PRIMARY GASTRIC LOW-GRADE FIBROMYXOID SARCOMA IN A JORDANIAN PATIEN

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ABSTRACT

Fibromyxoid sarcoma (Evans tumor) is a low-grade soft tissue tumor with a benign histopathological aspect that usually originates in the deep soft tissue of the proximal limbs or trunk in young adults, but which can initiate in the gastrointestinal tract (specifically in the small intestine or colon). It is relatively common in the lower extremities, and can be found in the head, neck, and abdominal wall, but it is very rare in the viscera, abdominal cavity, and bowel mesentery. This case study concerns an 18 year-old female Jordanian patient who experienced epigastric pain with a huge epigastric mass at King Hussein Military Hospital, Amman, in 2020. Abdominal computed tomography showed a hard mass starting from the lesser curvature of the gastric body. Macroscopically, there was a mass of 10*10 cm in maximal diameter with white-tan area, starting from the gastric wall. Histopathologically, there was a mesenchymal tumor of blunt spindle cells with collagen and inflammatory cells. Immunohistochemically, it was positive for MUC4 and FUS-CREB3L1 fusion in fibromyxoid sarcoma. The histopathological results confirmed low-grade gastric fibromyxoid sarcoma.

Keywords: fibromyxoid sarcoma; low grade; gastric.

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INTRODUCTION

Fibromyxoid sarcomas (FSs) are metastasizing tumors that may affect any area of soft tissue with a benign aspect. They commonly affect patients aged 3-78 years in the deep soft tissue of the inferior limbs, especially the thigh, occurring less commonly in the chest wall, axilla, shoulder, inguinal area, buttock, and neck, with rare cases in the gastrointestinal tract.[1] Typical FSs contain bland, spindle-shaped cells, which may recur or metastasize. Empirical evidence indicates that 14/33 FS patients (42%) died within 3-42 years, 21/33 patients (64%) experienced recurrence following 15 years, and 15/33 patients (45%) experienced metastases after 45 years.[2] .It is difficult to differentiate low-grade FS from mesenchymal tumors.

Recurrent fusion genes are recognized in FSs, 95% of which are FUS-CREB3L2, while less than 5% are FUS-CREB3L1.[3] Similar fusion genes have been seen in hyalinizing spindle cell tumors with giant rosettes (HSCTGR), a shaped variant of FSs. EWSR1 has been seen in FSs, but rarely.[4] Low-grade FSs include bland spindle cells with collagenous-myxoid stroma, and appear in young male and female adults

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equally as a slow-growing, painless mass in the deep soft tissue of the proximal limbs or trunk, appearing rarely in the viscera. These are managed with operative excision, but may show recurrences and increasing metastasis, with the lungs being the most frequent metastatic location. These tumors are genetically characterized by translocations of the FUS gene.

A 18 year-old Jordanian woman presented to hospital experiencing epigastric pain with distension one month prior to admission, reporting remarkable weight loss. Other than abdominal bulging, there were no other positive findings. She had no history of surgery or injury, or any pertinentfamily medical history. Abdominal computed tomography showed a hard mass originating from the lesser curvature of the stomach. Enhanced abdominal CT demonstrated that the mass was starting from the lesser curvature of the stomach and compressing the left lobe of the liver. Computed tomography of abdomen showed a 10x10 multiloculated mass at the lesser curvature of the stomach, close to the pancreas and major vessels (Fig. 1, Fig. 2).

MRI showed large lobulated soft tissue mass with central necrosis at lesser sac originating from the gastric wall, compressing the left lobe of the liver, encasing left gastric artery with multiple regional lymph nodes. The left gastric artery was penetrating inside the tumor mass. Esophago-gastric endoscopy found a submucosal mass of the stomach. Endoscopic ultrasound showed normal healthy mucosa. Upper endoscopy showed normal mucosa of the stomach with compression from the lesser curvature side. Biopsies were taken and showed minimal gastritis.

The patient underwent surgery to excise the tumor, but complete excision could not be performed. The challenges in achieving complete resection were primarily attributed to the tumor's substantial size and encasement of the left gastric artery including its origin from the coeliac trunk, so debulking surgery was done. Macroscopically, there was a mass with white-tan cut partial cystic area, starting from the gastric wall. There were no metastases. Microscopic sections of the tumor showed mostly bland fibroblasts, with little atypia and lobular patterns in heterogeneous myxoid and collagenous stroma. Mitotic figures were low to intermediate (0–4/10HPF). Rare pericollagenous rosettes were identified. Immunohistochemistry was positive for vimentin, and negative for BCl2, Desmin, S-100, smooth muscle action (SMA), c-Kit, beta-catenin, and synaptophysin. Immunohistochemistry was weakly positive for CD34.

