Diffuse Skeletal Muscles Uptake of $[^{18}F]$ Fluorodeoxyglucose on Positron Emission Tomography in Primary Muscle Peripheral T-cell Lymphoma

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Abstract

A 40-year-old man presented with weakness of neck extensor muscles. Cervical magnetic resonance imaging showed high-intensity areas in muscles of the left lateral cervical region on T2-weighted images. Fluorodeoxyglucose-positron emission tomography scan demonstrated striking fluorodeoxyglucose uptake by multiple skeletal muscles of the neck, chest, and abdominal region. Muscle biopsy demonstrated peripheral T-cell lymphoma, unspecified. The diagnosis was primary skeletal muscle peripheral T-cell lymphoma. Primary skeletal muscle non-Hodgkin’s lymphoma of T-cell immunophenotype is extremely rare and fluorodeoxyglucose-positron emission tomography demonstrated striking fluorodeoxyglucose uptake in multiple skeletal muscles and served as a quite useful modality for the diagnosis of this patient.

Key words: primary skeletal muscle lymphoma, non-Hodgkin lymphoma, peripheral T-cell lymphoma, fluorodeoxyglucose on positron emission tomography, skeletal muscle

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Introduction

Primary skeletal muscle non-Hodgkin’s lymphoma is extremely uncommon and it is associated with a poor prognosis (1). Additionally, the majority of these lymphomas demonstrate B-cell immunophenotype (1). We report a case with primary skeletal muscle non-Hodgkin’s lymphoma of T-cell immunophenotype, that showed slow progression, characterized by striking fluorodeoxyglucose uptake involving multiple skeletal muscles on positron emission tomography scans.

Case Presentation

In January 1996, a 27-year-old man had swelling of the right side of the labia and oral mucosa. In April 1996, the patient visited a hospital and was diagnosed with chronic lymphadenitis by a biopsy of the oral mucosa. Oral steroid therapy improved the symptoms. In July 1996, the patient had left ptosis, and left facial weakness. Steroid therapy again improved the symptoms. In September 1998, the patient had diplopia and bilateral facial weakness (manual muscle test (MMT): 1/1). The symptoms were not improved by steroid therapy and remained constant. In May 2000, the patient developed weakness of the neck extensor (MMT: 4/4). After several months, the symptoms disappeared spontaneously. Thereafter, the patient remained free of any new symptoms for the subsequent 9 years.

In May 2009, at the age of 40 years, weakness of neck extensor appeared again. The patient was admitted to our
magnetic resonance imaging (MRI) showed high-intensity areas in the muscles of the left lateral cervical region on T2-weighted images. The muscles of other regions did not demonstrate abnormal findings on MRI. Fluorodeoxyglucose-positron emission tomography demonstrated striking fluorodeoxyglucose uptake by multiple skeletal muscles of the neck, chest, and abdominal region, but not of the lower extremities (Fig. 1A, 1B). Maximum standardized uptake value of the left trapezius muscle was 10.32. Ga scintigraphy showed no abnormal uptake. Computed tomography of the whole body revealed no evidence of lymphadenopathies, tumor infiltrations, or masses. There were no disease abnormalities involving the osseous structure. Bone marrow biopsy revealed a normocellular marrow with trilineage hemopoiesis and no evidence of lymphoma. Biopsy of the left trapezius muscle demonstrated massive infiltration of atypical lymphocytes which varied in size and clusters of epithelioid histiocytes destroying muscle bundles (Fig. 1C). Immunohistochemical staining revealed that the atypical lymphocytes were positive for CD3 and CD8, but negative for CD4, CD20, CD56, CD79a, and EBER. Histopathologic and immunocytochemical characteristics were consistent with those of peripheral T-cell lymphoma, unspecified (PTCL-U). The final diagnosis was established as primary skeletal muscle peripheral T-cell lymphoma. Chemotherapy with 6 cycles of cyclophosphamide, doxorubicin, vincristin and prednisone completely resolved the weakness of the left pectoral girdle, but it did not improve the eye movement or facial weakness. On serum analyses, CK and IL2R were decreased to the normal range. Fluorodeoxyglucose-positron emission tomography and magnetic resonance imaging improved after chemotherapy. The maximum standardized uptake value of the left trapezius muscle was reduced. After 6 months of chemotherapy, the symptoms remained the same.

