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Mast-cell sarcoma of the tibia

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The mast-cell sarcoma of a bone is described here for the first time. The tumour presented in a 4-year-old boy, with pain, oedema and deformation of his right lower leg. Radiological findings revealed a destructive tumourous mass. Histopathological examination showed the tumour to be composed of large, atypical cells, with hyperchromatic oval and polygonal nuclei. The cytoplasm around them was eosinophilic with many basophilic and toluidine-blue-positive granules. These atypical mast cells were positive for chloroacetate esterase, c-kit, tryptase and negative for myeloperoxidase. The primary disease quickly progressed to mast-cell leukaemia, and despite intensive chemotherapy the patient died 18 months after first symptoms.

Mast-cell sarcoma is a rare disease characterised by local proliferation of atypical mast cells, destructive growth and poor prognosis. Here we present a case of mast-cell sarcoma arising in bone.

A 4-year-old boy was admitted to the Clinical Hospital Osijek, Osijek, Croatia with 8-month history of painful oedema and deformation of right lower leg. *x* Rays and CT-scan revealed a tumourous mass on the right tibia (fig 1). There was no hepatosplenomegaly, and blood counts were normal. After biopsy and pathohistological diagnosis at the Institute of Pathology, Medical School, University of Zagreb, Zagreb, Croatia, the patient was moved to the Children's Hospital, Zagreb, Croatia. Soon after admission, a skin rash developed, with high serum tryptase level, but still blood counts were normal. Sternal fine-needle aspiration biopsy was performed and an aleukaemic variant of mast-cell leukaemia was diagnosed. The patient was subjected to acute myeloid leukaemia-Berlin–Frankfurt–Munster (AML-BFM) 2004 therapy protocol. A follow-up CT-scan showed oval masses in both tibias, without the involvement of soft tissue. Follow-up biopsy of the right tibia was performed. A few weeks before death the patient's blood count revealed low leucocytes. Liver and spleen were enlarged. Despite intensive therapy the patient died 18 months after initial symptoms occurred.

Histologically, bone and soft tissue samples were partially destroyed or permeated by sarcomatous tumour tissue. The tumour was almost exclusively composed of large, atypical cells, with hyperchromatic oval to polygonal and few larger bilobulated and multilobulated nuclei. Cytoplasm was eosinophilic, filled with basophilic granules on haematoxylin-eosin

(fig 2A,B), and periodic acid Schiff staining, displaying metachromatic-like granules on toluidine-blue (fig 2C) and Giemsa-stained slides. Among the tumour cells, many eosinophils and some histiocyte-like cells were present. Mitotic activity was very low, and blast-like cells with high nucleus-to-cytoplasm ratio were only occasionally observed. No spindle-shaped mast cells were present. Chloroacetate esterase reaction was positive, whereas myeloperoxidase was negative. Immunohistochemically atypical mast cells were positive for tryptase, CD68, vimentin, c-kit (fig 2D), CD45 and CD43. They were negative for CD3, CD15 and CD34 (all antibodies from DAKO, Glostrup, Denmark). Cytopathological analysis showed 90% of atypical cells being toluidine-blue, c-kit and tryptase positive. A follow-up smear revealed hypercellular bone marrow infiltrated with atypical mast cells positive for toluidine-blue, CD45, CD68, occasionally for CD2, and negative for myeloperoxidase, with suppressed erythropoietic, granulopoietic and thrombopoietic cells.

Mast-cell sarcoma has been to the best of our knowledge, described in only three cases, presenting as subglottic tumour, as a tumour of the ascending colon and as an intracranial tumour.^{1–3} Our patient did not meet the criteria for systemic mastocytosis.⁴ Based on our histological, histochemical and immunohistochemical data we were able to exclude myeloid

Figure 1 CT scan showing tumourous mass in the right tibia.

Figure 2 (A,B) H&E staining presenting atypical mast cells (objective $\times 25$ and $\times 40$). (C) Toluidine-blue-stained slide showing highly atypical neoplastic mast cells, displaying polygonal nuclei and cytoplasm filled with metachromatic granules (objective $\times 100$). (D) Membrane CD117 positivity of neoplastic cells (objective $\times 40$).

sarcoma and diagnose mast-cell sarcoma. As in our patient, symptoms connected to limb pathology occurred 10 months before the first cytological examination of the bone marrow. We think that a sequence of events going from tumour towards generalisation of disease seems logical. Hence, we conclude that our patient presented with mast-cell sarcoma of a bone, quickly leading to secondary generalisation and leukaemic transformation with rapid fatal outcome.

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Take-home messages

- Mast-cell sarcoma is a very rare neoplasm and the differential diagnosis between systemic mastocytosis, mast-cell leukaemia and mast-cell sarcoma is difficult
- For diagnosis of mast-cell sarcoma histochemical and immunohistochemical analysis correlated with clinical data is always necessary
- Mast-cell sarcoma can originate in bone with rapid secondary generalisation

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