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Case Report
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A Case Report on an Intraluminal Colonic Polypoid Myxoid Sarcoma

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Abstract

Background: Liposarcomas (LPS) are aggressive soft tissue sarcomas which may demonstrate local and distant metastasis and often require a combination of neoadjuvant chemotherapy with surgical resection along with postoperative adjuvant chemotherapy. The disease is known for having a high recurrence rate and may present with symptoms specific to the involved system. Here, we present a patient who had an extremely rare presentation of recurrence within the colon.

Case Presentation: The case reports on a 58-year-old gentleman who had presented with a local myxoid liposarcoma which was initially treated with neoadjuvant chemotherapy and local excision. The patient was then found to have a metastatic lesion in his lung, which was resected by an experienced group of thoracic surgeons. Unfortunately, the patient had again presented with abdominal pain and was found to have a metastatic lesion near obstruction of his right colon, indicating surgical resection.

Conclusion: Liposarcomas rarely, if ever, present with metastasis to the colon. Treatment for such a presentation requires a multi-disciplinary approach involving both medical and surgical oncologists.

Keywords: Intraluminal sarcoma; Myxoid sarcoma; Chemotherapy; Surgical oncology; Histology; Pathology

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Abbreviations

FNCLCC: Fédération Nationale des Centres de Lutte Contre le Cancer; SRC: Sarcoma; FAK: Focal Adhesion Kinase; ROCK: Rho-Associated Protein Kinase

Introduction

Liposarcomas (LPS) are aggressive soft tissue sarcomas which may demonstrate local and distant metastasis and often require a combination of neoadjuvant chemotherapy with surgical resection along with postoperative adjuvant chemotherapy. The disease is known for having a high recurrence rate and may present with symptoms specific to the involved system. Here, we present a patient who had an extremely rare presentation of recurrence within the colon.

Case Presentation

This patient is an obese 58-year-old male with a history of epilepsy who had initially presented in 2014 to the medical oncologist after developing painless left lower extremity swelling. He had a workup which was negative for cardiogenic or vascular sources of his lower extremity swelling, and an MRI was completed at the time demonstrating a large soft tissue mass lateral to the mid-femur, involving the left vastus lateralis muscle. The mass was hypointense on T1 and hyperintense on T2 images, and lobular in nature.

The patient underwent resection of the mass with the surgical oncology team, and was found to have a 13.2 cm × 10.1 cm × 6.7 cm myxoid liposarcoma with mitoses less than 5 per 10 high-powered fields. There was tumor necrosis present along with lymphovascular invasion. At that time, all margins were free of tumor. Grading using the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) system demonstrated a differentiation score of 2, mitotic count score of 1, tumor necrosis

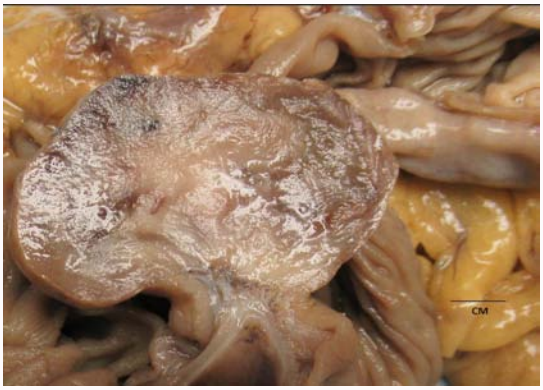

Figure 1: The gross image of a large intraluminal polypoid mass predominantly located in the submucosa with homogenous, myxoid appearance.

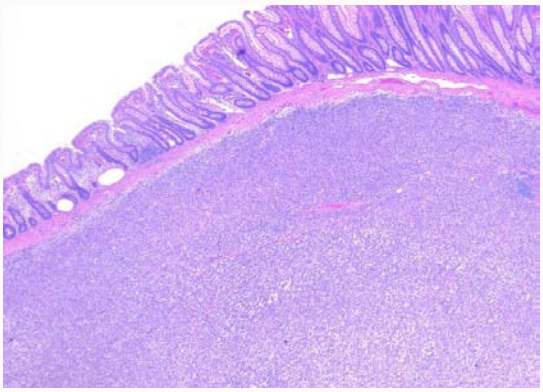

Figure 2: Microscopically, the cellular tumour occupies the submucosa without glandular formation. The overlying colonic mucosa is unremarkable. (Hematoxylin and Eosin stain, original magnification at 25).

score 1. He was defined as having a total histological grade 2 with staging as T2bN0M0. Cytogenic studies demonstrated a translocation 12;16. He had completed three cycles of Gemcitabine with Docetaxel.

