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Clinical note

FDG PET/CT findings in rare sarcomas

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ABSTRACT

The role of FDG PET/CT in management of soft tissue and bone sarcomas has been described in many studies up-to-date. However, contribution of PET/CT to diagnosis and treatment in some types of sarcomas that are seen with low incidence has not been identified properly yet. Clear cell sarcoma, synovial sarcoma of chest and myxoid lyposarcoma are rare types of sarcomas. We aimed to describe the FDG uptake patterns of these rare tumors and find out the role of FDG PET/CT in management of disease.

Hallazgos de FDG PET/CT en sarcomas raros

Palabras clave:
Sarcoma de células claras
Liposarcoma
Sarcoma sinovial
FDG PET

RESUMEN

Hasta la fecha se ha descrito en muchos estudios el papel de la FDG PET/TAC en el manejo de los sarcomas de tejido blando y hueso. Sin embargo, la contribución de la PET/TAC al diagnóstico y al tratamiento de muchos tipos de sarcomas con baja incidencia no ha sido identificada aún. El sarcoma de células claras, el sarcoma sinovial en tórax y el liposarcoma mixoide constituyen tipos raros de sarcomas. Tratamos de describir las pautas de captación de la FDG en estos tumores raros y de averiguar el papel de la FDG PET/TAC en el manejo de la enfermedad.

Introduction

Sarcomas are a heterogeneous group of malignant mesenchymal tumors consisting of bone and soft tissue sarcomas. Sarcomas can develop from various tissue types such as bone, cartilage, connective tissue, muscle, fat, nerves and vessels. There are many different histological types with a large variation in the degree of malignancy and aggressiveness of the different subtypes. In recent studies, the contribution of FDG PET/CT for defining different types of sarcomas has been reported with high values of sensitivity, specificity and accuracy for detecting lymph nodes and distant metastasis.[1] We report FDG PET/CT findings of 3 cases with rare types of sarcomas: clear cell sarcoma, synovial sarcoma of the thorax and myxoid liposarcoma.

Case reports

Case 1

A 16-year-old girl presented with pain and lump on the anteriorright knee. The MRI revealed a mass lesion obliterating the infrapatellary bursa and surrounding the patellary tendon. A tru-cut biopsy was performed from the lesion and histopathology demonstrated S-100 positive and HMB-45 positive tumor cells defining clear cell sarcoma of tendons and aponeuroses (CCSTA). The expert pathologist interpreted that since CCSTA and malignant melanoma possess the similar morphologic and immunohistochemical findings, the patient should be evaluated for systemic involvement of the disease. The patient had undergone a FDG PET/CT scan. The mass lesion on the right knee in infrapatellary region was observed with high FDG avidity (SUVmax = 12.4). There was no other pathological focus of FDG uptake related to the primary disease. Physiologic brown fat activation in bilateral supraclaviculary and axillary regions was observed (Fig. 1). The patient was considered as CCSTA and had taken neoadjuvant radiation therapy before the resection of the tumor.

Case 2

A 43-year-old woman with a history of multiple surgical operations in thoracal region with diagnosis of synovial sarcoma was referred to PET/CT after 31 cycles of chemotherapy. Her first operation was six years ago containing partial rib resection from right hemithorax and the pathology had revealed leiomyoma. After two years the patient presented with multiple lesions in right lung, mediastinum and axilla, and the pathology showed synovial sarcoma this time. After a wide

