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Extra Gastrointestinal Stromal Tumor of the Omentum

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1. Abstract

Gastrointestinal Stromal Tumors (GISTs) are tumors arising from the interstitial cells of Cajal. In very rare occasions, these set of tumors are detected outside the gastrointestinal tract, hence are referred to as Extra-Gastrointestinal Stromal Tumors (EGST). EGISTs may be primarily arising from structures outside the gastrointestinal tract or metastatic from a gastrointestinal stromal tumor. Radiological investigations like computed tomography scans, magnetic resonance scans and PET scans are used to aid the diagnosis of these tumors. Additionally, Histological and immunohistochemical tools are used to confirm the diagnosis of GISTs. Here we report a 75-year-old gentleman who was incidentally diagnosed with an EGIST in the Sultan Qaboos University Hospital, Muscat, Oman. This case report discusses and compares the modalities used in the process of achieving the diagnosis of these tumors.

2. Introduction

Gastrointestinal Stromal Tumors (GISTs) are tumors arising from the Interstitial Cells of Cajal (ICC) and it's precursors that are characteristically present within the gastrointestinal tract [1]. Despite GISTs being considered the most common mesenchymal tumors of the gastrointestinal tract, it only accounts for one percent of primary gastrointestinal malignancies [1, 2]. The most common site for Gastrointestinal stromal tumors is the stomach accounting for 60-70% of GISTs, other less common areas include the small intestines, large intestines and the esophagus [3, 4]. In addition to that, the literature reported rare occasions of GISTs occurring in extra gastrointestinal regions like the omentum, the mesentery and the retroperitoneum, giving them the term (EGISTs) that stand for Extra Gastrointestinal Stromal Tumors [3].

3. Case Report

A 75 year old gentle man who was diagnosed with prostate adenocarcinoma, has initially underwent an abdominal ultrasound which reported an incidental finding of a large epigastric mass measuring (15 cm x 5.6 cm x 11.2 cm). This mass appeared to have a solid-cystic appearance and did not have a clear plane of origin.

An abdominal Computed Tomography scan (CT) with contrast was performed and revealed a large complex mass measuring (16.4 cm x 8.5 cm x 7.2 cm) seen in the right upper abdomen. It was predominantly cystic as shown in (Figure 1, Figure 2). The liver was also found to have small multiple hypodense lesions that were most likely cystic, the largest seen in segment V measuring (1.9 x 1.7 cm).

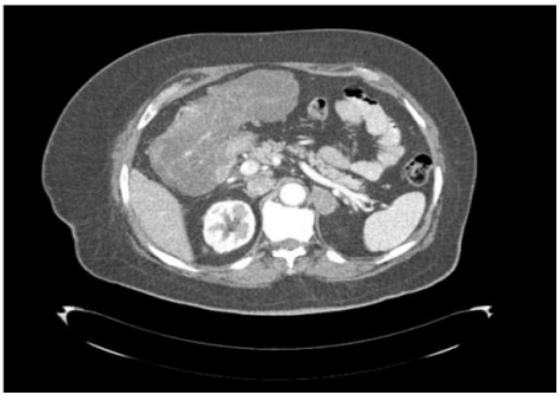

Figure 1: Transverse imaging of a contrast enhance CT abdomen demonstrating a large complex mass measuring (16.4 cm x 8.5 cm x 7.2 cm) seen in the right upper abdomen. No significant abdominal or pelvic lymph nodes visualized.

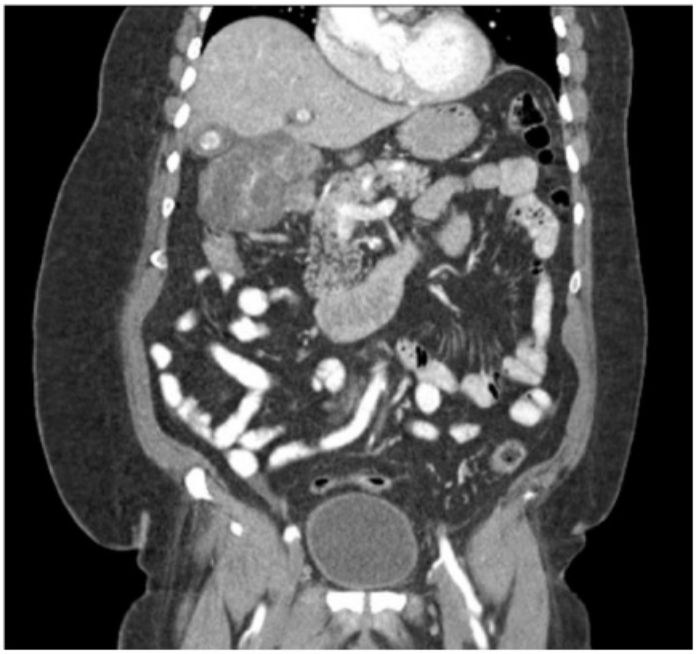

Figure 2 : Coronal imaging of contrast enhanced abdomen and pelvis CT demonstrating a large complex mass seen in the right upper abdomen that is predominantly cystic with enhancing soft tissue. It measures (16.4 x 8.5 x 7.2 cm). The mass appears to be abutting the lower surface of the liver and the gall bladder without a fat plane in between. The liver shows multiple hypodense varying size lesions, largest seen in segment V and measuring (1.9 x 1.7 cm).

An MRI was then performed, further characterizing the lesion to be inseparable from the omentum, and appears to be supplied by the gastro duodenal artery. The apparent consistency of the mass in MRI supported the CT findings, describing it as multilobulated and predominantly cystic with thick septations internally as shown in (Figure 3).

