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Case report

A case of undifferentiated pleomorphic sarcoma of a retro-gastric origin, case report and review of literature

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ABSTRACT

Introduction and importance: Undifferentiated pleomorphic sarcoma (UPS) is one of the most common sarcomas affecting elderly patients, majority of UPS usually in the extremities, trunk, and retroperitoneum. However, its rarely observed in the digestive system. There are minimal data published regarding this topic.
Case presentation: Here we present a 58 years old female patient with a rare case of the pleomorphic sarcoma of retro-gastric mass measures about (24.7 × 23.5 × 17.4 cm) who underwent exploration laparotomy founded a freely and mobile retro-gastric mass measuring 30 × 20 cm with a cystic and solid component. The mass was freely and mobile located with no true gastric relation is the first one to be reported.
Clinical discussion: The undifferentiated pleomorphic sarcomas or MFH has been regarded as the most common soft tissue sarcoma for many years (Kabashima et al., 2017). The incidence increases with age, with an inclusive incidence of about 1–2 cases per 100,000 patients per year, predominately 1.2:1 for males. Undifferentiated pleomorphic sarcoma of the stomach is rare and counted to have a worse prognosis when compared to the other types of pleomorphic malignant fibrous histiocytoma.
Conclusion: We present a rare case of high-grade undifferentiated pleomorphic sarcoma of the retro-gastric mass with no invasion to the surrounding structures, managed with surgical resection of the tumor.

1. Introduction

Undifferentiated pleomorphic sarcomas (UPS) or pleomorphic malignant fibrous histiocytoma (MFH) has been regarded as the most common soft tissue sarcoma for many years; extremities are the most affected sites with undifferentiated high-grade pleomorphic sarcomas, even more than the trunk or retroperitoneum; however, the incident in the gastrointestinal tract is sporadic [1,2]. UPS is counter to have a worse prognosis when compared to the other types of MFH [3]. Here, we report a rare case of retro–gastric undifferentiated pleomorphic sarcomas with no direct relationship with the stomach discuss the relevant literature. This case report has been written in line with the SCARE 2020 criteria [4].

2. Case presentation

A pleomorphic sarcoma originating from the stomach is rare. There are minimal data published regarding this topic. Here we present a rare case of the pleomorphic sarcoma of retro-gastric mass in a 58 years old female patient who complained of an abdominal distention for five months, associated with anorexia, nausea, subjective fever, and weight loss about 20 kg within three months. The surgical history and family history were insignificant. She denied smoking, alcohol consumption, and psychosocial issues. The abdomen's physical examination consisted of a mass centrally located down to the pelvis.

Her Initial The laboratory chemical examination did not show any abnormal results. CT scan of abdomen and pelvis showed a huge heterogeneous mixed cystic, solid mass seen extending from the lesser sac down to the upper pelvis, and it measures about 24.7 × 23.5 × 17.4 cm. The mass is inseparable from the pancreas, but it does not seem to be originating from it. There is a probably claw sign between the tumor and the gastric curvature of the stomach, where the main vascular supply of the mass developing from this point (Figs. 1, 2).

Ultrasound (US) guided core biopsy was done, and surgical pathology showed a malignant neoplasm with extensive necrosis. The cells are pleomorphic and spindles, focally arranged in bundles with a myxoid background. Atypical mitosis is identified. The possibility of gastrointestinal stromal tumor (GIST) is considered. The Immunohistochemistry

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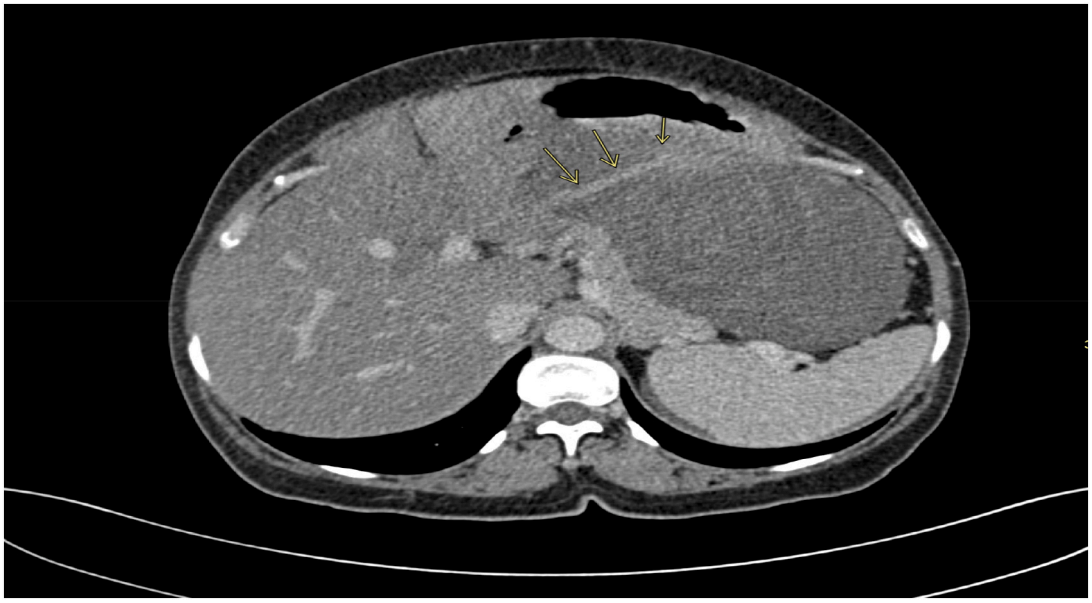

Fig. 1. Abdominal computed tomography (CT) Axial view showed a 23.5 × 17.4 (cm) retro-gastric tumor.

