CLINICAL IMAGE

Primary solitary extramedullary plasmacytoma progressing to multiple bone plasmacytomas: a rare condition with therapeutic dilemmas

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A 54-year-old woman presented with a pea-sized solid lump in the sacral region, surrounded by a red areola and neither painful nor itchy. A histopathologic examination showed diffuse infiltrate (FIGURE 1A) composed of atypical plasma cells positive for light chains (FIGURE 1B), CD56, CD138 (Supplementary material, *Figure S1*) and negative for CD3, CD20, CD30, CD123, anaplastic lymphoma kinase and κ light chains (Supplementary material, *Figure S2*), with a Ki-67 proliferation index of 50%. The lesion was identified as an extramedullary plasmacytoma. Intact immunoglobulins and free light chains remained within normal limits, and a slightly separated immunoglobulin A zone was revealed on immunofixation. No protein was detected on urinalysis. Bone marrow trephine biopsy showed 4% to 5% of dispersed CD138$^+$ plasma cells with a normal immunohistochemical reaction to light (κ/λ) chains (FIGURE 1C). Fusion positron-emission tomography–computed tomography (PET-CT) revealed a mass with an elevated standardized uptake, infiltrating the ascending colon wall, and an osteolytic lesion in the right ischial tuberosity (FIGURE 1D and 1E).

Given the risk of colon perforation, the patient underwent right hemicolectomy. The colonic lesion was identified as an extramedullary plasmacytoma infiltrating the intestinal muscular layer and focally invading the serosa, without angioinvasion and with negative surgical margins. The patient received radiotherapy to the right ischial tuberosity area (50 Gy in 25 fractions), which resulted in complete hematologic remission (CR). However, PET-CT performed a month later showed disease progression, with a focal increase in ^{18}F-fluoro-ethyl-tyrosine uptake in the sternum, multiple plasmacytomas in the left fourth rib, and a persisting osteolytic lesion in the right ischial tuberosity (FIGURE 1F–1H). After achieving complete remission with 6 cycles of VTD (bortezomib, thalidomide, dexamethasone), the patient underwent tandem autologous hematopoietic

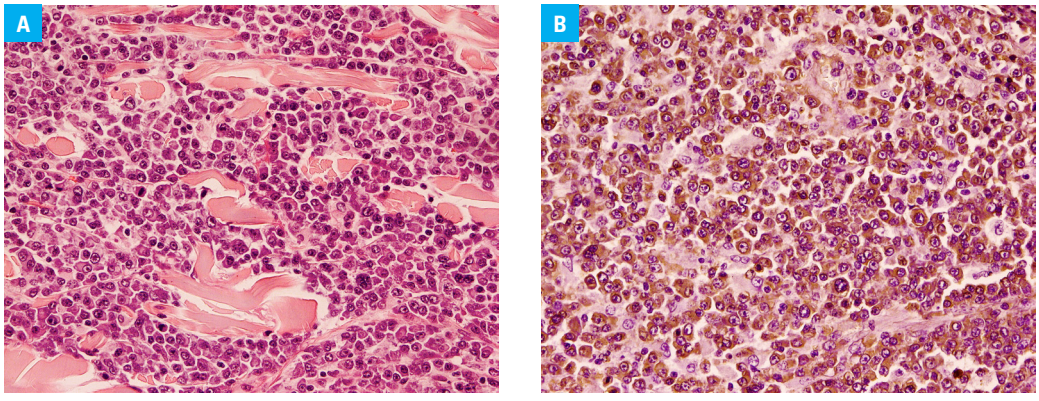

FIGURE 1 A, B – histopathologic specimen obtained from the skin lump: A – plasma cells with eosinophilic cytoplasm and round nucleus with granular chromatin (hematoxylin and eosin staining, ×400); B – positive immunohistochemical reaction to λ light chains (×400).