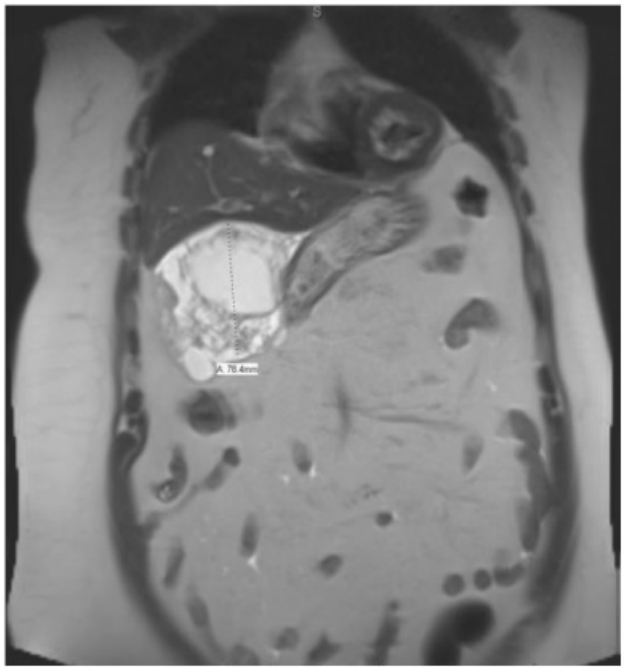

Figure 3: Transverse imaging of an abdomen and pelvis MRI demonstrating a relatively well-defined, multilobulated, predominantly cystic lesion measuring (17 cm x 8 cm). There's an evidence of thick internal septations and soft tissue components seen occupying the right subhepatic space.

CEA, CA19-9 and AFP tumor marker levels were ordered which all came back to be within normal limits. Despite the radio logically striking features, the patient remained clinically featureless and asymptomatic. He denied symptoms of abdominal pain or fullness, early satiety, nausea, vomiting, or constitutional symptoms. His bowel habits were normal and he did not report any episodes of per rectal bleeding.

The next step taken in light of the above findings was an exploratory laparotomy with the intent to directly evaluate and remove the abdominal mass. Upon exposing the peritoneal cavity, a large omental cyst extending from the liver border to the pylorus of the stomach was seen. The adhesions between the mass and the surrounding structures were easily released and the cyst was then separated from the omentum using ultrasonic device. Finally, the histological analysis of the mass revealed a neoplastic proliferation of ovoid to spindle cells with Extra Gastrointestinal Stromal Tumor of the Omentum moderate eosinophilic cytoplasm and elongated nuclei, suggesting features of a Gastro Intestinal Stromal Tumor (GIST). Immune histochemical staining confirmed the diagnosis by showing positive reactivity for CD117, DOG1 and CD34 antibodies, however Pan-CK antibody was negative.

4. Discussion

Gastrointestinal stromal tumors arise from the Interstitial cells of Cajal and its precursors [3]. There are a number of investigatory combinations that were found to be of diagnostic significance when approaching these set of tumors. Radiologically, the use of ultrasound in GIST has several limitations. Tumors that are less than 5cm in size are not easily detected using ultrasound, in addition to that, evaluation of tumor characteristics and complications such as perforation, necrosis, and ulceration are very difficult to achieve using this modality [5]. In our patient, ultrasound was effective in incidentally picking up the tumor which was (15 cm x 5.6 cm x 11.2 cm) in size. CT and MRI on the other hand, can optimize the radiological diagnostic experience of GISTs by further identifying the location of the tumor along with the features of perforation, invasion and metastasis [6, 7]. In this patient, the mentioned radiological modalities were useful for detecting the location along with the possible origin of the tumor, as it did not show a clear attachment band to the bowel, instead it appeared to originate from the omentum, which correlated ahead with the diagnosis of Extra Gastrointestinal Stromal Tumor (EGIST). The confirmation of GIST tumors is established with both histopathological and immunophenotypical bases [8]. Histologically, GISTS's are found to be either spindle celled (70%), epithelioid (20%) or mixed (10%) [9]. In our case, the tumor was found to show ovoid spindle shape cells, which is the most common histopathological type of GIST [9]. Immunophenotypically, c-kit receptor expression of certain protooncogenes is used to establish the diagnosis of GIST [10, 11]. The case we're presenting was immune positive for CD117 which is considered a characteristic feature of GIST. In addition to that,

immunohistochemical staining was also positive for DOG1 and CD34 antibodies.

EGISTs in particular, are tumors that share the same histological and immunohistochemical features of Gastrointestinal Stromal Tumors but are found to be unattached to the gastrointestinal tract. Upon the diagnosis of EGISTs, establishing a confident conclusion to the origin of the diagnosed tumor is crucial. As EGITSs can be originally arising from tissue outside the gastrointestinal tract, it can also be metastatic from other GIST tumors [12, 13]. In the case we're reporting, no evidence of a gastrointestinal stromal tumor was found, neither radio logically nor during laparotomy. Moreover, the attachment of the tumor intraoperatively was clearly visualized to be to the omentum. The mentioned finding along with the histology and the immunohistochemistry results confirms the diagnosis of EGIST.

5. Conclusion

EGISTs are a rare group of tumors that share the same morphology as Gastointestinal stromal tumors, they arise from outside the gastrointestinal tract. Diagnosis of these tumors usually involves CT, MRI, histology and immunohistochemistry tests, in addition to modalities like PET scans that are occasionally used, however, this investigation wasn't performed in our patient. Some studies suggest that EGISTs behave in a more aggressive manner compared to stromal tumors arising from the gastrointestinal tract. However, very limited data on the prognosis and pattern of these tumors are present due to the rarity of the disease.

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