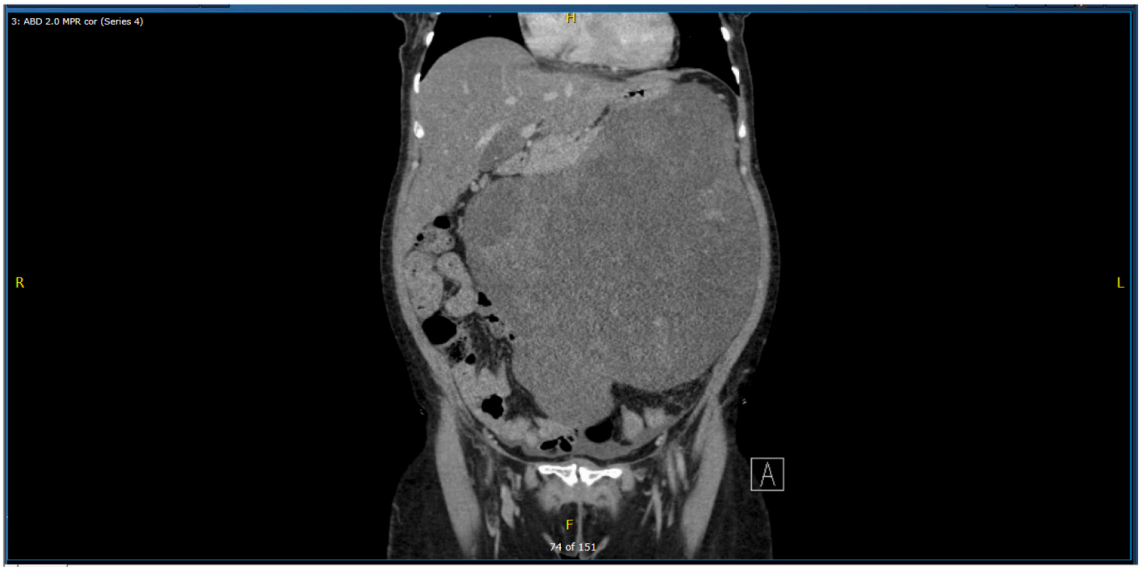

Fig. 2. Abdominal computed tomography (CT) Coronal view showed a 24.7 × 23.5 (cm) retro-gastric tumor.

stains show the neoplastic cells to be positive for smooth muscle actin (SMA) and Vimentin. They are negative for Desmoid, CD117, and CD34. The final diagnosis was high-grade undifferentiated pleomorphic sarcoma of abdominal mass. Confirmation of the diagnosis was made in association with the Mayo Clinic-Mayo Medical Laboratories.

The patient consented to receive the operation an exploratory laparotomy by the Surgical Oncology team with findings of a substantial retro-gastric mass measuring 30 × 20 cm with a cystic and solid component. This mass compressed the large bowel inferiorly, the small bowel to the side laterally, and the stomach was pushed anteriorly. The mass was freely and mobile located behind the stomach and has no relation to any structure except for a narrow pedicle of vascularity from the lesser curvature of the stomach. The mass protruded easily, and complete excision was done with ligation of vascular pedicle of the lesser curvature in addition to distal gastrectomy just in case of any micro invasion of the lesser curve.

Histopathology yielded mass sized 30 × 22 × 11 cm, weight 5780 g, encapsulated well-defined with a daughter cyst of 2.5 × 2 × 1 cm. Tumor differentiation score 3, mitotic Rate 20/10 high power field (HPF), microscopic necrosis is present, extent 30%, (Score I). The tumor is seen to be extending to the capsule, no lymph-vascular invasion, pathological Staging: pT2b NX, and the diagnosis was pleomorphic undifferentiated sarcoma (Fig. 3).

The postoperative course was uneventful. The patient was discharged home on day six postoperative for follow-up in the outpatient department. This case was discussed at the GI tumor board meeting for the potential of adjuvant radiotherapy. However, no sufficient data was supporting such treatment [5].

A Follow-up CT scan annually for the first two years and the last three years showed no evidence of recurrence or metastasis.