Histopathologically, a giant collagenous rosette in the hypercellular area was found. The tumor cells with a collagenous rosette were positive for MUC4. Positive Immunohistochemical MUC4 confirmed FSs of the stomach. Laboratory investigation demonstrated anemia (hemoglobin 9.6 g/L); otherwise, all other hematologic and biochemical parameters were normal. All tumor markers were normal.

After surgery, the patient was offered definitive chemoradiotherapy with standard fractionated chemoradiation (5000-5400 cGy in 180-200 cGy per fraction), concomitant weekly cisplatin chemotherapy, with 54 Gy in 2 Gy fractions as their primary treatment. Agents used in the first-line setting were doxorubicin by four, Caelyx by one, and combination of ifosfamide and doxorubicin by one. Ifosfamide was used as a second-line treatment, and rabectedin was used as a third- and fourth-line treatment.

DISCUSSION AND CONCLUSION

The studied case of primary gastric FS exhibited histopathological clusters of blunt spindle cells with hyalinization and collagenous rosettes, MUC4 immunopositivity,and unique FUS-CREB3L1 fusion gene diagnosed FSs. Benign or low-grade mesenchymal tumors of the stomach are differentiated from FS in terms of GIST, inflammatory myofibroblastic tumor, schwannoma, and

solitary fibrous tumor.[5] Low-grade FSs are malignant fibroblastic soft tissue neoplasms with alternating fibrous and myxoids and delayed recurrence and metastasis. An alternative name for low-grade FS is Evans tumor or hyalinizing spindle cell tumor with giant rosettes.

These tumors resemble other soft tissue tumors, and are commonly found in younger adults; they account for 20% of all tumors in adolescents. Males are a slightly more frequently involved with painless slow-growing mass over the course of years. Low-grade FSs are malignant tumors with alternating collagenous and myxoids with bland spindle cells organized in a fascicular or whirling growth shape.

The cause of low-grade FSs is unknown. In adults, low-grade FSs originate from the subfascial soft tissues of the proximal limbs and the trunk. Less frequent locations are the abdominal cavity, the retroperitoneum or mediastinum. In children, the superficial soft tissues of the head and neck region are affected (Table I).

Author Location	Evans (1993)[4]	Goodlad (1995)[6]	Devaney (1990)[1]	Fukunaga (1996)[5]	Dvornik (1997)[2]	Lee (2004)[7]	Total
Thigh	2	3		1			6
Shoulder	3					1	4
Inguinal area	2	1			1		4
Chest wall	1	3					4
Buttock	1	1					2
Neck	1		1				2
Popliteal		1					1
Axilla		1					1
Perineum	1						1
Psoas muscle		1					1
Mesentery	1						1
Total	12	11	1	1	1	1	27

Table I. Summary of cases of low-grade fibromyxoid sarcoma during history.

The studied case exhibited a solid well-demarcated fibrous tumor with mucoid areas macroscopically. Microscopically, there was a mixture of collagenous with some cells and more cellular myxoid areas with transitions between the two. In addition, there are spindle cells in a fascicular whirling or storiform shape, small vessels with staghorn vascularity, giant hyaline collagen rosettes surrounded by epithelioid tumor cells, and sclerosing epithelioid fibrosarcoma features. Immunohistochemistry stains with MUC4, an increasingly sensitive and specific marker for low low-grade FSs, revealed sclerosing epithelioid fibrosarcoma. Low-grade FSs demonstrated FUS-CREB3L2 or FUS-CREB3L1 gene fusions. Radiologically, there was a multinodular, heterogeneous, gyriform with complex solid cystic elements and well-defined tumor boundaries. Ultrasonically, there were solid soft tissue tumors with heterogeneous echogenicity. Computed tomography showed hypodense mass with calcifications. Magnetic imaging shows tumors with a striated or gyriform shape of signal intensity, with peritumoral edema and a split fat sign.

A few FSs demonstrated epithelioid shape in limited areas. The case did not have clear epithelioid element. FS is a scarce soft tissue neoplasia. These tumors commonly appear in young adults, often without clinical features; they are frequently detected by chance on imaging as a slow-growing mass. Despite their bland aspect, FSs demonstrate progression with increased recurrence (9%), metastasis (6%), and mortality (2%).[3] Histopathologically, FSs have cytological bland spindle cells with indistinct cytoplasm and no mitotic figures. Fibrous and myxoid regions are found. The fibrous regions are paucicellular, with abundant feathery stromal collagen with whorled patterns of growth. The myxoid areas are more cellular than the fibrous ones, with prominent vasculature. Hyalinizing spindle cell tumor with giant rosettes is a variant of FSs with prominent paucicellular hyalinized nodules covered by plump tumor cells.[6]