Subsequent follow up appointments revealed a metastatic 6.9 cm × 5.9 cm lesion in the right lower lobe requiring surgical resection by the thoracic service, and was upstaged to Stage 4 disease. The patient had no evidence of disease for one year before presenting with abdominal discomfort. Workup included a colonoscopy which demonstrated a large, friable fungating mass with 90% occlusion of the lumen which did not resemble adenocarcinoma of the colon. Multiple biopsies of the mass were with the pathology report demonstrating spindle cells, consistent with recurrent myxoid sarcoma.

The patient had then undergone a pre-operative PET-CT scan demonstrating several mass lesions within the abdomen, including a mass within the peritoneal cavity, the hepatic flexure of the colon and the right iliac fossa. In the operating room, a subxiphoid midline incision was carried down to the umbilicus. Upon entering the abdominal cavity, a large mass in the right lower quadrant retroperitoneum was present at the ileocecal junction. The remainder of the abdomen was evaluated, requiring access into the lesser sac and a total omentectomy. The mass was carefully dissected out in a superior, inferior, medial and lateral circumferential dissection. It was during this dissection that a left upper quadrant 11 cm lesion was noted to extend from the transverse colon into the deep mesentery, and so a separate left colectomy was performed from the

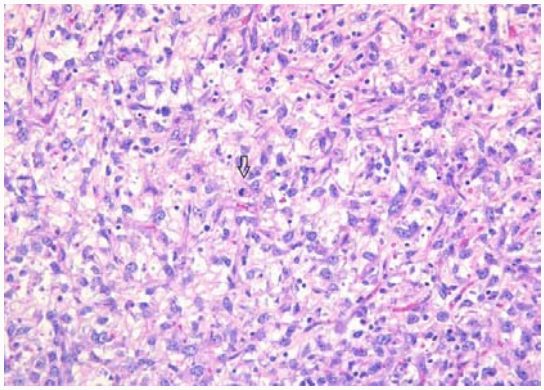

Figure 3: High power microscopic image shows pleomorphic spindle cells with hyperchromatic nuclei and myxoid stroma. Mitotic figures are easily seen (open arrow). (Hematoxylin and Eosin stain, original magnification at 200).

distal transverse colon to the proximal descending colon, along with resection of the affected mesentery.

The ascending colon was then mobilized and the terminal ileum was resected along with part of the distal ileum, the ileocolic and right colic vessels. An iso-colic, side-to-side ileocolic anastomosis was then fashioned between the distal ileum and the left colon. The mesenteric defect was then closed. No further implants were appreciated prior to closure and a Prevena VAC was placed. The patient's post-operative course was uneventful. Six months after surgery, he has completed adjuvant chemotherapy and is otherwise doing well.

Discussion

Liposarcomas (LPS) make up approximately 15% to 20% of all soft-tissue sarcomas and account for only 0.07% to 0.2% of all neoplasms [1,2]. LPSs are encompassed by 5 different histologic subtypes that form 3 different groups. The 3 groups include pleomorphic liposarcomas, well-differentiated and poorly-differentiated liposarcomas, myxoid and round-cell liposarcomas. All 5 subtypes have their own unique genomic alterations that will involve a mechanism of either amplification, loss of tumor suppressors, translocations, or oncogene expression [3]. The peak incidence occurs mainly in the 6^{th} and 7^{th} decades of life and shows no preference to sex or race. An association of LPS and exposure to radiation or chemical substances has been found and is currently undergoing further research. High dose ionizing radiation has been associated with high-grade and poorly differentiated LPS found located near the rim of the radiation field and on average has a latency period of more than eight years. Common chemical substances that have been associated with the formation of LPS include, but not limited to, herbicides, vinyl chloride, arsenic, asbestos, chlorophenols, and androgenic-anabolic steroids [4,5]. Current diagnostic methods for LPS are *via* cytologic analysis of tissue samples collected by percutaneous core needle biopsy in the outpatient setting or after surgical resection. Further staging of metastases will be conducted by performing chest X-rays, Computed Tomography (CT) scans, and Magnetic Resonance Imaging (MRI) scans [6]. Recent studies have shown that Fluorodeoxyglucose-Positron Emission Tomography (FDG-PET) CT scans are beneficial in the detection of soft tissue sarcomas and can accurately distinguish between low-grade and high-grade sarcomas [7].

