

Mast Cell Sarcoma: Clinical Management

Catherine R. Weiler, MD, PhD, Joseph Butterfield, MD*

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 There are 17 reported cases in the literature between 1997 and 2013.^{6–18} In addition, included are 3 unpublished reports of patients diagnosed at the authors’ institution ([Table 2](#)).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

The authors have no disclosures and no conflicts of interest. Dr J. Butterfield and Dr C.R. Weiler are funded by Mayo Clinic Rochester.

Program of Excellence in Mast Cell and Eosinophil Disorders, Division of Allergic Disease, Department of Internal Medicine, W-15 Mayo Building, 200 First Street Southwest, Rochester, MN 55905, USA

* Corresponding author.

E-mail address: Butterfield.joseph@mayo.edu

Immunol Allergy Clin N Am 34 (2014) 423–432

<http://dx.doi.org/10.1016/j.iac.2014.01.004>

immunology.theclinics.com

0889-8561/14/\$ – see front matter © 2014 Elsevier Inc. All rights reserved.

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.

Table 2
Patients reported in the literature and identified in clinical electronic records

Number Assigned to Patients	Gender	Age at Onset	Age at Death	Site(s) of Mast Cell Sarcoma and Associated Disorders	References
1	F	71	76	Larynx, then metastatic	8
2	M	64	64	Metastatic	18
3	M	63	63	Metastatic with systemic mastocytosis	23
4	F	32	34	Colon, subsequent metastasis	21
5	F	8	8	Cerebral	7
6	M	44	Alive 2013	Left buttock, systemic mastocytosis with negative c-kit mutation and eosinophilia. Rx imatinib mesylate	Mayo Clinic Rochester records
7	M	4	4	Left tibia and aleukemic mast cell leukemia	9
8	M	71	72	Initial CMML, then developed small intestine mast cell sarcoma	Mayo Clinic Rochester records
9	M	61	Last contact 2012	Buccal mucosa extending into the mandible, surgery, chemotherapy, and radiation therapy	Mayo Clinic Rochester records
10	F	39	Complete remission	Uterus with ascites, negative c-kit, Rx imatinib mesylate	22
11	M	Elderly	After 4.5 mo of the diagnosis	Small intestine	10
12	F	63	N/A	Scalp with systemic mastocytosis Novel Kit mutation	23
13	F	15	N/A	Temporal bone	17
14	M	0	2	Right external auditory canal	16
15	M	39	41	Cutaneous mast cell sarcoma with a history of cutaneous mastocytosis and a mastocytoma	11
16	M	25	25	Thoracic mast cell sarcoma with mediastinal adenopathy and metastasis known familial indolent mastocytosis	13
17	M	42	42	Splenomegaly, mediastinal mast cell sarcoma with adenopathy	13
18	F	12	Alive in 2013	Left middle ear	18
19	M	19	Alive in 2013	Lip	18
20	F	77	77	Pelvis	18

Abbreviations: F, female; M, male; N/A, not available; Rx, therapy.

Clinical Presentation

The presentation of each of the patients reported has been unique. Mast cell sarcoma can affect any organ. It generally presents as a mass with relatively rapid growth. The symptoms depend on the site of the mass. All the patients' reports in the literature and those found in the electronic medical records are summarized in [Table 2](#).

The first patient,⁸ a woman aged 71, presented with a mass in the larynx that was first diagnosed as Wegener granulomatosis and treated with high-dose corticosteroids. Subsequent biopsies, about a year later, demonstrated the diagnosis of mast cell sarcoma, which became metastatic soon after the diagnosis, and she died at age 76.

The second patient,¹⁹ a man aged 62, presented with systemic disease and died within 3 weeks of diagnosis.

The third patient,²⁰ a man aged 63, was known to have a history of systemic mastocytosis. He presented with metastatic mast cell sarcoma at the age of 63 and died within 5 weeks of the diagnosis.

The fourth patient,²¹ a woman aged 32, presented with a colonic mast cell sarcoma involving retroperitoneal lymph nodes. Within 2 years, she developed metastatic disease involving the lymphatics and died at age 34.

The fifth patient,⁷ a girl aged 8, presented with cerebral mast cell sarcoma and died the same year of presentation.

