Case Report

A Rare Case of Primary Skeletal Muscle Lymphoma: The Value of Octreotide Scintigraphy

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Summary
We report a case of non-Hodgkin lymphoma presenting as a painless mass of the quadriceps femoris muscle that was detected by a somatostatin analogue (octreotide) scintigraphy. We review the few reported cases of primary muscular lymphoma and discuss the potential value of octreotide imaging as a new diagnostic tool.

Key words
Lymphoma, Muscle, Somatostatin, Octreotide, Scintigraphy

INTRODUCTION
Extra-nodal presentation of lymphoma is well recognized (1) but primary skeletal muscle involvement is exceptional, with less than 30 cases being reported (2-14).

Here, we describe a case of primary non-Hodgkin's lymphoma of the quadriceps detected by a positive somatostatin analogue (octreotide) scintigraphy, that labels somatostatin receptor-bearing tumours. We review the literature on primary muscular lymphoma and emphasize the potential value of octreotide scintigraphy in diagnosis, staging and follow-up of the disease.

CASE REPORT
A 51-year-old woman presented with a 3-month history of a painless mass of the left anterior thigh. The patient denied constitutional symptoms such as weight loss, night sweats or fever. Clinical examination was otherwise unremarkable. In particular, the liver, the spleen and the lymph nodes were not enlarged. Blood tests revealed a raised ESR (70 mm/h), a massive IgM polyclonal hypergammaglobulinaemia (31 g/L) and an elevated eosinophil count (2370/ml). The haemoglobin level, the platelet count, the muscle enzymes (creatine kinase, aldolase, lactate dehydrogenase) and the liver tests (ALT, AST, alkaline phosphatase) were normal. Serological tests for HIV and parasitic infections were negative. Magnetic resonance imaging (MRI) of the thighs revealed a 15-cm soft-tissue mass infiltrating the left quadriceps femoris muscle but sparing the femoral blood vessels (Figs. 1 and 2A). An 111In-labelled octreotide scintigraphy showed an intense abnormal uptake in the left anterior thigh (Fig. 3A). Gallium scintigraphy was not performed. Histologic and immunohistochemical analyses of a percutaneous biopsy specimen disclosed a low-grade malignant B-cell lymphoma, with features analogous to those commonly observed in MALT (mucosa-associated lymphoid tissue) -deriving extra-nodal lymphomas (Fig. 4). These features include a diffuse or peri-follicular pattern of infiltration and a centrocytoid or monocytoid appearance of the predominant small lymphoid cells (15). Lymphoma cells stained positively for LN2, MB2 and LCA but were negative for CD3, MT1 and UCHL1. A careful search for other lymphomatous sites was negative. In particular, the bone marrow biopsy was normal. Chest X-ray and CT images of the chest and the abdomen failed to show significant lymphadenopathy. The patient was given local radiotherapy on the left thigh (40 Gy) which induced a complete clinical remission. Post-treatment octreotide scintigraphy (Figure 3B) showed a markedly decreased uptake of the tracer and post-treatment MRI (Figure 2B) confirmed the disappearance of the tumoural mass. However, hypergammaglobulinaemia and eosinophilia persisted. Eighteen months after diagnosis, the patient complained of fatigue and exertional dyspnoea. Chest X-ray showed enlarged hilar and mediastinal nodes. The bronchial mucosa was grossly abnormal at fibroscopic examination.

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Fig. 1: Magnetic resonance imaging. Comparative coronal T1-weighted image obtained after intravenous contrast material injection depicts an intermediate signal intensity mass in the left anterior thigh. and biopsy specimens confirmed the lymphoma relapse. Octreotide scintigraphy was not repeated. The patient was given chemotherapy.

DISCUSSION

Muscular involvement in lymphoma is relatively rare, occurring in only 1.4% of cases in a large necropsic series, usually in the setting of widespread disease (16). Primary muscular lymphoma is much more exceptional: in a review of 7000 lymphoma cases, Travis et al. (11) reported only 8 patients with primary muscle disease. The few reported cases in immunocompetent hosts (2-14) are summarized in Table 1. Recently, a number of additional primary cases has been described in association with HIV infection (17, 18). In AIDS, muscle lymphoma accounts for 8.8% of non-Hodgkin lymphomas, of which 80% develop primarily in the muscle (18).

Primary muscular lymphoma is always a diagnostic challenge for the clinician and the pathologist. Clinically, it usually presents as a painless soft-tissue mass of the thoracic wall or the extremities, the thighs and upper arms being the most common sites (Table). The rheumatologist and orthopaedic surgeon should therefore include lymphoma in the differential diagnosis of a peripheral muscular mass. In rare cases of pelvic muscle involvement such as the psoas, the tumour may cause hydronephrosis or renal failure (13). Computerized tomography and MRI imaging are of great value for the detection of deeper muscular structure involvement but modern imaging techniques do not preclude the need for a biopsy procedure since radiological findings are nonspecific.

Histological diagnosis requires much awareness from the pathologist since lymphoma may be confused with other small-cell tumours occurring in muscles, such as rhabdomyosarcoma and metastatic oat-cell carcinoma (4). Immunohistochemistry techniques that detect immuno-