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Gastric Metastasis of an Extremity Myxofibrosarcoma: A Case Report

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Introduction

Soft tissue sarcomas are rare soft tissue tumors that account for 1% of all malignancies. The most common primary site for this malignancy is an extremity, and metastasis most commonly involves the lungs. Morphologically, myxofibrosarcomas have both fibroblastic and histiocytic features and are one of the most aggressive tumors of the sarcoma family. Moreover, because of their ordinary gross appearance, they can be easily misdiagnosed as benign soft tissue tumors [1]. The average age at diagnosis ranges from sixth to eighth decade with median age being approximately 65 years old [2-4].Prognostic factors include size, grade, location and histologic subtype, with the foremost two determinants most influential on overall outcome. Lastly, necrosis is also an important factor for tumor grading [2]. In this report, we present our experience with this rare extremity tumor metastasizing to the stomach and manifesting as a gastrointestinal bleed-representing the first occurrence thus described in the medical literature.

Case Report

Our patient is a 73-year-old male who initially presented with a soft tissue tumor involving the posterior aspect of the left thigh. The patient had noted the mass growing over several months. Clinically and radiographically, this raised concerns for a soft tissue sarcoma. CT of the chest showed no distant metastasis. The patient was offered the option to undergo radical en bloc surgical resection. He underwent this procedure a few days after and a mass with a size of 12cm x 10cm x 10cm was removed successfully.

Histologic evaluation showed high grade sarcoma with myxoid features, favoring high grade myxofibrosarcoma with histologic grade 3(French Federation of Cancer Centers Sarcoma Group) Moreover, immunohistochemistry was negative for CAM 5.2, S-100, desmin, calponin, myogenin and CD31. Non-specific staining is noted with CD34. The MIB1/Ki67 proliferative index was markedly elevated (greater than 60%) within the high-grade range. Pathologic staging was pT2bNX.

Following surgical resection, the patient received radiation therapy to the limb which was well tolerated. After a lengthy discussion of risks, survival, and benefits of adjuvant chemotherapy, the patient elected against this therapy and chose surveillance alone. Regular 3-4 month surveillance CT scans were reviewed. Two years later a new 1.1cm pulmonary nodule consistent with metastatic disease was found. Our patient underwent radiation therapy with successful response. Another CT scan the following year showed a pulmonary nodule in the contralateral side and the patient underwent radiation once again with successful response. One month later, the patient presented to an outside facility with anemia and extreme fatigue. Lower and upper endoscopies were performed and a gastric mass was identified. The patient was transferred to our facility. During his hospitalization, the patient required multiple transfusions, esophagogastroduodenoscopy revealed a 6-7 cm non-bleeding gastric mass. The patient underwent an exploratory laparotomy, esophagogastroduodenoscopy, and partial gastrectomy, completely excising the mass (Figures 1 and 2). Pathology showed metastatic high-grade myxofibrosarcoma which was compared to the previous surgical pathology of the thigh. The thigh tissue morphologically resembled the gastric tissue. Immunohistochemical stains for Pan-CK, CK7 and CDX-2 performed were negative which supported the diagnosis. Following results and further studies, a CT scan showed a new lung nodule and a MRI showed possible recurrence of the left thigh lesion. The case was discussed at multidisciplinary tumor board meetings and was recommended to undergo radiation therapy for the new lung metastasis and surgical biopsy of potential recurrence on the left thigh. He continues to receive care at our institution while carrying a relatively normal lifestyle and playing golf on a regular basis.