The sixth patient (seen at Mayo Clinic Rochester) was a 44-year-old man. He presented with worsening pain and visible skin induration of the left buttock in 2005. He was found to have mast cell sarcoma of the buttock. A concomitant bone marrow biopsy test result showed the presence of systemic mastocytosis. C-kit analysis was negative for the Asp816Val mutation. He was last contacted, and was doing well, in 2013.

The seventh patient,⁹ a 4-year old boy, presented with a painful mass of his left tibia that was shown to be mast cell sarcoma. Because of subsequent elevation of serum tryptase and the development of a rash, he underwent a sternal bone marrow biopsy test. Study of the bone marrow and peripheral blood showed the presence of aleukemic mast cell leukemia. He died the year of diagnosis.

The eighth patient (seen at the Mayo Clinic Rochester) was a 71-year-old man with known chronic myelomonocytic leukemia (CMML). He developed gastroenterologic symptoms for which surgical intervention was required. The surgery showed an area of small intestinal thickening, which was resected. The pathology testing identified the presence of small intestine mast cell sarcoma from which he died the following year.

The ninth patient (seen at Mayo Clinic Rochester) was a 61-year-old man. He presented with a mass in the buccal mucosa in 2009. The mass subsequently enlarged and invaded the mandible. He underwent multiple therapeutic modalities. The last contact with him was in 2012 at which time he was asymptomatic. Imaging studies available included computed tomographic (CT) scan of the jaw, positron emission tomography scan of the head, and bone scan ([Fig. 1](#)).

The tenth patient,²² a 39-year-old woman, presented with a uterine mass associated with massive ascites. The biopsy specimen showed mast cell sarcoma that expressed c-kit. The Kit Asp816Val mutation was absent. After multiple therapeutic modalities, the authors considered her in complete remission.

The eleventh patient¹⁰ was referred to as an "elderly man." He was found to have small intestinal mast cell sarcoma from which he died within 4.5 months.

The twelfth patient,²³ a 63-year-old woman, presented with a mass in her scalp causing headaches. The biopsy specimen showed mast cell sarcoma and a bone

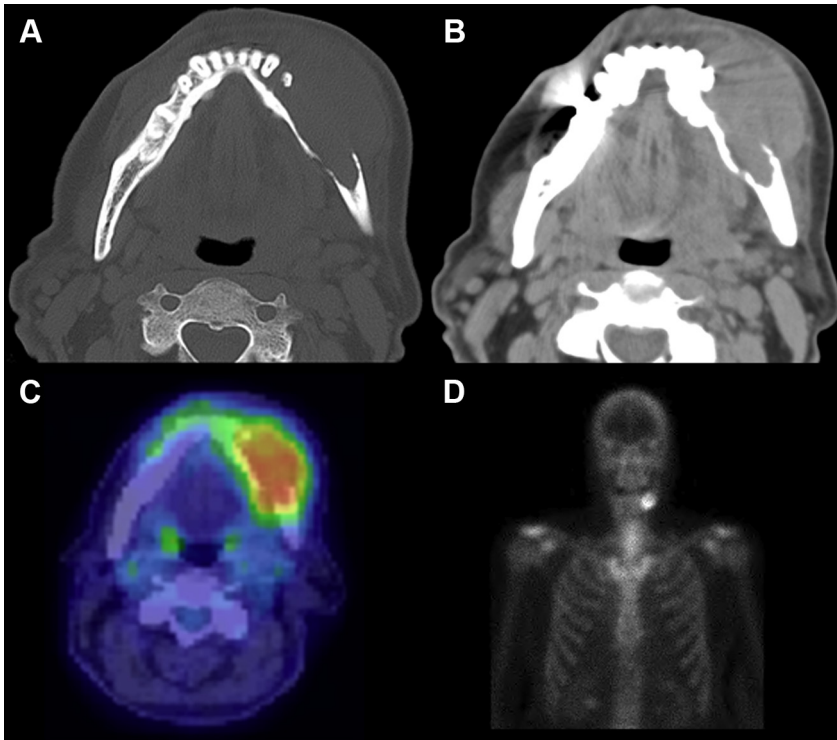


Fig. 1. (A) Patient #9-Axial noncontrast CT of the face, bone window, shows an expansile and lytic lesion of the left mandibular body, with associated cortical destruction laterally. (B) Axial contrast-enhanced CT, soft tissue window, shows an associated heterogeneously attenuating and enhancing soft tissue mass of the left lower face with the associated lytic involvement of the adjacent mandible. (C) Axial F-18 FDG positron emission tomography/CT image shows associated hypermetabolism of the soft tissue and lytic mass of the left mandible. (D) Coronal nuclear medicine Technetium-99m bone scan shows uptake in the left mandible corresponding to the mass noted on CT.