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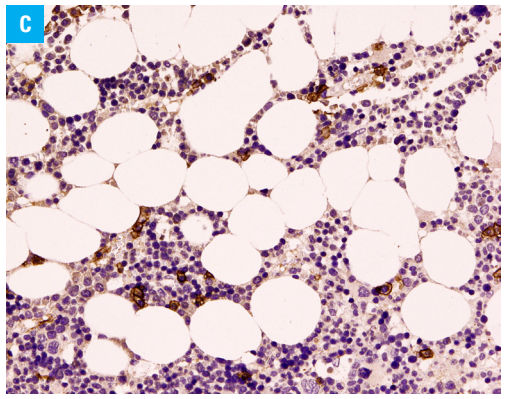

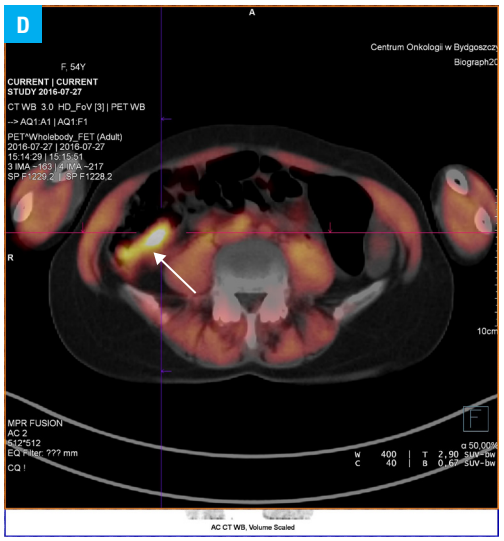

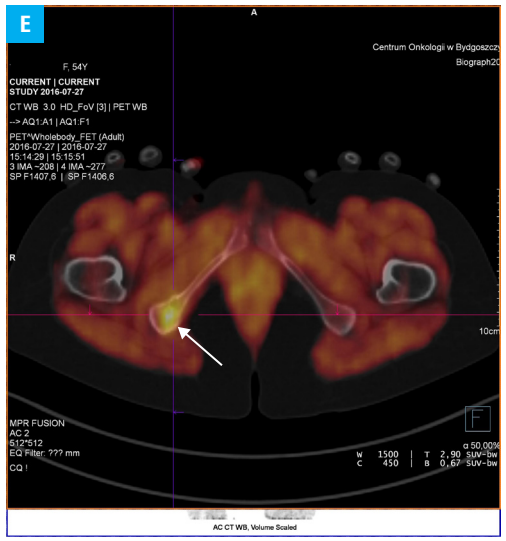

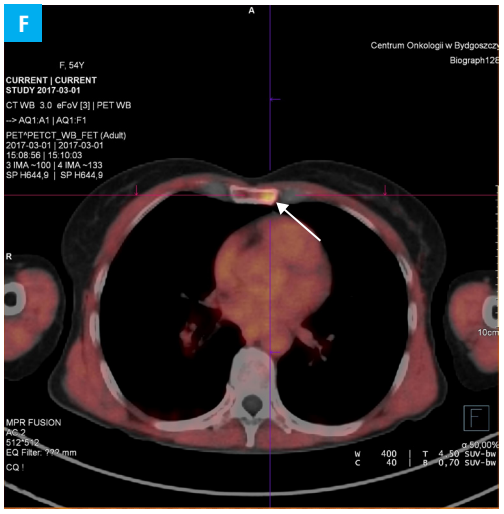

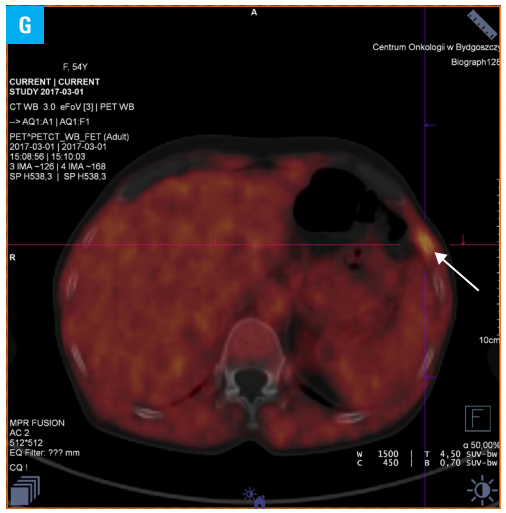

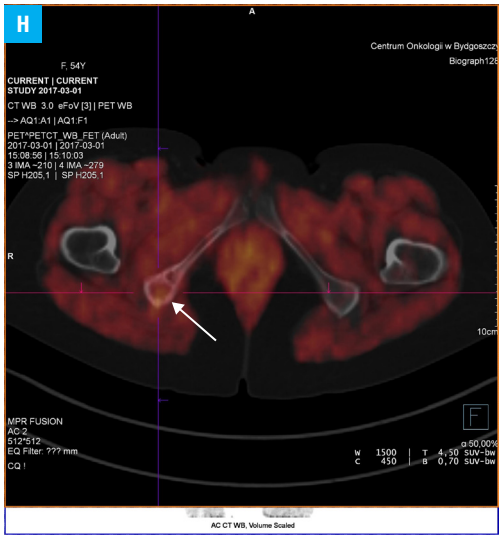

FIGURE 1
C – bone marrow trephine biopsy; positive immunohistochemical reaction to CD138 (×400). **D**, **E**, **F**, **G**, **H** – positron-emission tomography–computed tomography scan in axial plane: **D** – a mass (39×13×42 mm) infiltrating the ascending colon wall; enhanced ^{18}F-fluoro-ethyl-tyrosine uptake (^{18}F-FET; standardized uptake value [SUV], 4.12); **E** – an osteolytic lesion (diameter, 9 mm) in the right ischial tuberosity (arrow); enhanced ^{18}F-FET uptake (SUV, 2.9); **F** – focal increase in ^{18}F-FET uptake (11 × 9 mm; SUV, 3.64) in the sternum, at the level of the left fourth rib attachment (arrow); **G** – focal increase in ^{18}F-FET uptake (SUV, 2.94) in the left fourth rib (arrow); **H** – an osteolytic lesion (16 × 17 mm) in the right ischial tuberosity (arrow); enhanced ^{18}F-FET uptake (SUV, 2.27)

stem cell transplantation (auto-HSCT), followed by consolidation VDT-PACE therapy (bortezomib, dexamethasone, thalidomide, cisplatin, doxorubicin, cyclophosphamide, etoposide). Currently, 6 months after auto-HSCT, she shows no signs of progression.

