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Linitis Plastica of Muscle Fascia Presenting as Tiptoeing Gait

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Case Presentation

A 49-year-old woman presented initially with a 1-year history of painful spasms and progressive stiffness of her lower limbs. A never-smoker, she had had no significant prior illness. Clinical examination demonstrated a tiptoeing gait pattern, with normal muscle tone and downgoing plantar reflexes. Serial creatinine phosphokinase levels were normal. Prompted by a normocytic, normochromic anemia, serial GI endoscopies were performed and were normal. Computed tomography scans of the thorax, abdomen, pelvis, and a subsequent laparoscopy, were nondiagnostic. Magnetic resonance imaging showed a signal change in pelvic girdle muscles suggestive of myositis. Additional laboratory investigations were performed, including autoimmune and antineuronal antibodies, myositis-specific and associated autoantibodies, and serology testing for hepatitis, HIV, and Lyme disease. All were negative. Serum human chorionic gonadotropin (HCG) levels were elevated, but all other tumor markers were normal. Anti–gastric-parietal cell antibodies were elevated. Positron emission tomography–computed tomography scanning revealed two small foci of uptake in the anterior-superior mediastinum as well as bilateral pelvic girdle and lower-extremity myositis (Fig 1). A nondiagnostic biopsy of mediastinal lymph nodes was performed on two separate occasions.

Eight months after initial presentation, the patient's condition had gradually but significantly deteriorated. She was unable to rise from sitting and was mobilizing with a stick. She had worsening tiptoeing gait. A trial of high-dose prednisolone (1 mg/kg) yielded negligible clinical benefit. Magnetic resonance imaging of the femurs showed extensive muscle edema in the adductor and quadriceps musculature bilaterally (Figs 1A and 1B). Initial muscle biopsy showed mild type 2 fiber atrophy only but was otherwise normal. Cued by the clinical similarity of gait with cases of eosinophilic fasciitis, a subsequent repeat biopsy targeting skin and fascia was performed and it demonstrated fascial and subfascial endomysial infiltration by metastatic signet-ring cell adenocarcinoma (Fig 1C). Repeat gastric biopsy, undertaken without obvious mucosal lesion, confirmed the presence of human epidermal growth factor receptor 2–negative gastric adenocarcinoma. She commenced palliative chemotherapy with oxaliplatin and capcitabine, with initial serologic and clinical response. Chemotherapy was discontinued after 6 months because of deteriorating performance status, and the patient died 5 months later.

Discussion

Linitis plastica has been reported in signet-ring cell adenocarcinoma of the GI tract and bladder.[1,2] It was first coined in 1854 by William Brinton to describe the leather bottle shape of the stomach.[3] Over time, the term linitis plastica has been extended to include macroscopic thickening and reduced elasticity of any hollow organ.[3] In this case, inelasticity of the patient's fascia resulted in limb stiffness, which manifested as a