marrow biopsy test demonstrated existing systemic mastocytosis with c-kit D816V mutation. The authors did not mention the age at death of this patient.

The thirteenth patient¹⁷ was a 15-year-old girl who was found to have mast cell sarcoma of the temporal bone. The age of death of this patient was not reported in this article.

The fourteenth patient reported¹⁶ was a male infant who developed a mass in the right external auditory canal. Biopsies showed the mass to be mast cell sarcoma. Despite multiple therapeutic modalities, he died at age 2.

The fifteenth patient¹¹ was a 39-year-old man. He had a long-term history of cutaneous mastocytosis. He developed a mastocytoma that transformed into mast cell sarcoma. Despite therapeutic intervention, he died within 26 months.

The sixteenth patient¹³ was a 25-year-old man with familial indolent mastocytosis and urticaria pigmentosa. He presented with inguinal adenopathy and reduced lower extremity mobility. Further work-up revealed multiple lesions compressing the lower spinal cord and a large lesion compressing the thoracic spinal cord.

The seventeenth patient¹³ was a 42-year-old man. He presented with fever, sweats, rash, and cytopenias. He had a history of unexplained splenomegaly. At presentation he was found to have a mediastinal mass, generalized lymphadenopathy, multiple bone lytic lesions, and back muscle infiltration. He died within 1 to 2 months of presentation.

The eighteenth patient¹⁸ was a 12-year-old girl who presented with a large left middle ear mass, initially diagnosed as Langerhans cell histiocytosis, that proved resistant to multiple courses of chemotherapy as well as to local debulking surgery. The correct diagnosis was made only after 2 additional pathology reviews.

The nineteenth patient¹⁸ was a 19-year-old man who presented with a progressively enlarging submucosal nodule on the right lower lip. This patient also had a history of childhood-onset cutaneous mastocytosis that had resolved. An initial diagnosis was of ALK-negative anaplastic large cell lymphoma. Despite imatinib therapy and excision, the tumor recurred after 19 months.

The twentieth patient¹⁸ was a 77-year-old woman with right hip and groin pain who subsequently was found to have a pelvic mass. She declined bone marrow biopsy testing and died under hospice care after 4 months.

THERAPEUTIC MODALITIES

Multiple chemotherapeutic agents and tyrosine kinase antagonists have been tried and failed. The only 2 patients whose therapy seemed to be successful responded to a combination of multiple modalities including surgical debulking, radiation therapy, and combination chemotherapeutic agents including imatinib mesylate, a tyrosine kinase inhibitor.

Over the years during which the disease was reported the following agents were used.

In May 1970, the first patient, a 71-year-old woman, was misdiagnosed as having laryngeal Wegener granulomatosis and treated with surgical resection and high-dose systemic steroids. Steroids were continued until March 1972 at which point she was found to have a recurrence of the subglottic mass and what appeared to be metastatic disease with lymphadenopathy, which resulted in repeated pathologic evaluation revealing the diagnosis of mast cell sarcoma. She was treated with high-dose bleomycin over 6 weeks. The metastatic and primary lesion continued to grow. Bleomycin was replaced with high-dose radiation therapy, which resulted in transient remission. By December 1973, the metastatic disease recurred. Chemotherapy according to a modified "De Vita" regimen was successful transiently. She had recurrence of the metastatic disease and hepatosplenomegaly resulting in her demise in May 1974.⁸

The second patient¹⁹ report did not review the therapeutic intervention.

The third patient²⁰ report did not review the therapeutic intervention.

The fourth patient²¹ was a 32-year-old woman with mast cell sarcoma infiltrating the colon. She first underwent surgical resection of the lesion. After 2 years, she presented with a large abdominal mass and lymphadenopathy. Subsequent treatment included prednisone and radiation therapy. Despite the therapy, she died at age 34 from metastatic disease.

