Experimental and Molecular Pathology 98 (2015) 300–303

Contents lists available at ScienceDirect

Experimental and Molecular Pathology

journal homepage: www.elsevier.com/locate/yexmp

Low-grade fibromyxoid sarcoma of the sigmoid colon

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ARTICLE INFO

Article history:
Received 14 February 2015
Accepted 16 February 2015
Available online 20 February 2015

Keywords:
Low-grade fibromyxoid sarcoma
Sigmoid
CD99
BCL2
MUC4

ABSTRACT

Low-grade fibromyxoid sarcoma (LGFMS) is a rare soft tissue tumor with a slight male predominance. The tumor has a tendency to arise from deep soft tissue of the trunk and lower extremities. Rare cases are reported to arise from the mediastinal and retroperitoneal areas. Its deceptively bland histologic appearance makes this tumor difficult to diagnose. Also, there are several histologic mimics that may hinder in its diagnosis. We report a case of low-grade fibromyxoid sarcoma from a 48-year-old woman, first documented herein to arise from the sigmoid. We also report the value of CD99, BCL2 and MUC4 stains in the diagnosis of this tumor.

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1. Introduction

First described in 1987, LGFMS is considered a rare soft tissue tumor (Evans, 1987). Most patients are young to middle-aged adults, but can also be seen in children or elderlies (Billings et al., 2005; Evans, 1993; Folpe et al., 2000). It has a slight male predominance. The tumor typically occurs in the trunk, deep extremities (Nichols and Cooper, 1994; Zamecnik and Michal, 2000), head, and neck area (Evans, 1987, 1993; Folpe et al., 2000; Rando et al., 2005), and intrathoracic area. Rare cases have been reported in the retroperitoneal, abdominal (Fujii et al., 2008; Harish et al., 2003; Winfield and McKenney, 2007), and intracranial (Saito et al., 2008) areas. The two recognized subtypes are the classical low-grade fibromyxoid sarcoma and low-grade fibromyxoid sarcoma with giant collagen pseudorosettes, also called hyalinizing spindle cell tumor with giant rosettes (Lane et al., 1997). The tumor may be difficult to diagnose by cytomorphology because of its deceptively benign appearance. However CD99, BCL2, and MUC4 immunohistochemical stains may aid in the diagnosis of this tumor. The tumor can be confirmed by its characteristic t(7,16)(q34;p11) translocation (Reid et al., 2003).

2. Clinical summary

The patient is a 48-year-old obese female with a history of hypertension and menometrorrhagia who presented complaining of persistent lower abdominal pain and vaginal bleeding. She also noted an unintentional weight loss of 15 lb but denied hematochezia, melena, and change in stool caliber. Laboratory analysis revealed only moderate normocytic anemia. A transvaginal ultrasound was consistent with an 8.7 cm uterine fibroid. Magnetic resonance imaging of the pelvis revealed a 10.6 × 6.9 × 8.1 cm lobulated mass within the mid-pelvis. The right ovary abutted the mass but appeared separate. The patient

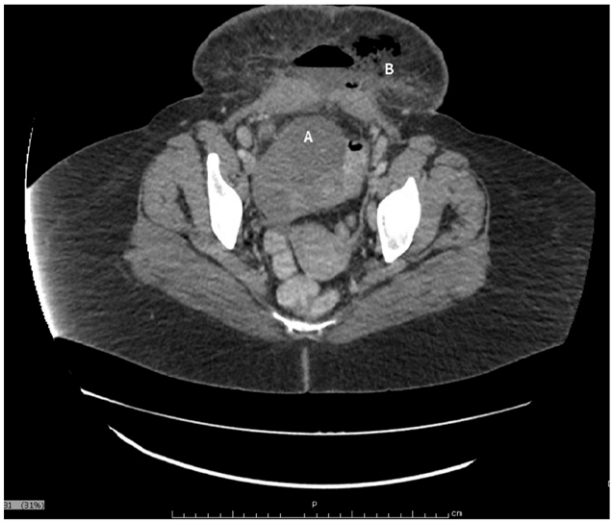

Fig. 1. Axial view of the CT abdomen and pelvis with intravenous contrast. A. Heterogeneous sigmoid mass. B. Postoperative changes at her Pfannenstiel incision.

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http://dx.doi.org/10.1016/j.yexmp.2015.02.007
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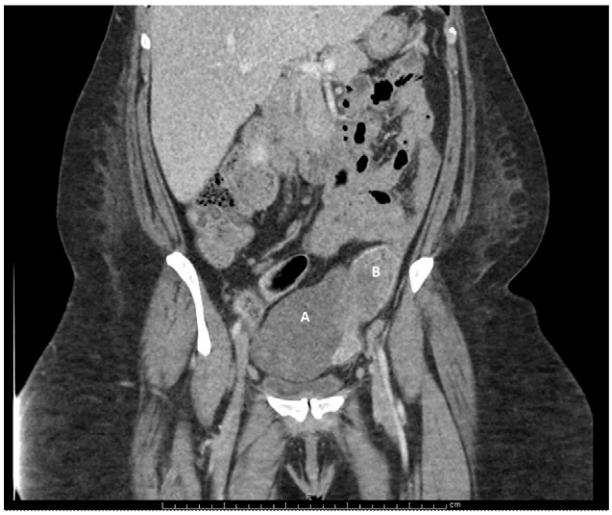

Fig. 2. Coronal view of the CT abdomen and pelvis with intravenous contrast. A. Heterogeneous sigmoid mass. B. Sigmoid colon.

