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RETROPANCREATIC FOLLICULAR DENDRITIC CELL SARCOMA: A DIAGNOSTIC CHALLANGE

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ABSTRACT Follicular dendritic cell sarcoma [FDCS] is a rare nodal or extranodal soft tissue tumor, uncommon in retropancreatic node. Etiopathogenesis of the disease is not clear and it is often misdiagnosed. Presentation is usually incidental. Imaging and histopathology features can be confusing. A high index of suspicion and a specific approach to diagnosis is of utmost importance. Immunohistochemistry is a must for diagnosis. CD 21 and CD 23 are specific for FDCS. Management is by complete resection. The role of chemotherapy and radiation is not clear. We report a case of retropancreatic FDCS and briefly review our approach to its management.

KEYWORDS : Dendritic cell, sarcoma, retropancreatic tumor, Pancreatic surgery

INTRODUCTION

Primary retropancreatic tumors are uncommon.1The clinical and imaging features of many of these tumors overlap leading to a difficulty in diagnosis. The management and prognosis of these tumors vary as per their cell of origin and the nature of the tumor. Hence, a histological diagnosis is important in these cases.1Follicular dendritic cell sarcoma [FDCS] which comprises 0.4% of soft tissue sarcomas overall, is one of the rare primary retropancreatic tumors seldom considered in the differential diagnosis.FDCS is often misdiagnosed even after complete imaging and histopathology andan appropriate immunohistochemistry panel is mandatory for a definitive diagnosis.2,3We present here a case of a retropancreatic FDCS.

CASE REPORT

A 44 year old lady presented with an incidentally detected retropancreatic mass. The computed tomography [CT] scan revealed an 4.1 x 3.2 x 3.2 cm lesion abutting the inferior part of head of the pancreas with heterogenous enhancement and few cystic areas(Figure 1).

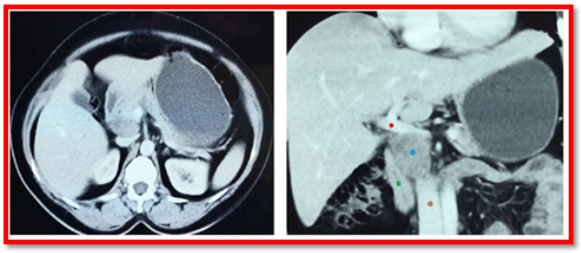

Figure 1: Contrast enhanced Computed tomography image shows axial (left) and coronal (right) image showing the relationship of the mass (blue) with inferior vena cava (Yellow), duodenum (green) and portal vein (red) Magnetic resonance imaging scan [MRI] showed a similar lesion elevating the uncinate process, second and third part of duodenum and compressing the infrahepatic inferior vena cava with restricted diffusion, bright on T2 and hypointense on T1(Figure 2).

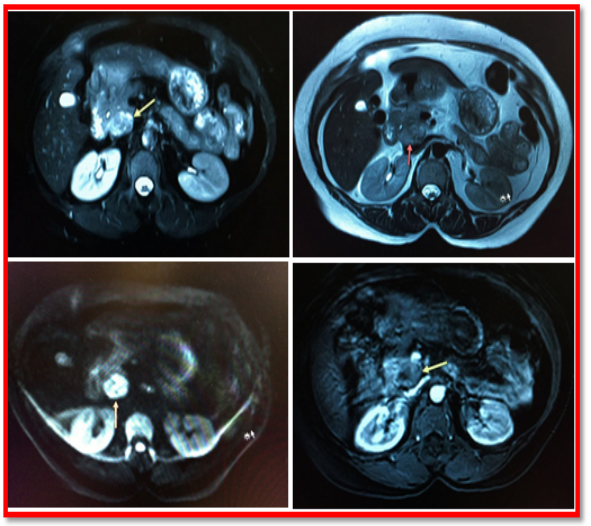

Figure 2 : Magnetic resonance imaging images show the mass in T2 SPAIR on top left, T2 on top right, Diffusion weighted image showing restricted diffusion on bottom left and contrast enhanced image with its relationship to renal vein and superior mesenteric vein in bottom right images Positron emission tomography [PET CT] showed a hypermetabolic mass with peripancreatic lymphadenopathy(Figure 3). Endoscopy and endoscopic ultrasound revealed a heterogenous 4 x 3.2 cm lesion. The core biopsy reported FDCSwith CD 21 and CD 23 positivity.As the lesion was very close to thepancreas with no well defined fat plane and peripancreatic lymphadenopathy, the patient was planned for pylorus preserving pancreaticoduodenectomy to achieve R0 resection.

