Rare multifocal extramedullary manifestation and progression of multiple myeloma after ASCT/h4

Penka Ganeva[1], Yavor Petrov[1], Branimir Spasov[1], Tihomir Dikov[1], Georgi Arnaudov[1], Margarita Guenova[1], Lidia Garcheva[1], Georgi MIhaylov[1], Rumen Simeonov[2]

[1]*BMT, Specialised Hospital for Active Treatment of Hematological Diseases, Bulgaria*

[2]*Urology department, Specialised Hospital for Oncology, Bulgaria*

Introduction

Extramedullary disease is an uncommon manifestation in multiple myeloma and can either accompany newly diagnosed disease (7-15%) or develop with disease progression or relapse (6-20%). The most common sites for extramedullary disease at diagnosis are skin, nasopharynx, larynx and upper respiratory tract. However gastrointestinal tract, pleura, skin, liver, lymph nodes are usually involved in the context of dissemination and are still rare. Regardless of therapy, extramedullary disease was associated with shorter progression-free and overall survival**.** We describe a case of rapid and unusual (genitourinary tract, orbits) extramedullary progression of MM after ASCT despite a concomitant medullary response**.**

Case presentation

A 62-year-old man presented with a back pain in thoracolumbar spine. Spinal MRI showed changes at Th7 vertebra with deformation of the body and tumour mass. The histology confirmed plasmocytoma with CD19(-);CD138(+);CD56(+) plasmocytic population. In siru hybridization was with kappa (-); lambda (+). Stage ISS2 MM with IgG lambda + FLC lambda (35.2% -33.8g/l) in the serum was diagnosed. A bone marrow aspiration showed 15% of plasma cells, immunophenotyping revealed a population of cells (3.36%) with aberant phenotype - CD19(-)CD45(-)CD56(+)CD27(+-)CD28(+)CD200(+). Cytogeneits 46XY, FISH confirmed IgH -MMSET (+) changes. CT of the body revealed no other extramedullary involvments. No bone lessions.

He achieved VGPR after 4 cycles of chemotherapy C-VEL/DEX type and ASCT. Two months later he was with pain, edema and swelling of the right eye with blurred vision and a split images. MRI of the brain and the orbits showed tumor formation (42/28/39 mm) of right orbita. The histology of the tumor and immunocytochemistry showed plasmocytoma. PET CT showed multiple dissemination of extramedullary tumours in the bones, soft tissue of hypoderma, peritoneal masses and residual tumor in the right orbit. Bone marrow biopsy/ aspiration non confirmed plasma cells. No paraprotein in the serum and urine. He received local radiotherapy of the orbit and chemotherapy, but 2 months later new tumour mass was determined in the pelvis (left), associated with the prostate gland enlargement and infiltration in the bladder, the left seminal vesicles and soft tissue lesion of the bladder. Enlarged parailiac lymph nodes and diffuse metastatic process in the hips and both femurs. The biopsy of testis confirmed plasmoblastic infiltration, prostate biopsy showed again plasmoblastic infiltration with CD138(+)CD56(+)MCK(+). PSI normal. At this moment he was with increased of serum/ urine paraprotein, beta2microglobulin 5.6mg/l, anemia, trombocytopenia. One month later he died.

Conclusions

This case illustrates the rare and rapid progression of multifocal unusual extramedullary presentation. At the same time he was with differential response of extramedullary compared to intramedullary myeloma after standad therapy and a very short survival. This is one of the few cases reported of a patient with rapid extramedullary progression of disease despite concomitant medullary response to standard therapies including novel therapeutics and ASCT.