Solitary plasmacytoma is rare, representing less than 5% of plasma cell malignancies. It may present as a single bone lesion, solitary bone plasmacytoma (SBP), or as a soft tissue mass, extramedullary plasmacytoma (EMP).[1] EMP may develop in any tissue but is primarily found in the head and neck region, gastrointestinal tract, and lungs.[2] SBP is more likely to progress to multiple myeloma (MM) (3-year progression rate, 60% vs 20% for EMP).[3] Solitary plasmacytoma can be diagnosed if biopsy shows clonal plasma cell infiltration of the lesion, with no or minimal (<10%)

bone marrow involvement, normal skeletal survey results, no additional lesions on spinal and pelvic magnetic resonance imaging (or computed tomography), and no features of end-organ damage.[3] Both SBP and EMP are treated locally (fractionated radiotherapy, 40–50 Gy, with at least 2-cm margin of normal tissue).[1-4] Surgery, followed by radiotherapy, should be considered in patients with pathological fractures, large and well-defined soft tissue lesions, or high risk of complications.[1] Chemotherapy, using the same protocols as for MM, including high-dose chemotherapy followed by auto-HSCT in younger patients, is indicated in refractory disease or relapse.[4] However, in some countries, especially those in Central and Eastern Europe, inadequate access to advanced antimyeloma therapy, including auto-HSCT, may limit treatment success.[5]

SUPPLEMENTARY MATERIAL Supplementary material is available with the article at www.pamw.pl.

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