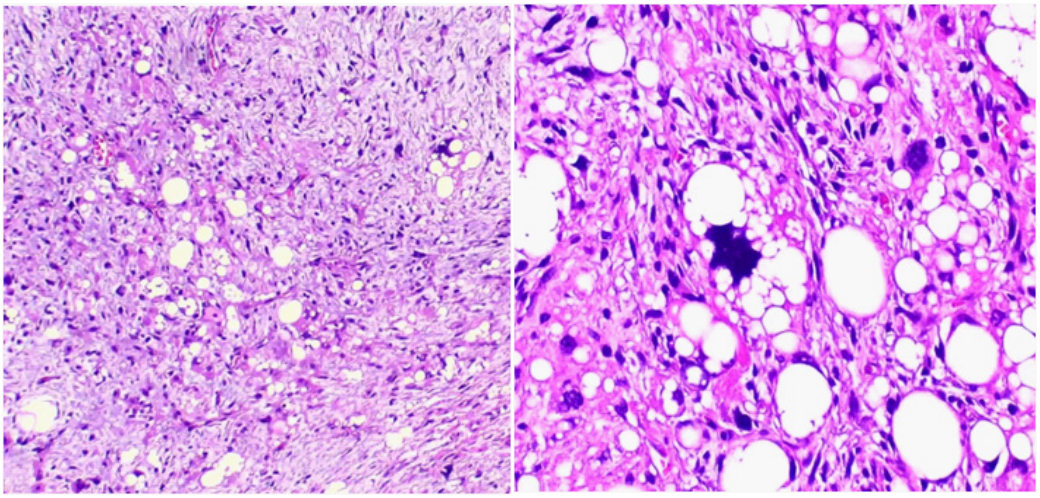

Figure 5: A) Uni and multivacuolated lipoblast intermingled within non-lipogenic areas. H&E stain. X40 magnification. B) Pleomorphic lipoblast with surrounding univacuolated and multivacuolated lipoblast. H&E stain. X400 magnification.

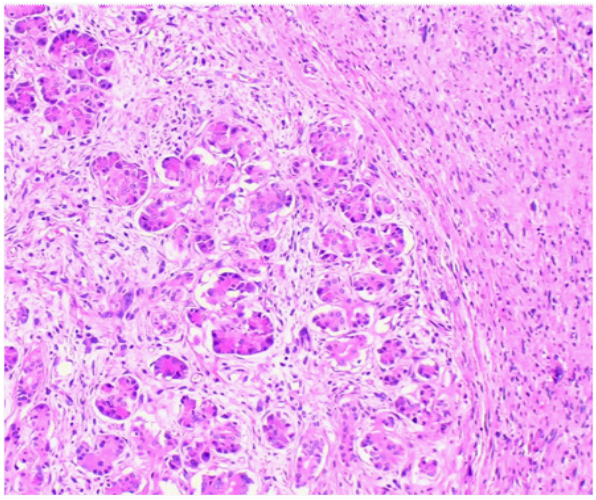

Figure 6: Entrapped pancreatic acini within the tumour. H&E stain. X200 magnification.

Discussion

Liposarcoma is a tumour of adipose differentiation. It is the most common soft tissue sarcoma, accounts for 15-20% of all mesenchymal malignancies [7]. They are divided into atypical lipomatous tumour/well-differentiated liposarcoma, myxoid/round cell liposarcoma, pleomorphic liposarcoma (PLS) and dedifferentiated liposarcoma. Deep soft tissues of the extremities are the most common sites of involvement with only rare cases of visceral location reported [7]. Both well-differentiated and myxoid cell types are low-grade tumours with relatively low risk of metastasis and more favourable prognosis [7]. However, the dedifferentiated and pleomorphic liposarcoma are high-grade tumours [7]. Dedifferentiation occurs in upto 10% of well-differentiated liposarcoma. Retroperitoneum is the most common location, followed by spermatic cords, head and neck and trunk. Occurrence in subcutaneous tissue is rare. The histological hallmark is an abrupt transition from well-differentiated liposarcoma to non-lipogenic sarcoma which is usually high-grade. It exhibits variably morphology, resembling undifferentiated pleomorphic sarcoma or intermediate- to high-grade myxofibrosarcoma. In addition, it may exhibit heterologous differentiation of myogenic or osteo-/chondrosarcomatous element. This type of tumour typically shows diffuse nuclear

expression of MDM2 (Mouse double minute 2 homolog), which is helpful to exclude pleomorphic liposarcoma. Dedifferentiated liposarcoma has a local recurrence rate of at least 40%. Distant metastases are seen in 15-20% of cases with overall mortality of 28-30% at 5-year follow-up. However, it appears to be less aggressive as in comparison with other types of high-grade pleomorphic sarcoma. Anatomical location is the most important prognostic factor with the retroperitoneal tumours associated with worst clinical behaviour.