Figure 1. Fluorodeoxyglucose-positron emission tomography (PET) and biopsy of the muscle. (A) (B) Fluorodeoxyglucose-positron emission tomography (PET): Fluorodeoxyglucose-positron emission tomography (PET) demonstrated striking fluorodeoxyglucose uptake involving the skeletal muscles of the neck, chest, and abdominal region. The maximum standardized uptake value of the left trapezius muscle was 10.32. (C) Biopsy specimen of the muscle: Muscle biopsy of the left trapezius muscle demonstrated massive infiltration of atypical lymphocytes of varying sizes and clusters of epithelioid histiocytes destroying muscle bundles.
Discussion

A 40-year-old man was diagnosed with primary skeletal muscle peripheral T-cell lymphoma. Multiple neurologic deficits had appeared for 13 years from the initial symptoms. The patient had been diagnosed with idiopathic or unknown disease because there was no specific finding. Although not definite, if those symptoms were related to lymphoma, this case showed very slow progression. Fluorodeoxyglucose-positron emission tomography demonstrated striking fluorodeoxyglucose uptake by multiple skeletal muscles and was useful in detecting disseminated lymphoma in the skeletal muscles.

Primary skeletal muscle non-Hodgkin’s lymphoma is a rare manifestation of non-Hodgkin’s lymphoma, accounting only for 0.1% of all lymphomas (1). The frequency of primary skeletal muscle non-Hodgkin’s lymphoma (7%) is increased in patients with Acquired Immune Deficiency Syndrome (AIDS)-associated lymphomas (2). It is difficult to differentiate between primary skeletal muscle lymphoma and secondary muscle involvement from bone lymphoma. The following criteria is used to diagnose primary muscular lymphoma: 1) histopathology proven non-Hodgkin’s lymphoma of the muscle; 2) the absence of systemic/nodal disease at initial presentation; and 3) the presence of a soft tissue mass with normal adjacent marrow or marrow disease much less extensive than soft tissue (3). The present case was diagnosed as primary skeletal muscle non-Hodgkin’s lymphoma because these three 3 criteria were fulfilled. In cases of primary skeletal muscle non-Hodgkin’s lymphoma the lower extremities and pelvic region are most commonly affected (4). Additionally, the majority of reported cases had B-cell immunophenotypes and behaved in an aggressive biologic manner (5). T-cell neoplasms constitute 10% to 12% of all non-Hodgkin’s lymphoma (6). PTCL-U represents the most common subtype of T-cell lymphomas, manifesting most often as a nodal disease (7). Extranodal PTCL-U is a rare presentation among neoplasms. Some cases of primary skeletal muscle T-cell lymphomas have been described in the literature (8-12). Furthermore, in a few reported cases fluorodeoxyglucose-positron emission tomography demonstrated striking fluorodeoxyglucose uptake in multiple skeletal muscles.

Fluorodeoxyglucose-positron emission tomography scans demonstrate multiple sites of hypermetabolic activity within musculature. A recent study suggests that fluorodeoxyglucose-positron emission tomography scans are sensitive to detect T-cell lymphomas in all regions except for the skin (13). Fluorodeoxyglucose-positron emission tomography is useful in aiding the diagnosis and staging of lymphomas of aggressive subtypes, including PTCL-U (14). However, other reasons for skeletal muscle fluorodeoxyglucose uptake may stem from a variety of factors such as physical exertion, talking, and chewing as normal physiological phenomena (15). As for pathological phenomena, elevated fluorodeoxyglucose muscle may be associated with anxiety-induced neck muscle spasm (15), accessory muscle use in respiratory distress (16), intercostal and diaphragmatic activity in chronic obstructive pulmonary disease (17), abdominal muscle contraction in intractable vomiting (18), sepsis-induced shivering (19), hypoglycemia (20), infection, and inflammation of the muscle tissue that increases glycolytic activity (21, 22). In the present case, the patient did not satisfy these conditions. Skeletal muscle fluorodeoxyglucose uptake was induced by T-cell lymphoma, which histopathologically proved PTCL-U.

In conclusion, primary skeletal muscle non-Hodgkin’s lymphoma of T-cell immunophenotype is extremely rare. Fluorodeoxyglucose-positron emission tomography demonstrated striking fluorodeoxyglucose uptake in multiple skeletal muscles, and served as a useful method for diagnosing this case.

Informed consent was obtained from this patient.

The authors state that they have no Conflict of Interest (COI).

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