CLINICAL VIGNETTE

Giant Dedifferentiated Liposarcoma Presenting as GERD

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Introduction

Sarcomas are rare, aggressive malignant tumors, found frequently in young people. They arise from skeletal and extra-skeletal connective tissue, including the peripheral nervous system and the retro-peritoneum. One review of 4550 soft tissue sarcomas found 59% of tumors occurred in the thigh, buttock, groin and upper extremities. Only 18% occurred in the torso, and 13% occurred in the retro-peritoneum.[1] We present an elderly male presenting with shortness of breath and dysphagia, found to be due to dedifferentiated liposarcoma, complicated by multiple pulmonary emboli.

Case Presentation

A 78-year-old male presented with acute shortness of breath and worsening dysphagia. Four months prior he saw another physician for "acid reflux" which was treated with omeprazole with improvement. The shortness of breath developed acutely and his dysphagia was described as a feeling that something was stuck in his esophagus. Past history included atrial flutter corrected by ablation 15 years ago, Hepatitis C treated with ledipasvar/sofosbuvir, resected colon cancer and hypertension. He was anxious with tachypnea, but had resting pulse oximetry of 96% on room air. He also reported a change of bowel habits and he was scheduled for urgent upper endoscopy and colonoscopy. Colonoscopy including biopsies was unremarkable. Upper endoscopy showed no signs of Barrett's esophagitis, intrinsic mass, or eosinophilic esophagitis, but suggested possible extrinsic esophageal compression as the distal lumen was difficult to distend. To pursue this finding, CT Chest revealed "a large, at least 22 cm mass, filling left lower chest and posterior mediastinum." It also showed small pulmonary emboli in segmental branches of the right middle and right lower lobes and moderate bilateral pleural effusions.

He was admitted to the hospital and lower extremity Duplex Scans demonstrated bilateral calf deep vein thromboses, and short segment right great saphenous thrombus at saphenofemoral junction. Bilateral thoracentesis yielded non-diagnostic cytology. However CT guided biopsy of the large pulmonary mass showed a dedifferentiated, high grade liposarcoma. He was evaluated by Oncology to discuss potential treatment options and decided to enroll in hospice and was discharged home on enoxaparin. He died within two months of diagnosis.

Discussion

This patient had a complex presentation, initially thought due to an intrinsic esophageal lesion, which initially improved with Omeprazole. Symptoms recurred and were eventually found to be due to a massive 22 cm liposarcoma which was displacing the heart and mediastinum anteriorly.[2] His evaluation included EGD with negative biopsies and negative cytology from bilateral pleural effusions. His dysphagia was due to displacement of the esophagus by the mass, with was a subtle finding in the distal esophagus. CT guided biopsy of the mass established the final diagnosis.

Liposarcomas are defined as well-differentiated (low grade) sarcomas, followed by dedifferentiated liposarcomas, and less commonly, myxoid, round cell and pleomorphic variations. Well differentiated liposarcomas frequently have an inflammatory cell infiltrate with very low potential to metastasize. Dedifferentiated liposarcomas are defined by the presence of sharply demarcated regions of non-lipogenic sarcomatous tissue within a well differentiated tumor.[4]

Well-differentiated and dedifferentiated liposarcomas have morphological and cytogenic similarities, but have different biologic behavior. Dedifferentiated sarcomas have higher local recurrence rates and potential to metastasize with 20-30% distant recurrence rates, vs 0 for well-differentiated liposarcomas as well as a six fold increased risk of death.[5] Some dedifferentiated liposarcomas are believed to arise from well-differentiated liposarcomas.

Many liposarcomas produce few symptoms, until they compress surrounding tissue, as in this case, with median size at diagnosis of approximately 15 cm. They frequently have laboratory abnormalities of LDH, AFP, and Beta-HCG, which were normal in our case. Early biopsy is important to establish the diagnosis including details of histopathology, with low risk for seeding.[6] This patient's DVTs and pulmonary embody may have been related to a neoplastic hypercoagulable state, though this was not specifically assessed.

Treatment consists of surgical removal or debulking if possible. Large dedifferentiated tumors can be treated with neo-adjuvant chemotherapy, with or without preoperative radiation therapy in attempt to prolong survival,[7] which this patient declined. Others have reported radiation therapy of dedifferentiated liposarcoma to be well tolerated.[8]

Conclusion

This patient's initial symptoms were misdiagnosed because his severe acid reflux symptoms responded to acid blocking therapy. He did not return for additional evaluation until his symptoms progressed and included worsening dysphagia and dyspnea. Endoscopy was initially performed due to concern for lower esophageal pathology. Although no gross findings were identified, a subtle finding of decreased distensibility of the lower esophagus lumen prompted additional imaging which resulted in the correct diagnosis.

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