PLS is very rare, contributing to less than 5% of all liposarcomas with a higher (30-50%) frequency of distant metastasis, local recurrence and poor prognosis [8,9]. The abdomen as a primary site contributes to about 12% of all PLS [9] and only one other case of pancreatic origin has been reported [10].Ghadmini et al found that the PLS in their study ranged between 0.7 cm to 30 cm [9].

PLS usually occurs in elderly with slightly higher incidence in males. It is commonly found in lower extremities while trunk wall, retroperitoneum is rarely affected. Macroscopically, the tumour is large, well-demarcated but non-encapsulated. They can be ill-defined and infiltrative. Cut section shows white to yellowish surface as well as myxoid change and foci of necrosis. Histologically, pleomorphic lipoblasts are seen in the background of high-grade sarcoma. Non-lipogenic areas resembles undifferentiated pleomorphic sarcoma with spindle and multinucleated giant cells. Lipoblasts is necessary for the diagnosis. In areas, myxofibrosarcoma of intermediate to high-grade can be present. In the immunohistochemical study, S100 highlights the lipoblasts. SMA and CD34 are at least focally positive.

Pancytokeratin, EMA and desmin immunostains can be positive [7]. Staining for MDM2 is typically negative, in contrast with dedifferentiation type.

Clinical presentations are not unlike other pancreatic tumours, and presenting symptoms may be vague such as nonspecific epigastric pain, nausea, and vomiting. Some patients may present with constitutional symptoms with weight loss and an abdominal mass. If the tumour is located at the head, uncinate or even the neck of the pancreas, jaundice is probable.

In this case, this patient is a middle-age lady presented with obstructive jaundice with constitutional symptoms. CT scan findings show head of pancreas tumour causing biliary tree obstruction. On gross examinations, a large tumour is found arising within the head of pancreas showing myxoid greenish gelatinous surface. Histologically, the tumour is focally infiltrative and rimmed by pancreatic tissue. There are areas resembling intermediate to high grade myxofibrosarcoma. The spindle cells are arranged in fascicles with intervening curvilinear walled blood vessels. In non-lipogenic areas, the tumours resembles undifferentiated pleomorphic sarcoma composed of cells with markedly pleomorphic vesicular nuclei and prominent nucleoli. Univacuolated and multivacuolated lipoblasts, some appearing pleomorphic are identified with S100 positivity, further supporting the diagnosis of pleomorphic liposarcoma. There are no abrupt transition from well-differentiated liposarcoma to non-lipogenic sarcoma areas to suggest dedifferentiated component. Histology was the mainstay of diagnosis in this case as MDM2 was not readily available.

Factors that influence the prognosis are degree of differentiation and adequacy of surgical resection. Five-year survival following complete surgical excision had been reported as 41-50%. Surgical resection, radiation and close follow up were the current recommended treatment. In the pre-existing literature re-

view, 2 cases were reported as well differentiated liposarcoma, 3 cases of dedifferentiated with high grade components, 1 case of myxoid and 1 of pleomorphic liposarcoma [2]. Most of the cases were diagnosed through surgery combined with microscopic examination. One of the cases in the literature was diagnosed by positive amplification of MDM2 [5].

At the moment, surgical resection is the only potentially curative management for resectable primary pancreatic liposarcomas. In cases of incomplete resection, radiotherapy has been shown to potentially increase the duration of remission [6]. However, due to the scarcity of case series and randomised trials there are no established chemotherapy regimens currently employed.

In our case, CT imaging revealed a pancreatic head mass resulting in the obstruction and dilatation of the patient's biliary tree. The tumour was subsequently resected via Whipple's procedure. Due to the extensive dissection embarked during surgery, this patient had an unfortunate development of chyle ascites with super imposed infection that complicated her post-operative period. She had a prolonged hospital stay and rehabilitation. The combination of modified enteral feeds with Pancreatic enzyme supplementation and parenteral nutrition was key to her recovery. Although she endured two further laparotomies for infected chyle ascites resulting in organ/space SSI, she remained in good spirits and this enabled us to complete her recovery process. She remains under close follow-up for recurrences as she did not receive any adjuvant therapies after consultation and discussion with our oncology colleagues.

Conclusion

Pleomorphic liposarcomas of the pancreas are rare entities and this case report illustrates imaging work-up, surgery, possible complications, management and histopathology examination of the tumour.

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