3. Discussion

The World Health Organization Classification of tumors, the

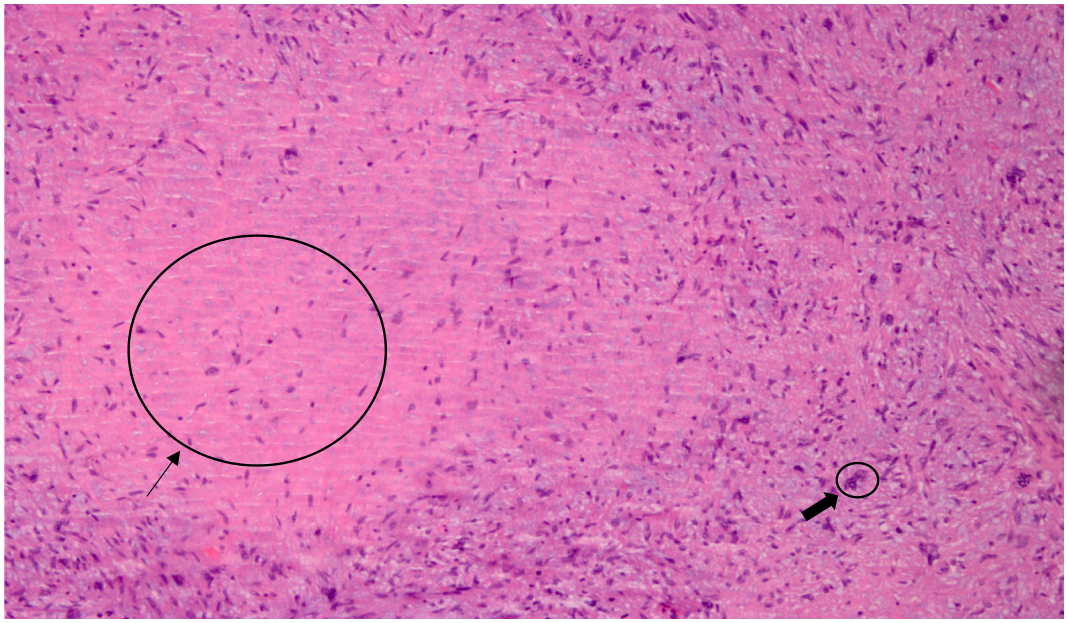

Fig. 3. Histopathology slide of pleomorphic undifferentiated sarcoma, bold arrow Showed multinucleated giant cell. Narrow arrow showed area of necrosis.

undifferentiated pleomorphic sarcomas (UPS) substitutes the term pleomorphic malignant fibrous histiocytoma (MFH), and both are used as synonymous. In adults, the undifferentiated pleomorphic sarcomas or MFH has been the most common soft tissue sarcoma for many years [1]. The median age at diagnosis of MFH was 64-year-old. The incidence increases with age, with an inclusive incidence of about 1–2 cases per 100,000 patients per year with a predominance of approximately 1.2:1 for the males [1,2]. According to a published paper of literature review and case report regarding primary gastric undifferentiated high-grade pleomorphic sarcoma with a total of 16 cases were reported as direct UPS or MFH, main chief complaints were a pain in 9 patients, two patients with bleeding, two subjects with amenorrhea, one with weight loss, one with no complain and the last case was reported as fever [3]. Unlike our patient, her main complaint at time presentation was progressively increasing abdominal distention.

The tumor origin was the body of the stomach in 9 patients, antrum of the stomach in 5 patients, two patients in the cardia [3]. In comparison, the tumor origin in our case was retro-gastric origin with no true gastric relation, which is the first one to be reported.

The mean age was 61 years old, the mean diameter of the tumor was 6.7 cm and ranged from 1 to 15 cm, invasion or metastasis was present in seven patients of the sixteen, and died by the time they were reported. 2-year survivors were only 4 of the 16 patients. 5-year survival has not been reported in the literature yet [3]. However, our case exceeds three years of survival now.

In a retrospective study of 12 different medical oncology institutions of 112 patients in Turkey, the original site of the high-grade undifferentiated pleomorphic sarcoma tumors for the majority of cases was located in the upper extremity, which accounted for 46 patients (41.1%), the remainder were 33 patients in the lower extremity (29.5%), 16 patients in retroperitoneum (14.3%), 13 patients in viscera (11.6%), four patients in the head and neck (3.6%). The size of the tumors ranged from 0.6 to 30 cm. Regarding the histological subtypes, the predominant subtype was the pleomorphic variant (90.2%); at the same time, the other less frequent variants were the giant cell (6.3%) and inflammatory (3.6%). For staging concerns, (50.9%) were stage II at diagnosis, and the rest were stage III [4].

4. Conclusion

Undifferentiated pleomorphic sarcoma of the retro gastric is rare, counted to have a worse prognosis when compared to the other types of malignant fibrous histiocytoma; for this; proper clinical evaluation, imaging workup, and diagnostic procedures are necessary to a suspicious case to achieve the diagnosis and manage the patient's condition before the progression of the disease.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Ethical approval

No ethical approval is required. Our institution does not demand ethical approval for case reports, as these projects are not investigational.

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CRediT authorship contribution statement

Wafa Almalki, Majdi Alzahrani, wrote and edited the manuscript. Bilal Baker and IhabAlssaqqaf managed the case, Bilal Baker guided, edited and reviewed the manuscript. All authors read and approved the final version to be published.

Declaration of competing interest

The authors declare no conflicts of interest.

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