Fibromyxoid sarcoma in the retroperitoneum
A case report
Guyi Wang, BMa, Zhenhua Zhao, MDb, Jianguo Wei, MSc, Jianfeng Yang, MDh,∗

Abstract
Rationale: Low-grade fibromyxoid sarcoma (LGFMS) is a pathological type of fibrosing fibrosarcoma that appears as a distinctive soft tissue mass with bland histological features. It is mostly located in the deep soft tissues of the extremities. Computed tomography (CT) plays an important role in diagnosing fibrosing fibrosarcoma in the abdomen. To date, several studies in the literature have reported on CT features of LGFMS.

Patient concerns and Diagnoses: We report another case of LGFMS, which presented with certain unique CT feature. The anterolateral region and the wall of the cystic nodules showed gradual enhancement and several nourishing vessels were seen after contrast administration. These imaging features were consistent with the histologic findings of LGFMS.

Interventions and Outcomes: The patient underwent tumorectomy and implantation of radioactive implants. Two years after the operation, 1 metastasis mass occur in the right psoas major.

Lessons: These CT features in LGFMS may be useful to assess the histological characteristics of LGFMS to facilitate preoperative diagnosis in the clinical setting and provide the supplemental imaging knowledge for future studies.

Abbreviations: CT = computed tomography, LGFMS = low-grade fibromyxoid sarcoma.

Keywords: computed tomography, x-ray, fibrosarcoma, fibrosis

1. Introduction

Low-grade fibromyxoid sarcoma (LGFMS) is a subtype of fibrosing fibrosarcoma[1] that mostly occurs in the deep soft tissue of the extremities or trunks, but rarely in the retroperitoneum.[2] This tumor usually affects young to middle-aged adults and is more common in men than in women.[3] The etiology and real incidences of LGFMS are generally unknown. Only 3 cases of LGFMS in the abdomen have been described with their computed tomography (CT) findings.[2,4,5] The purpose of this report is to present the different CT features of LGFMS.

2. Case report

We report a new case of LGFMS. Ethical approval and informed consent was waived by the local institutional review board. A 48-year-old man presented with a 13.4 × 10.1 cm mass in the right lower quadrant of the abdomen identified by ultrasonography during a routine health examination. The patient denied symptoms, any significant medical history or medications. Laboratory data were within normal limits except for ferroportin (a tumor marker), which was measured to be 453.22 ng/mL (normal value = 4.63–274.66 ng/mL). Abdominal and pelvic CT demonstrated a large, well-circumscribed, lobulated mass measuring 8.4 × 13.2 × 15.7 cm in the right retroperitoneum. The mass was mostly composed of hypoattenuating areas with a CT value of 22 HU. Contrast administration [1.8 mL/kg of a nonionic contrast agent (Ultravist 300, Schering, Berlin, Germany) via a power injector (Missouri TMXD2001, Ulrich Medical Germany)] at a rate of 2.5 mL/s revealed a hypo-isodense area in the anterolateral region as well as isoattenuated septa (Fig. 1A). There were several nourishing vessels without any area of calcification or hemorrhage focus in the mass (Fig. 1B). On the arterial phase, the CT values of the hypoattenuating area, hypoiodoside area, and septa were 25, 39, and 40 HU, respectively. On the venous phase, the corresponding values were 28.5, 55, and 59 HU, respectively (Fig. 1A, C). The adjacent intestine was compressed and displaced. We did not find any metastasis in the abdominal organs or any lymphadenopathy near the pelvis, retroperitoneum, and para-aortic region.

The patient underwent tumorectomy and implantation of radioactive implants. One 10.1 × 15.0 × 16.0 cm encapsulated lesion was identified in the right retroperitoneum. The histologic findings showed that the tumor contained collagenous fiber nodules and fibrous septa with low cellularity, as well as myxoid areas with moderate cellularity composed of bland spindle cells...
with small hyperchromatic nuclei (Fig. 2A). The cells showed mild nuclear pleomorphism with little mitotic activity and a whorled arrangement in a random manner. Perivascular hypercellularity was noted in some areas. An immunohistochemical staining examination was performed to show that the neoplasm stained strongly and diffusely for vimentin (Fig. 2B). Staining for other markers, including CD 34, CD 68, and NSE, were positive, whereas staining for SMA, NF, Ki-67, MBP, DM, EMA, CK, CD 117, and S 100 were negative. Based on these features, the tumor was diagnosed as LGFMS.

Ultrasonography and abdominal CT performed 2 years after the operation indicated a metastatic mass in the right psoas major (Fig. 3A, B).

3. Discussion
LGFMS was first described by Evans in 1987 as a distinctive mass with bland histological features and a paradoxically aggressive behavior.[5] Grossly, LGFMS is well circumscribed with a capsule.[6] On histological examination, LGFMS typically presents with contrasting fibrous and myxoid areas, moderate to low cellularity, bland-appearing spindle cells with little or no nuclear pleomorphism and rare mitotic figures, and a swirling, whorled growth pattern.[7]

LGFMS in the abdomen usually presents as large well-demarcated masses composed of a hypoattenuating areas or a mixture of hypo- and isodense areas without calcification and/or hemorrhage on CT imaging.[2,3] Several cases of LGFMS have been found in the trunk or falciform ligament with a hemorrhagic focus on its cut section.[2,8] However, all of these masses failed to indicate any hyperattenuating areas of hemorrhage on CT. It may be that CT could not identify areas of micro-hemorrhagic foci.