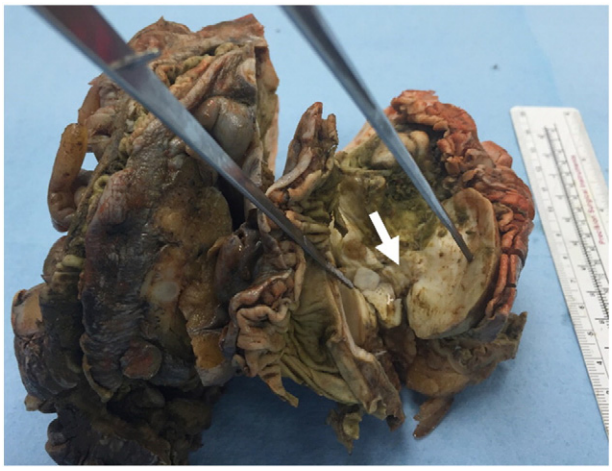

Fig. 4. Mucosal extension. Cut section of the polypoid mass in the mucosal surface showing direct connection of the stalk (arrow) to the subserosal mass.

was taken to the operating room for a total abdominal hysterectomy with bilateral salpingo-oophorectomy for a mass of presumed gynecologic origin. Intraoperatively, a large 12 cm mass located in the pelvic was seen to be arising from the sigmoid colon and there were no other significant abnormalities. A shave biopsy of the mass was obtained and the abdomen was closed with the expectation that further staging of this colonic mass would be needed prior to definitive treatment.

Therefore, computed tomography (CT) of the chest, abdomen, and pelvis were obtained which revealed a 12 cm heterogeneous exophytic mass arising from the sigmoid colon without metastatic disease (Figs. 1 and 2). The carcinoembryonic antigen level was within normal limits at 0.5 ng/mL. An outpatient colonoscopy with biopsy of the colonic mass was planned. However prior to the colonoscopy, she was readmitted for a surgical site infection of the Pfannenstiel incision that necessitated a surgical debridement. The patient later developed a bowel obstruction requiring another emergent operation during this admission. Intraoperatively, there were loops of small bowel adherent to the pelvis and to the sigmoid mass. Consequently, a sigmoidectomy with descending end colostomy and Hartmann's pouch was performed. A short segment of small bowel with an unclear serosal lesion was removed and a segment of small bowel that had been trapped into the pelvis was also resected. Primary anastomoses of both small bowel areas were performed. The patient ultimately recovered after a protracted hospital course and was discharged to home.

3. Pathological findings

Grossly, the tumor was well circumscribed with white whorled cut surface. It measured 11 × 7 × 4.5 cm expanding to the subserosa, to the muscularis propria, and up to the mucosal layer (Fig. 3). Mucosal extension showed a 3.8 × 3.2 × 2.7 cm polypoid mass with a whorled white appearance on cut sections (Fig. 4). The margins were negative of the sarcoma and there was no lymph-vascular invasion. Histologically, the tumor showed a swirling whorled growth pattern with alternating fibrous and myxoid areas. The myxoid area had a low to moderate cellularity composed of bland spindle cells with scant cytoplasm, uniform elongated nuclei, and small inconspicuous nucleoli (Fig. 5). In some areas, the tumor also showed plump epithelioid cells with abundant cytoplasm and strands and nests of clear epithelioid cells separated by eosinophilic hyalinized stroma (Fig. 6). The tumor cells stained negative for SMA, S100, CD34, Desmin, Caldesmon, Calponin, CKit, DOG-1, EMA, and B-Catenin immunohistochemical stains. Ki67 also showed a low mitotic index (Fig. 7). Further review of the case gave a high suspicion for LGFMS. CD99, BCL2 and MUC4 stains were subsequently ordered which all came back positive (Fig. 8).

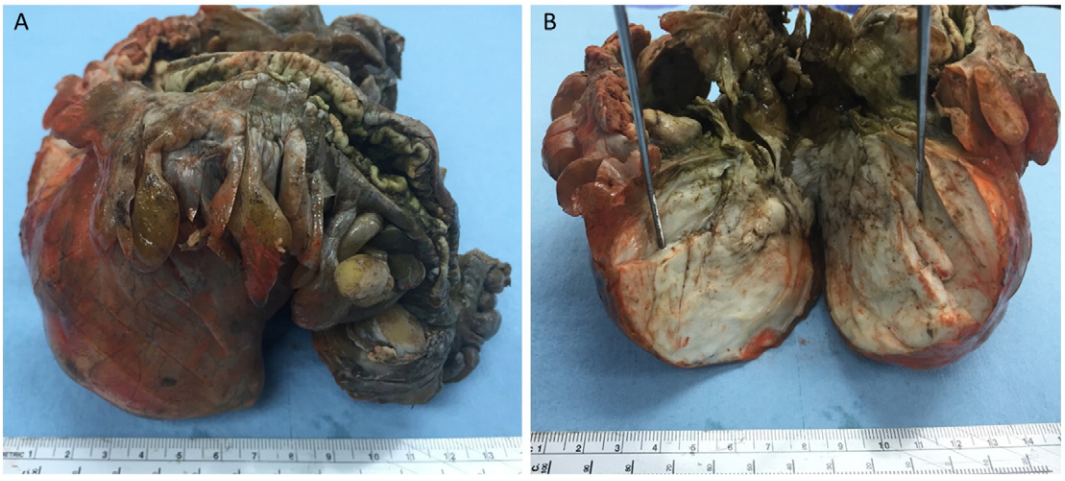

Fig. 3. A. The mass has a locally aggressive growth expanding to the subserosa, muscularis propria, and into the mucosal surface. B. Cut section of the mass showing uniform white whorled surface.