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#### RESEARCH ARTICLE

# UNDIFFERENTIATED PLEOMORPHIC SARCOMA OF THE PANCREAS WITH METACHRONIOUS LIVER METASTASIS, A RARE CASE REPORT

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# Abstract

Undifferentiated Pleomorphic Sarcoma(UPS) is an exceptional histopathological entity of pancreatic tumors, Pancreatic sarcomas tend to be more aggressive and have a poor prognosis(1). We present the case of a 65-year-old man who was treated with a pancreatic head tumor with metachronious liver metastasis for which he was operated in two times, he was first operated for his pancreatic head tumor a UPS of pancreatic head witch consisted on a cephalic duodenopancrtoctomy then he received adjuvant radiochemotherapy, the patient developed a metachron liver metastasis 1 year after first surgery with 2 hepatic lesions, the management of his liver metastasis after DRCs for cancer multidisciplinary consultation meetings is a Right lobectomy with metatsasectomy of the VI segment lesion.this case repport and literature revews take us to conclude about the role of multidisciplinary treatment in the long-term prognosis of primary UPS of the pancreas, we believe that multidisciplinary treatment could improve the survival rates of other patients.

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# **Introduction:-**

Undifferentiated Pleomorphic Sarcoma (UPS) or previously known as malignant fibrous histiocytoma, is the most prevalent type of soft tissue sarcoma in adults(HASSAN ET AL, 2020), However, it has been rarely observed in the digestive system (). Pancreatic sarcoma represents less than 1% of all pancreatic tumors, and primary UPS of the pancreas is even rarer (3).

The first case of pancreatic UPS was described in 1976 by Margules (4), only 21 cases have been documented in the literature(2). One case recurrence was reported 11 months after surgery with hepatic and pulmonary metastasis(5).

We report a case of pancreatic UPS with liver metastasis.

#### **Case Report:**

The patient was a 65-year-old man. He had no significant past medical history. The history of the disease seems to date back to a year and 3 months, the patient did present with jaundice plus itching and weight loss of 12 Kg in three months, he benefited from a CT scan of the abdomen revealed a large process of the pancreatic head measured 34x36mmwithout signs of mesenteric vessels infiltration, There was dilation of the main pancreatic duct up to 0.7 cm with intra and extra hepatic duct dilation, the common bile duct (CBD) was 19mm diameter (figure 1).

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The patient underwent the first surgery which consists on a Cephalic duodenopancreatectomy. (Figure 2)

Postoperative pathological studies were established and indicated a white tumor with the greatest dimension of 5 cm with irregular borders and central necrosis with a pancreatic margin section of 1 cm and we count 13 local adenomegalis.

Microscopic examination showed a malignant undifferenciated neoplasm with a sarcomatoid aspect constitut with a range of pleomorphic tumoral cells and fibroblast-like cells that were arranged in a storiform pattern

Invasion of perineural sites was seen without vascular invasion

Surgical margins were intact and No lymph node metastases wereidentified.

The immunohistochemical staining was performed, and the tumor was positive for CD-10, lysozyme, alpha 1-antichymotrypsin, and vimentin . Also, it was negative for S-100P, pancytokeratin, epithelial membrane antigen, desmin, CD34, smooth muscle, antigen, MDM2, and CD34, DOG1, myogenine and HMD45.therefore, the tumor diagnosis was compatible with UPS

The post-operative course was uneventful, and patient was discharged on the fifteenth postoperative day, he received adjuvant therapy, it consist in chemotherapy with Intensity modulated radiotherapy with a dose of 45Gy in 25 seances

At 1year follow-up the patient had benefit of an abdominal CT scan which exhibited a hypodense lesion at the second liver segments relevant to a distant recurrence of his disease (figure 3), magnetic resonance imaging (MRI) demonstrated 2 hepatichyposignal lesions in T1 and hypersignial in T2, which one of them located in the II segment and the other one in the VI segment, they measured 36x27 mm and 12x9 mm respectively (figure 4).

The patient after DRCs for cancer multidisciplinary consultation meetings did go under surgery,

In the surigical exploration; we did found two lesions which the first one was poorly limited in the second and the III segment and another lesion on the underside of VI segment of the liver. The surgical strategy consisted of a right lobectomy with metatsasectomy of the VI segment lesion (figure 5,6). The postoperative complications were unremarkable and the patient was declared discharged on D4 postoperative. Postoperative follow-up is favorable with a 8 months decline.

Pathological examination revealed hepatic localization of a malignant pleomorphic cell tumor proliferation which may be consistent with metastasis of the already diagnosed pleomorphic sarcoma of the pancreas.

#### **Discussion:-**

Primary pancreatic sarcomas are an exceptional disease representing less than 1% of pancreatic neoplasms. They derive from the mesenchymal support tissue in the pancreas, Pleomorphic sarcomas are more rarest, which only 15 cases report find in the literature (picture 1). In our case we are presenting an undifferentiated pleomorphic sarcoma in the pancreas with hepatic metastases without local recurrence which is the second case in the literature.(5)

Pleomorphic sarcomas are a heterogeneous group of mesenchymal neoplasms with widely varied clinical behavior. Undifferentiated pleomorphic sarcoma is not histologically distinctive and may show a fascicular, storiform, or sheet-like appearance(6).

In a serie of 109 patients diagnosed with UPS bilal et all notes metastatic disease developed in 29 patients (32%). Pulmonary metastases were the most common and occurred in 26 of 29 patients (90%). Bone metastasis occurred in 2 patients (7%), whereas one patient only (3%) developed liver metastases(2).