In our case, the anterolateral region and septa of the mass had a gradual enhancement from the arterial to venous phases. There were several visible nourishing vessels in the mass on the arterial phase. This enhancement modality is consistent with that of fibrous tissues confirmed by histopathologic findings. To our knowledge, these CT features have never been reported on LGFMS in the literature. Harish et al[2] believed the pattern of the vascular distribution on the tumor surface may account for the enhancement modality of the lesion that showed an initial peripheral enhancement with centripetal filling on time delay. However, Fujii et al[3] reported a case of LGFMS in the small bowel mesentery in which the myxoid area had an intense enhancement and believed that the enhancement modality depended on its compact cellularity with prominent capillary networks. As for the enhancement pattern of this case, we...
presume that it may be formed by the distribution of fibrous components and nourishing vessels of the tumor.

These CT findings may aid in differentiating LGFMS from other tumors, such as liposarcoma, the most common tumor in the retroperitoneum. Liposarcoma usually does not contain fibrosis and myxoid components simultaneously. Myxoid liposarcoma is one type of liposarcoma that mainly consists of myxoid and soft tissue components, and it can be distinguished based on its homogeneous or mildly heterogeneous nature given its extracellular myxoid material and fat components that are sometimes present in small amounts without gradual enhancement. This enhancement modality and the existence of nourishing vessels can be seen in myxofibrosarcoma, which contains myxoid and fibrosis components; however, this tumor typically occurs in the elderly and is prone to necrosis or hemorrhage, whereas LGFMS usually is not associated with either outcome. Some scholars have proposed the use of needle biopsy to diagnose LGFMS. However, Harish et al believes that needle biopsy is not specific for diagnosing LGFMS in the abdomen because the excisable tissues are too small for analysis and aspiration biopsy may increase the likelihood of implant metastasis.

CT plays an important role in diagnosing abdominal tumors. In the present case, we found that hypoisodense areas composed of fibrous tissues showed gradual enhancement patterns and several nourishing vessels visible in the mass. Although these CT findings are not specific features of LGFMS, they may be useful to determine the histological characteristics of LGFMS to aid preoperative diagnosis in the clinical setting. Radical tumor resection and postoperative histopathologic examination are the optimal means by which LGFMS of the abdomen is diagnosed and managed.

References

[1] Antonescu CR, Baren A. Spectrum of low-grade fibrosarcomas: a comparative ultrastructural analysis of low-grade myxofibrosarcoma and fibromyxoid sarcoma. Ultrastruct Pathol 2004;28:321–32.
[2] Harish K, Ashok AC, Alva NK. Low grade fibromyxoid sarcoma of the falciform ligament: a case report. BMC Surg 2003;3:7.
[3] Fuji S, Kawawa Y, Horiguchi S, et al. Low-grade fibromyxoid sarcoma of the small bowel mesentery: computed tomography and magnetic resonance imaging findings. Radiat Med 2008;26:244–7.
[4] Koncna J, Liberale G, Haddad J, et al. Diffuse intra-abdominal low grade fibromyxoid sarcoma with hepatic metastases: case report and review of the literature. Int J Surg Case Rep 2015;14:40–3.
[5] Evans HL. Low-grade fibromyxoid sarcoma. A report of two metastasizing neoplasms having a deceptive benign appearance. Am J Clin Pathol 1987;88:615–9.
[6] Maeda E, Ohta S, Watadani T, et al. Imaging findings of thoracic low-grade fibromyxoid sarcoma: report of three cases. Jpn J Radiol 2009;27:375–80.
[7] Evans HL. Low-grade fibromyxoid sarcoma: a clinicopathologic study of 33 cases with long-term follow-up. Am J Surg Pathol 2011;35:1450–62.
[8] Kim SY, Kim MY, Hwang YJ, et al. Low-grade fibromyxoid sarcoma: CT, sonography, and MR findings in 3 cases. J Thorac Imaging 2005;20:294–7.
[9] Barile A, Zugaro L, Catalucci A, et al. Soft tissue liposarcoma: histological subtypes, MRI and CT findings. Radiol Med 2002;104:140–9.
[10] Waters B, Panicek DM, Lefkowitz RA, et al. Low-grade myxofibrosarcoma: CT and MRI patterns in recurrent disease. AJR Am J Roentgenol 2007;188:w193–8.

Figure 3. Two years after the operation, ultrasonography and abdominal computed tomography (CT) were performed. A, Ultrasonography depicted a heterogeneous low echo mass near the right psoas major (white arrow). B, The axial CT image on the venous phase indicated a mass with isohyperdense parenchyma (white arrow).