Liposarcomas are histologically classified into three grades based on the FNCLCC system. FNCLCC utilizes a scoring system that is

based on three histological criteria which are tumor differentiation, mitosis count, and necrosis. Tumor differentiation is based on the histologic type of the tumor and is scored on a scale of 1 to 3 with a score of 1 for a well-differentiated LPS, a score of 2 for a myxoid LPS, a score of 3 for a round-cell LPS, a score of 3 for a pleomorphic LPS, and a score of 3 for a poorly-differentiated LPS. Mitosis count was scored on a scale of 1 to 3 with a score of 1 for 0 to 9 mitoses per 10 High-Power Field (HPF), a score of 2 for 10 to 19 mitoses per 10 HPF, and a score of 3 for ≥ 20 mitoses per 10 HPF. Tumor necrosis was examined on histologic sections of the tumor and is scored on a scale of 0 to 2 with a score of 0 showing no necrosis, a score of 1 showing ≤ 50% necrosis of the examined tumor slide, and a score of 2 showing ≥ 50% necrosis of the examined tumor slide. Scores for lesions were then totaled up and are classified into grades. Grade I liposarcomas have a total score of 2 or 3. Grade II liposarcomas have a total score of 4 or 5. Grade III liposarcomas have a total score of 6, 7 or 8 [8]. One of the most important negative prognostic factors in LPS is a high histological grade [9].

Polypoid Myxoid Liposarcomas (MLPS) account for around 30% to 35% of all liposarcomas and are diagnosed at an average age of 42 years old [10,11]. There is a variant MLPS, called the Round cell LPS and it is recognized as a high-grade, cell-specific type that has a very poor prognosis. MLPS primary tumors occur two thirds of the time in the musculature of the thigh [12]. Other primary locations of these tumors include the upper limbs and the trunk. Studies have shown a local recurrence of around 10% and a metastases rate of 14% after wide or radical surgical resection of the primary tumor. A common location for metastasis is the lungs and it can rarely be found in the liver, spine, kidney, and retroperitoneum (Figure 1) [13]. MLPS found within the retroperitoneum has a lot of room to expand and may take many years before they start to show clinical signs of invasion and metastases [14,15]. Histologically, the MLPS will contain lobules of uniformly spindled, ovoid, and round cells that are within a blue-grey to pink matrix (Figure 2). A key histologic finding for traditional MLPS is the delicate plexiform capillary networks that branch at 45° to 90°. Other findings include a non-clearly visible nucleoli with nuclei containing a uniform chromatin pattern and increased cellularity at the periphery of the tumor (Figure 3) [16]. CT imaging will show hypoattenuation due to a high water content. MRI T1 and T2 imaging are important for the enhancement of thick septa that go through the mass and demonstrate the characteristic fibrous bands [17,18].

A unique diagnostic feature of the myxoid LPS is the translocation t(12:16)(q13;p11) seen in >95% of the tumors [19,20]. The CHOP (C/EBP Homologous Protein) gene is located on 12q13 and it is combined with the FUS (Fused in Sarcoma) gene on 16p11 to form FUS-CHOP. The CHOP gene is a pro-apoptotic transcription factor and will also be referred to as the DDIT3 (DNA Damage-Inducible Transcript 3) gene in literature [20]. The FUS-CHOP fusion promotes cell invasion through the activation of the SRC/FAK/RHO/ROCK signaling axis [21]. These pathways stimulate the upregulation of catalytic domains, cellular migration, proteolytic proteins, and prosurvival signals that all aid in tumor cell invasion and metastases. Pharmaceutically targeting this signaling axis has shown promising results in treating myxoid liposarcomas and could be the future of treatment. A promising drug, dasatinib, has been shown to successfully inhibit the SRC-dependent phosphorylation of FAK, which has an anti-invasion effect on the tumor [21].

Surgical resection with wide margins is the mainstay of treatment for primary and metastatic MLPS. Depending on the extent of invasion by the tumor, resection of the entire muscle it is nearby may be required to obtain negative margins [22]. Chemotherapy has been shown to also play a very large role in the treatment of MLPSs. This specific tumor is very chemosensitive to anthracycline-based treatments and due to this it has been a staple for neoadjuvant and adjuvant therapy [1,19]. Gemcitabine, an anthracycline anticancer drug, has been proven clinically effective with the addition of Docetaxel for the management of metastatic soft tissue tumors such as MLPS [23].

Conclusion

Here, we present a rare presentation of myxoid liposarcoma metastasizing to the colon. Prognosis and resection depends on multidisciplinary planning with neoadjuvant chemotherapy, surgical resection followed by adjuvant chemotherapy to perform successful resection.

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