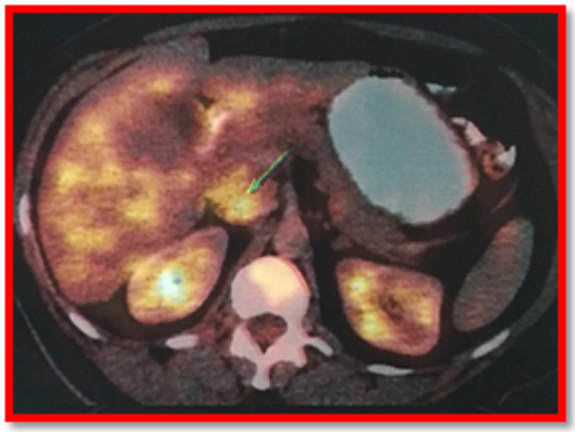

Figure 3: Positron emission tomography [PET CT] scan image shows the hypermetabolic mass in retropancreatric position [Arrow]
Intraoperatively, a hard,nodular 4 x 4 x 3 cm mass with dense perilesional fibrotic reaction was identified posteroinferior to the pancreatic head, firmly adherent to pancreas and duodenum and free from IVC and portal vein(Figure 4).

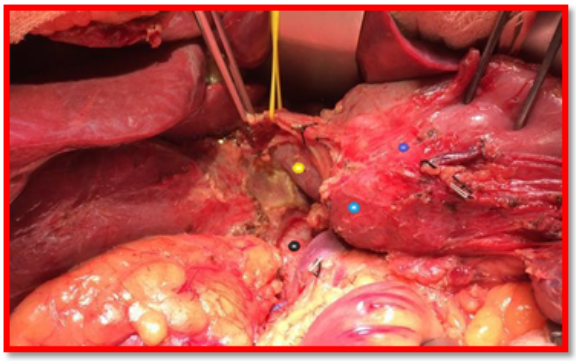

Figure 4: Intraoperative image showing the mass [light blue] in retropancreatic region and its relation to duodenum [dark blue], inferior vena cava [black] and portal vein [yellow] On gross examination, there was a retropancreatic pale brown solid mass with few cystic areas. Microscopically, lobular aggregates of whorls & fascicles of spindle cells with elongated, pleomorphic nuclei and eosinophilic cytoplasm were seen with sprinkling of small lymphocytes (Figure 5). Immunohistochemistry [IHC] revealed CD 21 and CD 23 strong positivity. The patient recovered uneventfully.

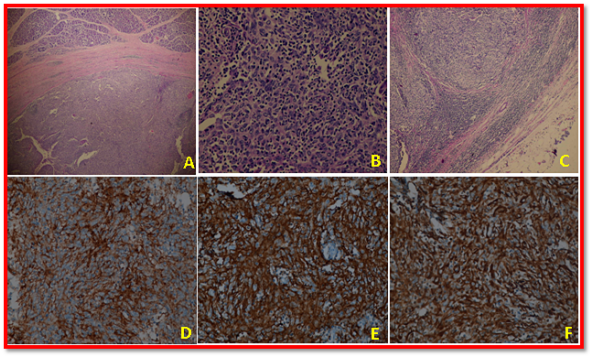

Figure 5: Microscopy images of the specimen show the mass separated from the posterior border of the pancreas by a rim of fibrocollagenous tissue [A], with lymphocytes sprinkled in the mass amongst the dendritic cells [B] and also contains normal lymphoid tissue peripherally [C]. Immunohistochemistry show CD 21 positivity [D], CD 23 positivity [E] and vimentin positivity [F].