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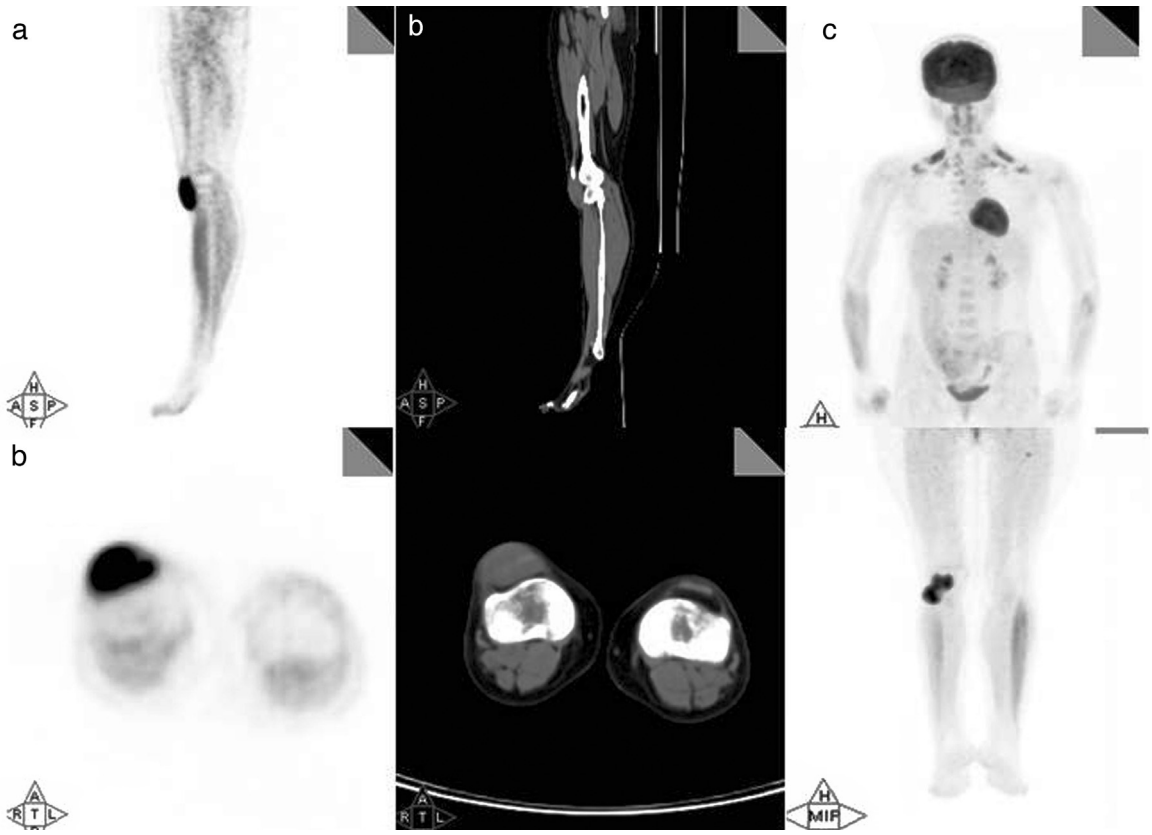

Fig. 1. FDG avid lesion in infrapatellary region (a, b). No other pathologic hypermetabolic focus in whole body (c).

resection of the tumor the patient received radiation therapy and chemotherapy. Her last operation was two years ago and metastatic pulmonary nodules were resected. After a long term chemotherapy the patient again presented with mass lesions in right hemithorax. In FDG PET/CT examination we observed multiple gross mass lesions in right hemithorax invading the muscles and lipomatous tissue showing high FDG uptake (SUVmax = 10.7) heterogenously with hypometabolic areas in cystic components of tumors. A similar mass was observed on the right posterior wall of abdomen originated from the right diaphragmatic crus (Fig. 2). A wide resection of the ribs and surrounding tumoral tissue in right hemithorax and posterior abdominal wall was performed and histopathology demonstrated synovial sarcoma involvement.

Case 3

A 13-year-old boy presented with two-year-history of surgery, chemotherapy, radiation therapy and follow-up for myxoid liposarcoma in left hemithorax. He had received last cures of chemotherapy and radiotherapy about 8 months ago and afterwards a wide resection of tumor from left lung and mediastinum was performed. He was referred to PET/CT imaging for control. In FDG PET/CT scan we observed multiple gross hypodense mass lesions similar to each other showing mild FDG uptake (SUVmax = 2.6) localized in anterior mediastinal and left paramediastinal regions, left axilla, and also in anterior abdominal region and posterior of the right kidney and in pelvis (Fig. 3). The disease was in progression and it spread to abdomen and pelvis from thorax.

Discussion

Although the use of FDG PET/CT in sarcomas has been reported in several studies up-to-date, most of them include a mixed population of tumor types. For identifying the diagnostic value of PET/CT in various histological types of sarcomas, more prospective studies having large number of patients are necessary. We reported three cases presenting rarely seen types of soft tissue sarcomas.

Clear cell sarcoma of tendons and aponeuroses (CCSTA) is a rare type of tumor accounting for less than 1% of soft tissue sarcomas. The majority of the patients are children and young adults. The tumor involves the extremities in 90–95% of cases and foot and ankle are the most common primary sites, accounting for 33–43% of cases.[2] Tumor size and the presence of necrosis are statistically significant predictors of prognosis. Surgery is the elected treatment in most of the cases.[3] Primary clear cell sarcoma and metastatic malignant melanoma represent similar histopathological features.[2] Thus, detection of the whole body for tumor involvement is an important way of discriminating malignant melanoma from the local soft tissue involvement of CCSTA. FDG PET/CT findings of the tumor are reported in very few cases up-to-date and in a recent report high FDG uptake was observed in two cases as in our case.[4] Whole body imaging with FDG PET/CT contributes to diagnosing and staging of CCSTA.

Synovial sarcomas usually originate from paraarticular soft tissues of extremities in young adults. Primary synovial sarcoma of thorax is a rare tumor and can originate from lung parenchyma, pleura, mediastinum or chest wall. A complete resection of the tumor is the best treatment and response to chemotherapy and radiation therapy is poor. Polverosi et al.[5] have recently reported