The fifth patient was an 8-year old girl who presented in 2001 with severe headaches and was found to have intracranial mast cell sarcoma. At first, she was treated with prednisolone and then surgical resection. This treatment was followed by radiation therapy, subcutaneous interferon α 2b, and cytarabine intrathecally. The tumor continued to proliferate instead of regress. She was treated with the regimen for

myelogenous leukemia/sarcoma in childhood. Interferon was discontinued and she received cytosine arabinoside, etoposide, idarubicin, and prednisolone. This regimen did not result in a remission and was therefore changed. Prednisolone was continued; thioguanine, vincristine, and Adriamycin were added. This regimen was followed by a combination of arabinoside and mitoxantrone and subsequently followed by arabinoside and etoposide. This regimen resulted in a partial remission. However, the tumor recurred 5 weeks after the discontinuation of the chemotherapy. The parents declined additional therapy and the child died within 6 weeks of the recurrence.⁷

The sixth patient (from the Mayo Clinic records) was a 44-year old man. He presented in 2005 with left buttock pain and infiltration of the skin with hardening. Imaging studies showed a mass in the left gluteal area. He was treated with aggressive surgical resection, which was followed by imatinib mesylate therapy. The patient was still alive when contacted in 2013.

The seventh patient, according to a report published 2007,⁹ was a 4-year old boy that was hospitalized for an 8-month history of pain, edema, and deformation of his right tibia, which was diagnosed as mast cell sarcoma with destructive bone invasion. A sternal bone marrow biopsy test revealed the presence of associated aleukemic mast cell leukemia. Despite aggressive surgical resection and chemotherapy (not specified in the case report), he died 10 months after hospitalization.

The eighth patient (seen at Mayo Clinic Rochester) was a 32-year old man. He was known to have CMML, which was under observation. After he developed abdominal pain and hemoptysis, thickening of the wall of the small bowel was identified. He underwent a diagnostic laparoscopy that revealed a mast cell sarcoma infiltrating the small bowel wall. He underwent surgical resection with end-to-end anastomosis. He initially did well but was subsequently found to have metastatic disease. He was treated with imatinib mesylate. Despite the therapy, he died within a year.

The ninth patient (seen at the Mayo Clinic Rochester) was a 61-year-old man. He was first seen in 2009 for a buccal lesion, which had been found to extend into and invade the mandible. He underwent aggressive surgical resection and radiation therapy, followed by imatinib mesylate therapy. He was last contacted in 2012 and was doing well on imatinib therapy.

The tenth patient²² was a 39-year-old woman who was found to have mast cell sarcoma of the uterus. She underwent total hysterectomy and bilateral oophorectomy. Treatment with imatinib mesylate was added. The authors report that she achieved total remission.

The eleventh patient was an elderly male patient with small intestinal mast cell sarcoma, reported in 2011.¹⁰ He underwent a diagnostic and therapeutic surgical resection with end-to-end anastomosis. Reverse transcription-polymerase chain reaction was used to study c-kit. He had an unusual mutation at c-kit Asn822Lys. He was initially treated with 40 mg prednisolone followed by 400 mg imatinib mesylate. He experienced what was a partial remission after 2 months of therapy. At the third month, however, the disease recurred. Therapy with interferon $\alpha 2b$ was instituted and resulted in an apparent resolution of the sarcoma after 6 months of interferon therapy. However, 2 months later, he was again started on therapy with imatinib mesylate. He died within 4 to 5 months of the diagnosis.

For the twelfth patient, therapy and prognosis were not discussed in the article.²³

Regarding the thirteenth patient, a report of a 15-year-old girl was published in March 2013.¹⁷ She presented with a headache and was found to have mast cell sarcoma of the temporal bone. It expressed c-kit, and mutation analysis revealed L799F mutation. The initial diagnosis and therapy followed a protocol for Langerhans cell histiocytosis 3. Subsequent evaluation showed tumor progression. The therapies tried

included vinblastine, dexamethasone, cladribine, clofarabine, multiple combination chemotherapeutic agents combined with tyrosine kinase antagonists, radiation therapy, and multiple surgical resections. Despite that, the tumor did not regress. No additional information about the patient was provided.