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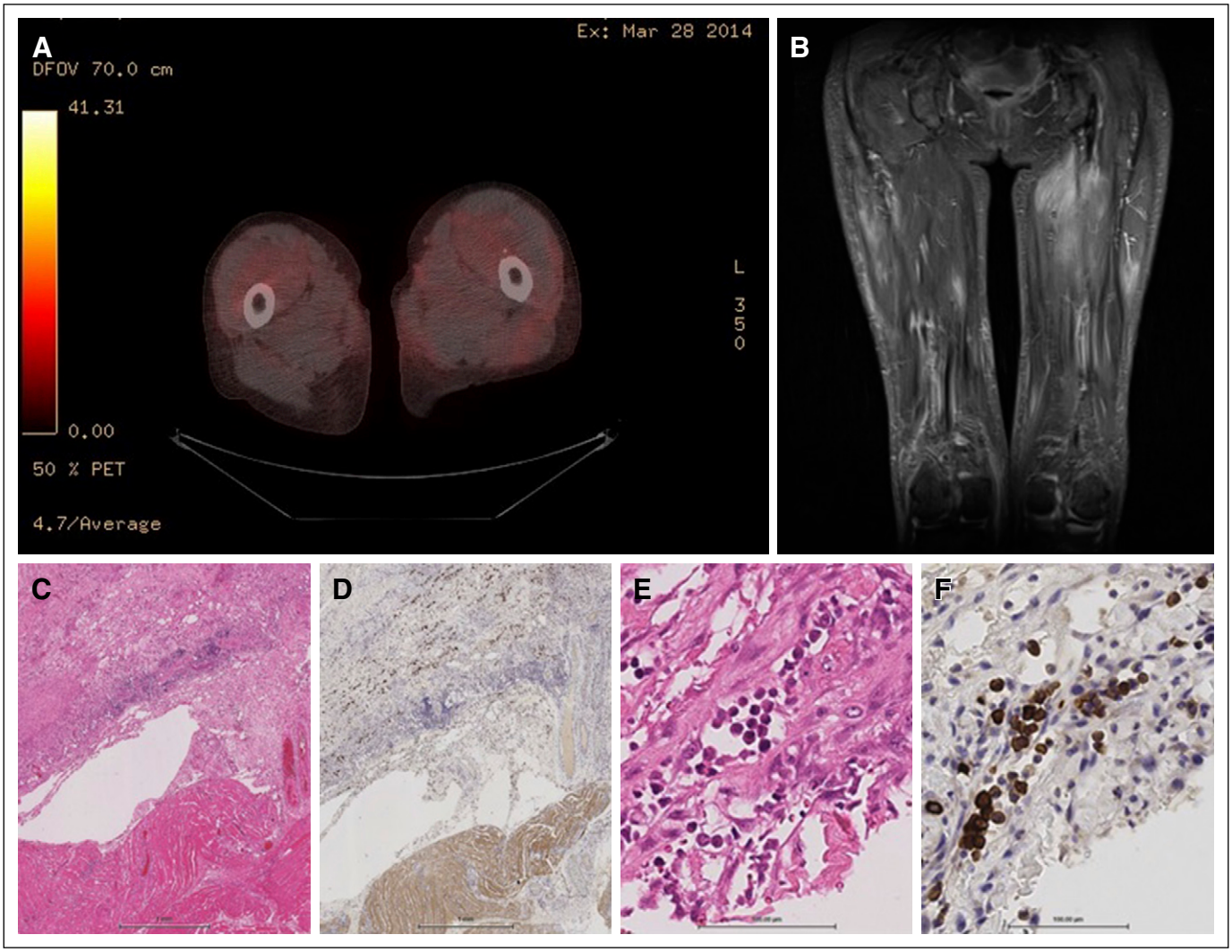


Fig 1. (A) Positron emission tomography showing bilateral lower-extremity myositis. (B) Magnetic resonance imaging of femurs showed extensive muscle edema in the adductor and quadriceps musculature bilaterally. (C–E) Biopsy of skin, fascia, and muscle showing desmoplastic thickening of the fascia with marked chronic inflammation and infiltrating signet-ring cells, (F) highlighted with immunohistochemical staining for AE1/3 cytokeratin.

tiptoe-walking gait. Lower-limb stiffness in signet-ring cell gastric adenocarcinoma has been attributed previously to a paraneoplastic fasciitis-panniculitis syndrome.[4] Our finding of direct fascial infiltration by signet-ring cells suggests that metastases can have an identical presentation. It is essential to distinguish between a paraneoplastic phenomenon and metastases because different therapeutic strategies are often necessary.

To the best of our knowledge, this case is the first report of linitis plastica causing malignant fasciitis. Linitis plastica is a challenging diagnosis to establish because the submucosal infiltration by signet-ring cells can lead to a normal biopsy.[3] In our case, the patient had a persistently elevated beta HCG. This finding has been described in gastric adenocarcinoma and is a potential indicator of an underlying linitis plastica.[5] Signet-ring cell metastases to fascia should be suspected in cases of limb stiffness without pyramidal neurologic signs, especially in the setting of an elevated beta HCG. Currently, linitis plastica refers to infiltration by signet-ring adenocarcinoma cells of the walls of hollow organs.[3] Our case illustrates that this infiltrative process is not restricted to hollow organs and can also involve muscle fascia. More than 150 years after William Brinton's initial description, we propose that the definition of linitis plastic be broadened. JOP

Authors' Disclosures of Potential Conflicts of Interest

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AUTHORS' DISCLOSURES OF POTENTIAL CONFLICTS OF INTEREST

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