MAST CELL SARCOMA OF TIBIA

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Mastocytosis is a rare and mainly sporadic disease. Its main feature is an abnormal proliferation and tissue accumulation of mast cells. It can present with wide spectrum of local and systemic symptoms. Therefore, it is clinically easily misdiagnosed and pathohistological confirmation (morphological, immunohistochemical) is always necessary. WHO classification differentiates cutaneous and systemic disease. Mast cell sarcoma is a very rare disease, with only 3 well-documented cases described in literature. It is characterized by local proliferation of atypical mast cells, with destructive growth and very poor prognosis (1, 2).

A 4-year-old boy, presented with painful edema and deformation of his right lower leg. X-rays and computed tomography revealed destructive tumorous mass. Biopsy was performed. Histopathology showed tumor very rich with large, atypical cells, with hyperchromatic oval and polygonal nuclei. Cytoplasm around them was eosinophilic with many PAS-positive and Toluidine-Blue-positive granules (Fig. 1).

Figure 1. Toluidine-Blue-positive granules (left) and CD 117 positivity (right) in mast cells. (objective 40x)

Immunohistochemically mast cells were positive for CD68, vimentin, CD 117 (c-KIT) and CD 45 (common leukocyte antigen) (Fig. 1). These revealed these cells being malignant mast cells. Soon urticaria developed with elevated serum tryptase level. One month after biopsy, his primary disease progressed to mast cell leukemia, proved in bone-marrow analysis. Patient was subjected to chemo- and supportive therapy. Follow-up computed tomography showed oval masses in both lower limbs. Another targeted biopsy of the right tibia was performed, with same pathohistological findings as in the first one. Despite intensive therapy patient died 8 months after initial symptoms occurred.

Based on pathological findings, histochemistry and immunohistochemistry, we diagnosed for the first time a mast cell sarcoma of a bone, leading to leukemic transformation and death.
References:
