Whole-body MR-imaging allows to differentiate indolent from aggressive systemic mastocytosis

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Introduction:

Mastocytosis is a group of rare clonal disorders caused by the presence of too many mastocytes and CD34+ mast cell precursors in a person's body ¹. Systemic mastocytosis (SM) involves the extracutaneous tissues and can be further subdivided into indolent systemic mastocytosis (ISM), smoldering systemic mastocytosis (SSM) and aggressive systemic mastocytosis (ASM). No larger studies on MRI-findings in patients with SM exist so far. Therefore the aim of this prospective study was to evaluate if whole-body (wb) – MRI allows to detect typical pattern of organ involvement in SM and to differentiate between different WHO-classes of SM based on the wb-MRI findings.

Methods and Materials:

In this IRB approved study, 36 consecutive patients (20F, 16M, age range 31-80 years) with clinically proven mastocytosis underwent a whole-body (wb) MR-exam at 1.5T (Siemens Avanto) including coronal wb-T1w-TSE imaging (TR/TE – 579/11ms, 1.6x1.6x6mm³ spatial resolution), coronal wb-STIR-imaging (TR/TE – 5220/105ms, 2.2x1.0x6mm³ spatial resolution) in four single steps, sagittal T1 and TIRM sequences of the spine and dynamic contrast-enhanced T1w-VIBE imaging (TR/TE 5.15/1.87ms, 1.5x1.0x3mm³ spatial resolution) of the abdomen using a single dose of 0.5M gadoterate (Dotrarem). Image evaluation done by one radiologist included the pattern and extent of pathologic bone marrow signal in the spine (normal, activated bone marrow, sclerotic bone marrow) and the extremities, osteolyses, the presence of ascites, pathologic lymph nodes, hepatosplenomegaly and focal organ lesions. The clinical diagnosis of ASM and ISM was based on the current WHO-criteria. T-tests were performed for statistical analysis.

Results:

17 patients were diagnosed with ASM, 2 with SSM and 17 with ISM. 17/17 ASM patients revealed a pathologic bone marrow pattern (sclerosis in 10/17, activated bone marrow in 7/17 patients) extending into the extremities in all patients. 2/2 patients in the SSM group and 4/17 patients in the ISM group revealed a pathologic bone marrow pattern which extended into the extremities in 1/2 (SSM) and 1/17 (ISM) group (p<0.001ln ASM pathologic lymph nodes, hepatosplenomegaly and ascites were present in 10/17, 16/17 and 15/17 patients while in the ISM group these findings were seen in 0/17, 2/13 and 1/17 patients (p < 0.001 for all three items). Exemplary images of a patient with ASM and characteristic findings are presented in Figure 1. In the small SSM group these findings were encountered in 0/2, 2/2 and 1/2 patients. Focal organ lesions were exclusively found in 4/17 ASM patients. Osteolyses were only seen in one patient.

Conclusion:

Wb-MRI seems to be a valuable clinical tool for reliable differentiation between ASM and ISM based on the specific pattern of bone marrow changes in combination with the presence of pathologic lymph nodes, focal organ lesions and ascites.

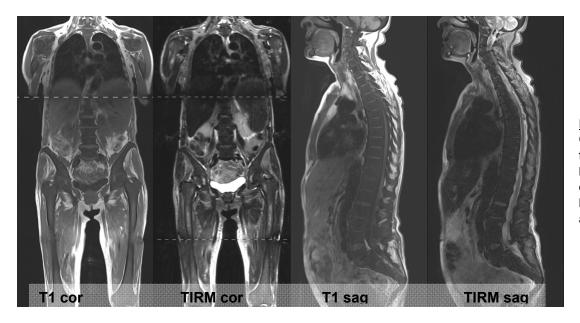


Figure 1:
Characteristic imaging findings of ASM: sclerotic bone marrow of spine and extremities, hepatosplenomegaly and

hepatosplenomegaly and ascites.

References

1. Horny HP et al. *Pathobiology* 2007