DISCUSSION

Retropancreatic masses can be primary or secondary, solid or cystic and neoplastic or non-neoplastic. Common primary solid retropancreatic masses include primary sarcoma [most common], leiomyosarcoma, malignant fibrous histiocytoma, gastrointestinal stromal tumor, schwannoma, lymphoma or germ cell tumor.[1]

Follicular dendritic cell sarcoma is an uncommon primary nodal neoplasm in this area and not routinely considered in diagnosis. The tumor can be nodal, extranodal conventional or extranodal inflammatory pseudotumor. The most common sites include cervical, axillary and mediastinal nodes. Weak associations have been made with Castleman's disease and Epstein Barr virus infection.[2,3,4]

The differentiation of retropancreatic tumors is important because treatment differs as per the type ranging from only chemotherapy for lymphoma or identification of primary and treatment tailored to primary in case of metastasis, only surgery for follicular dendritic cell sarcoma, fibroid, liposarcoma or a combination of chemotherapy and surgery for GIST or neuroendocrine tumors or retroperitoneal sarcomas.[1,5]

The differential diagnosis can be achieved by a series of well-planned investigations.CT scan, MRI and PET/CT scan can be useful noninvasive investigations in this regard. Inspite of these investigations, the diagnosis is often difficult due to an overlap in clinico-epidemiological and radiological findings. In those cases, invasive imaging, histopathological examination and immunohistochemical tests may help in achieving a definitive diagnosis.[5,6]

Immunohistochemistry is very important here as it gives a very clear information about the site of origin of tumor [synaptophysin {neuroendocrine}, vimentin{mesenchymal}, epithelial membrane antigen {epithelial origin}], nature of tumor [CD 20 {lymphoma}, CD1a {langerhan cell histiocytosis(LCH)}], prognosis[ki-67 (mib1)] and can also provide a therapeutic benefit [c-kit in GIST].[6]

An asymptomatic middle aged female patient with a retropancreatic well defined lesion with heterogenous enhancement, few cystic areas, no calification and a mass effect on adjacent structures on CT scan can have leiomyosarcoma, GIST, lymphoma, malignant fibrous histiocytoma, desmoid, FDCS or metastasis as the diagnosis.[1,3,5] Poorly differentiated neuroendocrine cancers also cannot be ruled out. MRI helps in narrowing down the differential diagnosis as can be seen in our case where the lesion shows restricted diffusion which is a feature of a highly cellular lesion, hence more likely to be malignant lymphoma, metastasis, poorly differentiated neuroendocrine tumor or retropancreatic sarcoma. GIST is ruled out as it doesn't show restricted diffusion. PET CT is next and rules out the neuroendocrine tumors which don't show intense uptake on PET which is the finding in our case.[1,5]

EUS and EUS or CT guided biopsy is the next investigation as the yield of EUS alone is inferior compared to EUS with biopsy. Also, asfindings of histopathology can show a considerable overlap amongst mesenchymal tumors, immunohistochemistry is a must for management.[5,6,7] For our case, the diagnosis was achieved on the basis of a suspicion of mesenchymal tumor on histopathology and a well judged immunohistochemistry. The markers CD 21, CD 23 and CD 35 are positive in 83%, 90% and 44% of FDCS.[4,10] The tumor is negative for c-kit, CD 34, CD 1a, cytokeratin and desmin which rules out GISTs, lymphomas andLCH.[4,6,8,9]

FDCS is more common in 5th decade and females. The most common presentation in abdominal disease include incidental detection.[3,4,8] Characteristic CT findings include a well-defined mass with internal necrosis, occasional calcification, cystic components and regional lymphadenopathy.[3]

Histopathology shows findings similar to what was seen in our case. These findings should raise a suspicion for FDCS and appropriate IHC panel than applied.[8,9]

The treatment guidelines are not established. However, most reports consider the tumor as an intermediate grade and recommend R0 resection.[3,8,10] The role of chemoradiation therapy is not clear.[1,4]Extranodal and/or intra-abdominal location, tumor diameter > 6 cm, presence of coagulative necrosis, a high mitotic count [>4 mitoses/10 high-power fields] and cellular atypia are adverse prognostic factors.[8,10]

CONCLUSION

FDCS is an intermediate grade soft tissue tumor uncommon in the retropancreatic region.It usually presents incidentally and is often misdiagnosed on histopathology. Immunohistochemistry is necessary for the diagnosis in suspicious cases and surgical resection with negative margins is the treatment.

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