Multiple myeloma is a disease characterized by bone marrow infiltration with neoplastic plasmatic cells. It represents approximately 10% of all hematologic malignancies and occurs mainly between 40 and 80 years.¹

Extradural manifestations of multiple myeloma are diagnosed in the 10–16% of patients. Extraosseous disease is more common in young patients with more aggressive subtypes (such as nonsecretory myeloma and myeloma IgD) and after a stem cell transplant. In patients with multiple myeloma, autopsy series detected reticuloendothelial system as the most affected, represented liver, spleen and lymph node of a 31%, 29% and 23%, respectively.¹

We report the case of a 60-year-old woman with pathological right humerus fracture. Bone scintigraphy show right humerus, skull involvement, left humerus, left eight costal arch, L3 vertebral body, right sacral ala and left femur, compatible with myelomatoid disease (Fig. 1A). After laboratory confirmation, the patient was treated with chemotherapy (Bortezomib [Velcade®] and dexamethasone) and autologous hematopoietic stem cell transplantation, with poor response. Bone scintigraphy performed in order to monitor treatment response confirmed bone progression (Fig. 1B), being accompanied of general discomfort and pruriginous skin rash. For these reasons, an ¹⁸F-FDG PET/CT was performed in our center. This study not only allows better assessment of bone disease but also diagnoses extradural disease highlighting the presence of metabolically active peritoneal mass, mainly in the left upper quadrant and pelvis (Fig. 2).

There has been an increased incidence of extramedullary myeloma, probably related to improved diagnosis with MRI and PET studies as well as prolonging patient survival due to new chemotherapeutic agents and autologous hematopoietic stem cell transplantation. In a recent meta-analysis the sensitivity and specificity of ¹⁸F-FDG PET/CT in the detection of bone marrow disease were 61.1% and 94.1%, respectively; whereas detection of extramedullary disease was 96.0% and 77.8%, respectively.²

In our case, the diagnosis of extraosseous disease has allowed a combined therapy with thalidomide, cyclophosphamide and dexamethasone, which is proved to be effective for refractory myeloma.³ The patient shows decreased abdominal bloating and lower limb edema, so monitoring by ¹⁸F-FDG PET/CT was performed. The study allows assessment of therapeutic efficacy, with virtually complete response of the peritoneal mass, variable changes in bone lesions and the detection of a new metabolically active tumor muscle lesion (Fig. 3). Given this evolution is decided to continue with therapy maintenance, but the appearance of pancytopenia requires discontinuation treatment.

Therefore, ¹⁸F-FDG PET/CT has significant prognosis in multiple myeloma, enabling early detection of extramedullary disease and facilitates monitoring in a single scan of medullary and extramedullary involvements.
Fig. 1. (A) Whole body $^{99m}$Tc-MDP bone scintigraphy. Pathological uptake was observed in the proximal third of the right humerus including the humeral head, related to pathological fracture. Abnormal active images were located in the skull, left eighth posterior costal arch, L3 upper platform, right sacral ala, middle third of left humerus and mild uptake in the middle third of the left femur, consistent with bone myelomatoid involvement. (B) Bone scintigraphy performed after chemotherapy and autologous hematopoietic stem cell transplantation. In relation to the previous study, remain active images of the proximal right humerus, skull, L3 upper platform, and right sacral ala without significant changes. The slight left femoral uptake is not evident at present. However, it has increased in extent and intensity uptake in left eighth posterior rib and left humeral. There are new pathological uptakes in left humeral head, right scapula and tenth right posterior costal arch, suggesting progression of myelomatoid disease.

Fig. 2. Maximum intensity projection whole body PET, axial CT and fused images. Due to the progression of multiple myeloma $^{18}$F-FDG PET/CT study was performed. Images show multiple metabolically active bone lesions, in medullary location (A), predominantly in diaphysis of long bones; other lesions in spinal structures had nodular sclerosis, and also lytic appearance in the trabecular right iliac bone (C). Extensive peritoneal solid masses with intense and irregular uptake of $^{18}$F-FDG, including: left upper quadrant, located in the lesser sac and the gastrospenic ligament, infiltrating the renal hilum and the splenic flexure of the colon (B). Pelvis: large areas of contact with the posterior wall of the bladder (D).
Fig. 3. Maximum intensity projection whole body PET, axial CT axial and fused $^{18}$F-FDG PET/CT images for the evaluation of therapy response. Marked reduction in metabolic activity and size in the left upper quadrant mass (B) and pelvis (C). There is also an activity reduction of bone lesions (A). However, appears multiple muscular masses and infra and supradiaphragmatic tumor metabolically active implants (D).

References