In May 2013, another report of an infant, the fourteenth patient, with mast cell sarcoma was published.¹⁶ The child presented with purulent otorrhea at 4 months of age. After treatment of the infection, a small mass was identified in the auditory canal that grew and invaded the bone by the age of 8 months. It was initially diagnosed as Langerhans cell histiocytosis. At the age of 11 months, the mass enlarged and serum tryptase was found to be elevated (34 ng/mL, with normal level <11.5 ng/mL). C-kit Asp816Val mutation was absent. The child was treated with surgical debulking and the experimental drug PKC412 for 1 year. This treatment reduced the tumor size and allowed for further debulking. At the time of the report, the child was 2 years of age.

Another report published in 2012¹¹ reviewed the clinical history of a 40-year-old man, the fifteenth patient, with known persistent cutaneous mastocytosis. He experienced a malignant transformation of a cutaneous mastocytoma into mast cell sarcoma. Surgical excision, radiation therapy, and imatinib therapy failed to induce a remission. The patient died within a few months.

A publication in February 2013¹³ reported 2 patients. The first (sixteenth) was of a 25-year-old man with a history of familial indolent systemic mastocytosis and urticaria pigmentosa. He was found to have a large thoracic mast cell sarcoma with metastasis to the lymph nodes and the bones. He failed therapy of 1 month with dasatinib and corticosteroids. After failed dasatinib therapy, cytosine arabinoside in combination with cyclophosphamide was tried but failed. When the tumor invaded the sphenoid sinus, therapy with cladribine was started but also failed and he died within a few days.

The second (seventeenth) patient was a 42-year-old man with a history of unexplained splenomegaly. He developed mast cell B symptoms, which include fever, night sweats, leukopenias, anemia, and hepatosplenomegaly. A CT scan of the chest and abdomen revealed the presence of mediastinal and retroperitoneal lymphadenopathy. It also showed a muscular dorsal mass with lytic bone lesions. After an erroneous initial diagnosis (anaplastic lymphoma), positive stains for c-kit, tryptase, and CD30 suggested the diagnosis of mast cell sarcoma. C-kit mutation analysis showed Val560Gly mutation. Therapy with corticosteroids and dasatinib was started but failed. The patient died within 10 days.

Patient 18, a 12-year-old girl, did not respond to therapy with 2CDA/Ara-C, ICE, clofarabine, ALCL 99, idarubicin/Velcade/Ara-C, and decitabine. Subsequently she was given radiation treatment and imatinib mesylate and scheduled for allogeneic stem cell transplantation. She is alive with persistent disease 45 months following initial presentation.

Patient 19, a 19-year-old man, experienced recurrent tumor 19 months after excision and treatment with imatinib 400 mg daily.

Patient 20 died 4 months following diagnosis after treatment with pamidronate and radiotherapy.

SUMMARY

Mast cell sarcoma is the rarest and most difficult of mast cell neoplasms to treat. Mast cell sarcoma might occur in any age group and could initially present in various organs. It tends to metastasize. An approach using combined surgical debulking, radiation therapy, and newer chemotherapeutic agents targeting mutated c-kit (tyrosine

kinase inhibitors) seems to have the best chance to induce remission. The authors' data revealed an almost 2:1 male-to-female ratio. Furthermore, 4 of the 20 patients with mast cell sarcoma had underlying systemic or cutaneous mastocytosis.