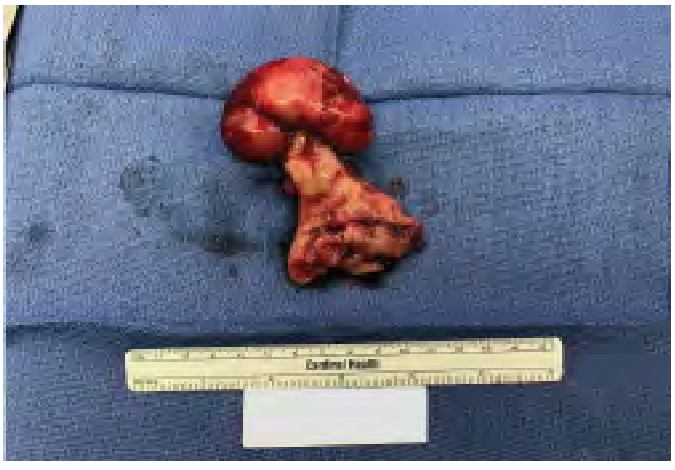

Figure 1: Gross image of stomach mass after complete resection

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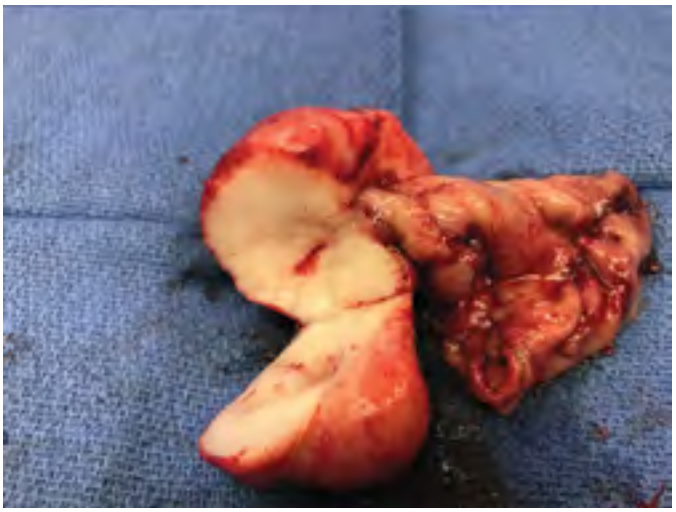

Figure 2: Gross image of stomach mass, sectioned

Discussion

Myxofibrosarcomas arise in the extremities and metastasize to lungs and bone. Sarcomas arising from the gastrointestinal system most commonly metastasize to the liver and peritoneal surfaces. This patient presented with a unique case in which the myxofibrosarcoma metastasized to the alimentary tract and presented as a new entity with anemia and gastrointestinal bleed three years after treatment for the initial primary extremity tumor. Sanfilippo et al., performed a large retrospective study to understand the prognostic factors and survival of patients with myxofibrosarcomas. This study concluded that these tumors have a slight male predominance, most metastasized to the lungs at a rate of 14.6%, while median time for development of metastasis was 11 months. The most important prognostic factors are size and grade which dictate treatment [5]. NCCN guidelines indicate that patients with local disease of the extremities, superficial trunk, head, and neck should undergo En bloc surgical resection, which is a large bulky tumor resection sometimes involving surrounding structures and lymph nodes, and consider radiation therapy with or without adjuvant chemotherapy. For metastatic disease, if single organ, options abound including several permutations of neoadjuvant versus adjuvant chemotherapy as well as radiation alike. If disseminated metastasis is present, options include palliative chemotherapy with or without an admixture of radiation, surgery, observation, or supportive care [6].

The choice to use neoadjuvant chemotherapy for soft tissue sarcomas is usually made by a multidisciplinary team of experts. Anthracycline-based regimens are preferred for the neoadjuvant setting. There are few studies showing potential benefits of this treatment with slight increases of 5-year survival rates [7,8].

We reviewed the literature and found case reports of metastasis to the lung, lymph nodes, brain, bone, mediastinum, mesentery, pancreas, liver, and colon [4,9-13]. We also found that the different literature reviews and clinicopathological analysis suggest that tumors with necrosis or larger than 5cm have significantly higher rates of metastatic relapse [5,14].

Conclusion

Myxofibrosarcomas are rare tumors which show low and high grade histology. High grade malignancies are associated with higher rates of metastasis mainly to lungs and lymph nodes [9]. Metastasis to the gastrointestinal tract is rare and, to our knowledge, this is the only report of a metastasis to the stomach presenting with anemia from a gastrointestinal bleed. Thorough surveillance for recurrence and metastasis is imperative for the best prognosis of patients with myxofibrosarcomas, as well as key recognition of rare cases like the one presented in this manuscript to avoid a delayed diagnosis of metastasis once they present.

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