In the present case, the patient was treated initially with cephalic duodenopancrtoctomy for the Primary pancreatic tumor, followed by 25 cycles of adjuvant radiotherapy with chemotherapy. Followed by left hepatic lobectomy and metastasectomy for the metastatic lessions. Moreover. Because Undifferentiated pleomorphic sarcoma is extremely rare and more rare in pancreatic location and even more rare with hepatic metastasis, the standardization of its

treatments including surgery, chemotherapy, and radiotherapy requires further investigations. Previous studies and our experience suggested excision combined with adjuvant therapy could improve its prognosis. Furthermore, this case could provide a helpful reference to aid in the decision-making process for the diagnosis and treatment of pancreatic undifferentiated pleomorphic sarcoma.

## **Conclusion:-**

The Undifferentiated Pleomorphic Sarcoma of the pancreas is exceptional and his distance metastasis is rarest, however We pretend that our experience with this case will enrich our knowledge about UPS of the pancreas to perform a representative clinical analysis and adequate therapeutic approach

## **Echonography:**

Echonography.						
Authors (ref. no.)	Year	Age	Sex	Location	Treatment	Follow-up (months)
Ishiguchi [7]	1986	44	M	Body-tail	Left pancreatectomy, splenectomy	NED 17
Garvey et al. [8]	1989	77	M	Uncinate lobe	Enucleation	NED 48
Pascal et al. [9]	1989	39	M	head	Pancreaticoduodenectomy	DOC
Allen et al. [10]	1990	46	M	Body-head, Local invasion	80% pancreatectomy, splenectomy, subtotal gastrectomy	DOD 5
Tsujimura et al. [11]	1992	43	F	tail	Pancreatectomy, splenectomy	NED 5
Ben [49]	1993	72	M	Body-tail	Left pancreatectomy, splenectomy	DOD 12
Bal'en [12]	1993	37	M	Body-tail	Extended left pancreatectomy	DOD 7
Haba et al. [13]	1996	70	M	head	Pancreaticoduodenectomy	NED 22
Bastian et al. [14]	1999	67	M	body	Left pancreatectomy, splenectomy, transverse colectomy, subtotal gastrectomy	NED 34
Darvishian et al. [15]	2002	74	M	head	Pancreaticoduodenectomy	NED 4
Akatsu et al. [16]	2005	67	M	Body-tail	Left pancreatectomy, splenectomy, transverse colectomy, total gastrectomy	NED 37
Yu et al. [17]	2008	67	M	head	Pancreaticoduodenectomy	DOD 11
Jarry et al. [17]	2010	45	M	head	Multidisciplinary treatment	NED 11
BehnamSanei [18]	2016	72	F	Head and neck	Pancreaticoduodenectomy	NED 22
Ze Liang [19]	2022	37	M		body and tail	
Our case	2022	63	M		head	

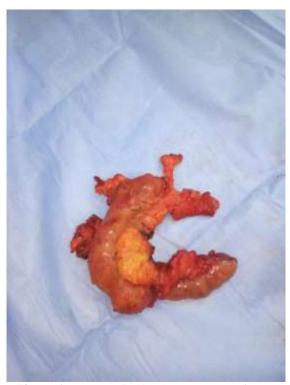


Figure 1:- Pancreaticoduodenectomy specimen.



**Figure 2:-** The preoperative abdominal CT scan exhibited a hypodenselesion in the pancreas headwithintra and extra hepatic duct dilation.



Figure 3:- Months follow-up abdominal CT scan which exhibited a hypodense lesion at the second liver segments.

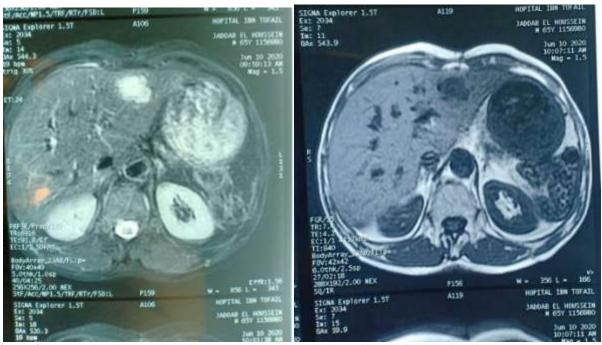
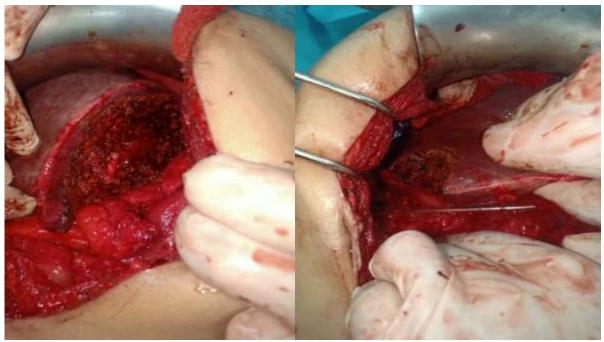
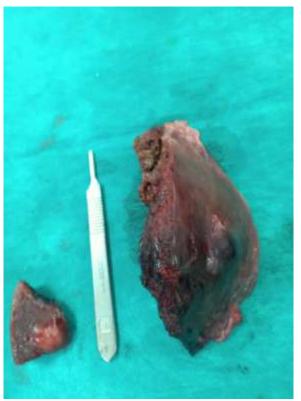


Figure 4:- Magnetic resonance imaging (MRI) demonstrated 2 hepatic hyposignal lesions in T1 and hypersignial in T2



**Figure 5:-** The preoperative; photograph showing the slices of surgical resection.



**Figure 6:-** Photograph showing the pieces of resection with 2 metastatic lesions.

Figure 6: table of cases reported in the literature

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