REFERENCES

1. Mulligan RM. Neoplastic diseases of dogs; mast cell sarcoma, lymphosarcoma, histiocytoma. *Arch Pathol (Chic)* 1948;46(5):477–92.
2. Sandes AF, Medeiros RS, Rizzatti EG. Diagnosis and treatment of mast cell disorders: practical recommendations. *Sao Paulo Med J* 2013;131(4):264–74.
3. Valent P, Arock M, Akin C, et al. The classification of systemic mastocytosis should include mast cell leukemia (MCL) and systemic mastocytosis with a clonal hematologic non-mast cell lineage disease (SM-AHNMD). *Blood* 2010;116(5):850–1.
4. Valent P, Akin C, Sperr WR, et al. Mast cell proliferative disorders: current view on variants recognized by the World Health Organization. *Hematol Oncol Clin North Am* 2003;17(5):1227–41.
5. Pardanani A. Systemic mastocytosis in adults: 2013 update on diagnosis, risk stratification, and management. *Am J Hematol* 2013;88(7):612–24.
6. Galanis E, Li CY, Philyik R. Mast cell disease mimicking granulocytic sarcoma. *Am J Hematol* 1997;56(3):194–5.
7. Guenther PP, Huebner A, Sobottka SB, et al. Temporary response of localized intracranial mast cell sarcoma to combination chemotherapy. *J Pediatr Hematol Oncol* 2001;23(2):134–8.
8. Horny HP, Parwaresch MR, Kaiserling E, et al. Mast cell sarcoma of the larynx. *J Clin Pathol* 1986;39(6):596–602.
9. Brcic L, Vuletic LB, Stepan J, et al. Mast-cell sarcoma of the tibia. *J Clin Pathol* 2007;60(4):424–5.
10. Bugalia A, Abraham A, Balasubramanian P, et al. Mast cell sarcoma of the small intestine: a case report. *J Clin Pathol* 2011;64(11):1035–7.
11. Auquit-Auckbur I, Lazar C, Deneuve S, et al. Malignant transformation of mastocytoma developed on skin mastocytosis into cutaneous mast cell sarcoma. *Am J Surg Pathol* 2012;36(5):779–82.
12. Georgin-Lavialle S, Aguilar C, Guieze R, et al. Mast cell sarcoma in an infant: a case report and review of the literature. *J Pediatr Hematol Oncol* 2013;35(4):315–20 doi:10.1097/MPH.1090b1013e318279e318392.
13. Georgin-Lavialle S, Aguilar C, Guieze R, et al. Mast cell sarcoma: a rare and aggressive entity—report of two cases and review of the literature. *J Clin Oncol* 2013;31(6):e90–7.
14. Chott A, Guenther P, Huebner A, et al. Morphologic and immunophenotypic properties of neoplastic cells in a case of mast cell sarcoma. *Am J Surg Pathol* 2003;27(7):1013–9.
15. Krauth MT, Fodinger M, Rebuzzi L, et al. Aggressive systemic mastocytosis with sarcoma-like growth in the skeleton, leukemic progression, and partial loss of mast cell differentiation antigens. *Haematologica* 2007;92(12):e126–9.
16. Bautista-Quach MA, Booth CL, Kheradpour A, et al. Mast cell sarcoma in an infant: a case report and review of the literature. *J Pediatr Hematol Oncol* 2013;35(4):315–20.
17. Kim YS, Wu H, Pawlowska AB, et al. Pediatric mast cell sarcoma of temporal bone with novel L799F (2395 C>T) KIT mutation, mimicking histiocytic neoplasm. *Am J Surg Pathol* 2013;37(3):453–8.

18. Ryan RJ, Akin C, Castells M, et al. Mast cell sarcoma: a rare and potentially under-recognized diagnostic entity with specific therapeutic implications. *Mod Pathol* 2013;26:533–43.
19. Horny HP, Kaiserling E, Sillaber C, et al. Bone marrow mastocytosis associated with an undifferentiated extramedullary tumor of hemopoietic origin. *Arch Pathol Lab Med* 1997;121(4):423–6.
20. Sotlar K, Horny HP, Lebherz J, et al. Assoziation einer Knochenmarksmastozytose mit extrem unreifem extramedullarem Mastzellsarkom. *Pathologe* 1997;18(3):252–6.
21. Kojima M, Nakamura S, Itoh H, et al. Mast cell sarcoma with tissue eosinophilia arising in the ascending colon. *Mod Pathol* 1999;12(7):739–43.
22. Ma HB, Xu X, Liu WP, et al. Successful treatment of mast cell sarcoma of the uterus with imatinib. *Int J Hematol* 2011;94(5):491–4.
23. Falletti J, Borgia L, Lalinga A, et al. Mast cell sarcoma of the scalp: the first sign of undisclosed systemic mastocytosis? *Pathol Res Pract* 2012;208(11):683–6. <http://dx.doi.org/10.1016/j.prp.2012.1006.1010>.