Mast Cell Sarcoma: Clinical Management

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KEYWORDS

• Mast cell sarcoma • Mastocytosis • Clinical presentation of mast cell sarcoma
• Treatment of mast cell sarcoma • Prognosis of mast cell sarcoma

KEY POINTS

- Mast cell sarcoma is the rarest mast cell disorder.
- Prognosis of mast cell sarcoma is poor.
- Mast cell sarcoma affects both genders.
- Mast cell sarcoma is more common in canine, murine, and bovine species.

INTRODUCTION

Special stains for mast cells were described by Paul Ehrlich around the late 1800s and early 1900s. He studied dyes that stain tissues, cells, and infectious organisms. In 1908 he shared the highest scientific distinction, the Nobel Prize, with Metchnikoff, for this work. Mast cells have been described and named by him. Since then, they were identified in different tissues in many species.[1] The earliest report of mast cell sarcoma using different electronic computer searches dates back to 1948.[1] The species affected was dogs.

The disorders affecting mast cells have been described under the terms "cutaneous mastocytosis," "systemic mastocytosis," "mast cell leukemia," and "mast cell sarcoma" (**Table 1**).[2–5]

CLINICAL PRESENTATION AND PROGNOSIS

Human mast cell sarcoma is a very rare disorder of mast cells. There are 17 reported cases in the literature between 1997 and 2013.[6–18] In addition, included are 3 unpublished reports of patients diagnosed at the authors' institution (**Table 2**). The disorder

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Table 1
Classification of mast cell disorders

Groups of Mast Cell Disorders	Suggested Subgroups
Cutaneous mastocytosis	Urticaria pigmentosa Telangiectasia eruptiva perstans Bullous cutaneous mastocytosis Diffuse cutaneous mastocytosis
Systemic mastocytosis	Indolent Smoldering Aggressive Mastocytosis with an associated hematologic non-mast cell disorder
Mast cell leukemia	Aleukemic mast cell leukemia Leukemic mast cell leukemia
Mast cell sarcoma	

is difficult to diagnose because the mast cells in the tumor occasionally lose some of their surface diagnostic markers.

Organs Affected

Mast cell sarcoma can affect any part of the body. The reported affected areas are outlined in **Table 2**. Those include the following:

- Larynx
- Colon
- Small bowel
- Bones (tibia and temporal bone in 2 different patients)
- Buccal mucosa with invasion of the mandible and the external auditory canal

Occasionally mast cell sarcoma becomes metastatic, and in one report, the tumor was identified after it became metastatic. On other occasions mast cell sarcoma was associated with systemic mastocytosis and mast cell leukemia.

Age Groups Affected

The reports of mast cell sarcoma span from infants to elderly patients. The youngest patient was diagnosed at age 8 months and the oldest was diagnosed at 77 years. The median age of the patients was 39 years.

Survival

Patients who develop mast cell sarcoma have a short life expectancy (see **Table 2**), likely because of the lack of effective therapeutic measures, and the aggressiveness of the disorder. The survival ranged from 2 months to 8 years. Two patients had the longest survival because the tumor expressed c-kit without a mutation that is resistant to imatinib mesylate. Those 2 patients responded to imatinib mesylate therapy after surgical excision and radiation therapy. A third patient with uterine mast cell sarcoma was reported to achieve complete remission after surgery, radiation, and therapy with imatinib mesylate. This last patient's tumor did not express the Asp816Val c-kit mutation. The median survival of the patients for whom survival data was reported is 6 months.