10% of FSs have zones with atypia, high cellularity or epithelioid morphology. Recurrences and metastases commonly look like the initial neoplasm; recurrences may demonstrate hypercellular nodular zones with high mitotic activity. These tumors are MUC4-positive, and include FUSCREB3L2 fusion of FSs. MUC4 immunostain is a specific and sensitive marker for FSs. It has a potential use as a confirmation marker for FSs following global gene expression profiling, which is recognized differential upregulation of the mucin 4 gene in FSs. Recurrences and metastasis are reduced during the first five years following excision, but rates of recurrence subsequently increase in the long term, with a total recurrence incidence of more than 60%, and mortality of more than 40%. FS metastases have been recorded more than 40 years following primary resection. The main frequent locations for metastasis are lung, pleura, bone and liver.

In conclusion, the studied case presented with a primary gastric low-grade FS, a scarce disease and must be differentiated from visceral soft tissue tumors. Histopathological examination and an immunohistochemical analysis with molecular study was able to confirm the diagnosis.

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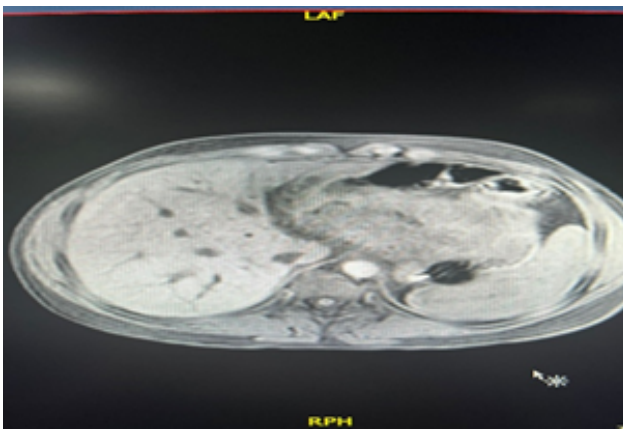

Fig. I. Abdominal CT scan.

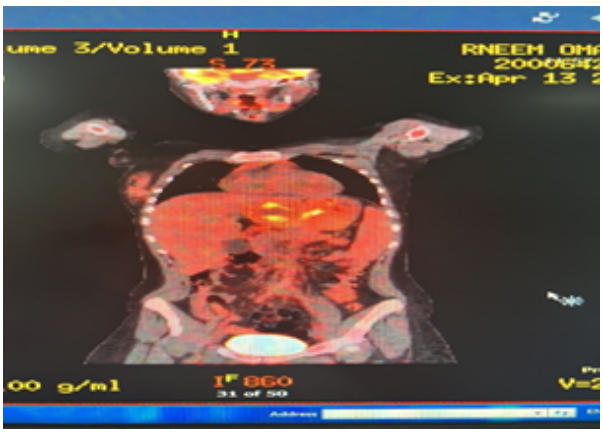

Fig. III. PET scan

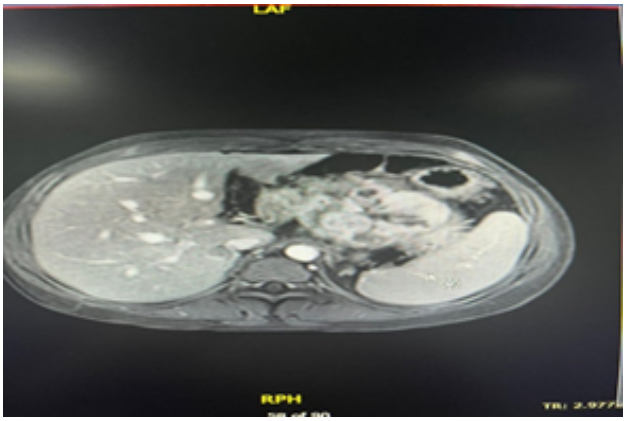

Fig. II. Abdomino-pelvic CT scan.

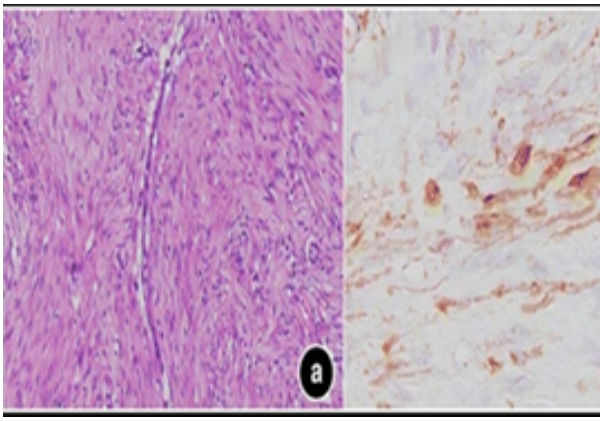

Fig. IV. Histopathology

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