Extragastrointestinal Stromal Tumor Of The Greater Omentum Presenting A Large Multilocular Cystic Mass: A Case Report And Review Of Literature

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Citation

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Abstract

Primary neoplasms of the greater omentum are rare and and may pose difficulty for the preoperative histologic diagnosis. We report a case of extragastrointestinal stromal tumor of the greater omentum in a patient with chronic pancreatitis, which has been tentatively diagnosed of a benign omental cyst preoperatively. Computed Tomography showed a multiloculated large cystic mass of the greater omentum with multiple thick septa and a solid component and pancreatic calcification. Fine needle aspiration cytology showed a few histiocyte in a bloody background and it was negative for malignant cells. The patient underwent resection of the huge cyst including a major part of the greater omentum. Histologically, the tumor was consisted of sheets and strands of relatively uniform medium sized round epithelioid cells. Immunohistochemical staining of the tumor cells was positive for CD117 (c-kit, a transmembrane tyrosine kinase receptor), CD34 and vimentin, but it was negative for S-100 and Calretinin. These findings were compatible with extragastrointestinal stromal tumor (EGIST). We emphasized the possibility of extragastrointestinal stromal tumor of the greater omentum, although the preoperative radiologic findings suggested as a benign omental cyst.

INTRODUCTION

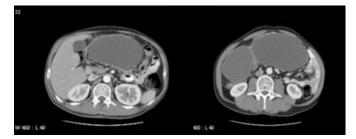
Extragastrointestinal stromal tumor (EGIST) is a unique tumor that occurs outside the gastrointestinal tract. EGIST shows c-kit expression and a histologic appearance similar to gastrointestinal stromal tumor (GIST)(₁). Only a few cases of extragastrointestinal stromal tumor involving the mesentry, retroperitoneum and the bladder have been reported (_{1,2,3}). In previous reported cases, most extragastrointestinal stromal tumors presented as a circumscribed or lobulated firm mass, and only rarely were cystic changes(₁). In this paper, we describe a case of the extragastrointestinal stromal tumor of greater omentum that presented with a huge cystic mass.

CASE REPORT

A 49-year-old man presented with abdominal distension and a palpable mass in his abdomen. He had history of alcoholic pancreatitis. Computed Tomography showed a multiseptated large cyst of the greater omentum and pancreatic calcification (Fig1).

Figure 1

Figure 1: Computed Tomography showed multiseptated large cyst of the greater omentum and thickened septa



Fine needle aspiration cytology showed a few histiocyte in a bloody background and the biopsy was negative for malignant cells. The levels of pancreatic enzymes (amylase, and lipase) were within normal limits upon cystic fluid analysis. The cystic mass was diagnosed as a likely omental cyst, but also had some possibility of being a pancreatic pseudocyst. During the operation, the tumors were completely resected along with major part of the greater omentum. The tumors are well circumscribed and they were not adhered to the surrounding tissue, and they contains two cysts that were 22cm and 19cm in diameter (Fig. 2,3).

Figure 2

Figure 2: Gross view of the tumors located in the greater omentum. The cystic tumor was well circumscribed and multiseptated.

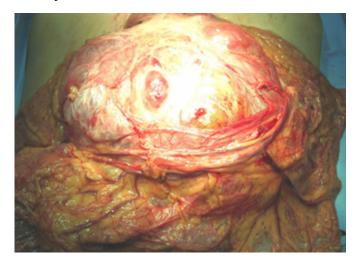


Figure 3

Figure 3: A cut surface of the resected tumor. The tumor consisted of two cysts with watery and hemorrhagic fluid and without a solid portion.

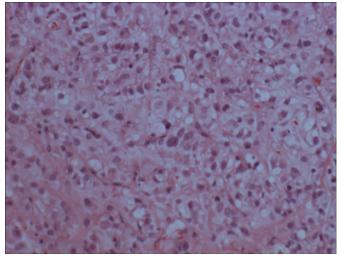


The two cysts were filled with watery fluid and hemorrhagic materials and had no solid portion. We analyzed this omental tumor histologically and immunohistochemically.

Histologically the tumor was biphasic, and consisted of sheets and strands of relatively uniform medium sized round epithelioid cells with focal vacuolar change. There were two mitoses per fifty high-power fields with hemorrhage and necrosis (Fig. 4).

Figure 4

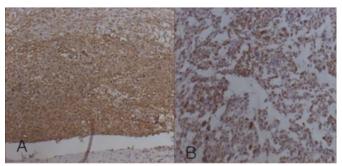
Figure 4: Microscopic appearance of the stromal tumor presenting epitheloid cells with focal vacuolar change(H & E, original magnification, x200)



Immunohistochemical staining of the tumor cell was positive for CD117 (c-kit, a transmembrane tyrosine kinase receptor), CD34 and vimentin but negative for S-100 and Calretinin (Fig. 5). These findings were compatible with extragastrointestinal stromal tumor (EGIST). Six months later, the patient was in good condition, and there was no sign of any recurrence of the tumor.

Figure 5

Figure 5: Immunohistochemical staining of the stromal tumor.



The stromal cells are positive for CD34(original magnification x 100)

The stromal cells are positive for CD117(original magnification x 100)

DISCUSSION

Extragastrointestinal stromal tumor (EGIST) is an infrequent neoplasm and the preoperative diagnosis is very difficult. Preoperative fine needle aspiration cytology has been reported as useful diagnostic tool($_{4}$). In our case, the preoperative fine needle aspiration cytology was not helpful for diagnosis. In Nakagawa's study, preoperative angiography was used as a helpful diagnostic tool. Angiography showed the origin of the tumor by revealing that the blood supply came from the right gastroepiploic arcade(₅). Upon gross examination, most EGISTs present as a circumscribed firm masses, but our case presented as a huge cystic mass without a solid portion. Preoperatively, our case had been diagnosed as benign cyst of the greater omentum by the cytologic and radiologic findings. In the previous reported cases, a high mitotic rate(>5/50 HPF) and a large size(>5cm) were significantly associated with the clinical outcome for EGIST(1). However, Reith et al have suggested that more than 2 mitoses per 50 HPF may be useful in predicting behavior in EGIST(6). In Yamamoto et al's study, most EGISTs were >5 cm in size because of the tumor having enough space to grow, and tumor size was not significantly associated with the prognosis(1). In some studies the two mitoses per fifty high-power fields suggested a high malignant potential, so it was necessary to closely follow-up our case that had the two mitoses per fifty highpower fields and a size of 22cm. This case demonstrated that even though extragastrointestinal stromal tumor of the greater omentum is a rare disease, it should be considered in the differential diagnosis for patients presenting with huge sized cystic lesions of the greater omentum.

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