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Mazabraud's Syndrome mimicking metastases on FDG PET/CT in a patient with colon cancer

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Abstract

Mazabraud's, a syndrome consisting of fibrous dysplasia and soft tissue myxomas, is a rare disorder with less than 100 cases reported in the literature. Appearance of lesions in Mazabraud's syndrome on FDG PET/CT scan in an oncological scenario can be misinterpreted as malignancy. We present the case of a 69-year-old woman, diagnosed with metastatic colon carcinoma and suspected osseous as well as soft tissue metastases, who was later found to have concomitant Mazabraud's syndrome. This case describes the diagnostic dilemma associated with this unusual entity and importance of differentiating benign and malignant etiologies on FDG PET/CT scan.

Keywords

Mazabraud's syndrome; FDG PET/CT scan; colon carcinoma; myxomas

Considering known history of colon carcinoma and reports from prior imaging describing these lesions as metastases, this study could be easily misinterpreted as representing widespread metastatic disease. The scan findings are however, typical of Mazabraud's syndrome, characterized in this case by mildly avid rounded intramuscular soft tissue lesions probably myxomatous in etiology and osseous disease consistent with aggressive polyostotic fibrous dysplasia. Mazabraud's syndrome is a benign disorder, described by Mazabraud et al. only in 1967 (1). So far, less than 100 cases of Mazabraud's syndrome have been reported in the literature (2–5). It is more frequent in women and often remains asymptomatic. While lesions are benign, complications such as skeletal deformities and fractures may occur. The treatment of Mazabraud's syndrome is dependent on the symptomatology and extent of the lesions, predominantly includes conservative management for myxomas and bisphosphonates for fibrous dysplasia. Surgical intervention is indicated only for severe and progressive osseous disease with persistent pain, skeletal deformity, or impending fracture. Sometimes, excision may be considered for myxomas if pain or pressure symptoms develop.

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Fibrous dysplasia, an uncommon fibro-osseous disease, is seen in 7% of benign bone tumors and 2.5% of all bone tumors. At imaging, the osseous lesions demonstrate expansile remodeling and diffuse cortical thinning, maintaining an intact overlying periosteum and a defined rim of reactive sclerosis (6). Polyostotic fibrous dysplasia can sometimes, closely mimic the appearance of widespread distant bone metastases (7). Fibrous dysplasia may be FDG-avid (8). Malignant degeneration is uncommon, reported in only a few cases of fibrous dysplasia. Soft tissue myxomas are a benign connective tissue with mucinous components. The mucinous components result in a lower than muscle attenuation of CT. Myxomas may be FDG-avid (9).

Important clue to the benign nature of the FDG-avid osseous lesions is the knowledge of the propensity of colon carcinoma metastases. Colon carcinoma metastasizes mainly to the liver, lung, peritoneum, and rarely to the bone. Skeletal metastases occur in only 5–10 % of cases and usually after widespread metastatic disease (10). The distribution of osseous lesions was predominantly in the extremities, sparing the spine, while osseous metastases normally have a predilection for the axial skeleton with less involvement of the appendicular skeleton.

Our case, highlights the potential to misdiagnose Mazabraud's, a benign syndrome, as malignancy on FDG PET/CT. Physicians need to be aware of this rare syndrome to help prevent misdiagnosis of this benign condition.

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Figure 1 -

A 69-year-old woman with mucinous adenocarcinoma of the ascending colon, status post right hemicolectomy, was referred for restaging FDG-PET/CT scan to evaluate the extent of disease. She had received multiple lines of chemotherapy over the past seven years for hepatic metastases and suspected osseous as well as soft tissue metastases. (A) FDG MIP demonstrates multiple abnormal FDG-avid foci overlying the upper and lower extremities and pelvis. (B) Axial PET, (C) Axial CT, (D) Coronal PET, and (E) Coronal CT through the pelvis demonstrate multiple foci correspond to expansile osseous lucencies with cortical thinning (SUV range 2.7–9.7). (F) Axial PET and (G) Axial CT through the lower extremities demonstrate a lower than muscle attenuation lesion involving the right proximal vastus with SUV 1.6. Additional lower than muscle attenuation lesions were seen (not shown).