Histiocytic Sarcoma of Tibia: A Rare Case Report and Review of Literature

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Abstract

Histiocytic sarcoma is a rare disorder and there has been a lot of confusion and debate regarding its diagnosis and treatment. The World Health Organization (WHO) in 2008 aided in the standardization of diagnosis of histiocytic sarcoma; however, the treatment protocols are still not clear and the treatment is on the line of other hematological malignancies. This study intends to report a rare case of histiocytic sarcoma and the treatment protocol used and analysis of available literature. The usual sites of histiocytic sarcoma are the lymphoreticular system, skin, and gastrointestinal tract, but solitary bone involvement is rare. This disease being a localized one was treated locally with surgical curettage followed by radical radiation therapy. Systemic therapy was not offered to this patient and has been reserved in case a patient gets a systemic recurrence as done in most cases of B cell lymphoma. Based on follow-up until now, the patient is disease-free and doing well. Thus, this treatment protocol appears apt for this concerned patient; however, there is a need for a large-scale analysis of various reported cases to establish a standardized treatment protocol for this rare and aggressive disease.

Keywords
- Hematopoietic malignancy of tibia
- Histiocytic sarcoma
- Neoplasm of histiocytic origin
- Primary histiocytic sarcoma of bone
- Rare Neoplasm of bone
- Reticulo sarcoma

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Introduction

Histiocytic sarcoma is an extremely rare malignant neoplasm of the hematopoietic system with poor prognosis, literature quotes its incidence of less than 1% of all hematolymphoid neoplasms. It is characterized by morphological and immunophenotypic features similar to those of mature tissue histiocytes. There are expressions of histiocytic markers without dendritic cell markers. Neoplastic proliferation associated with acute monocytic leukemia should be excluded. Because a subset of cases occurs post mediastinal germ cell tumor, particularly malignant teratoma and teratocarcinoma are known to differentiate along hematopoietic lines. Thus, it is suggested that histiocytic sarcoma arises from pluripotent germ cells.

Clinically, it presents as a solitary mass that may be associated with fever and weight loss. Other less common systemic features are lytic bony lesions, hepatosplenomegaly, and pancytopenia. Three typical sites of involvement are lymph nodes, skin, and the gastrointestinal system. Immunohistochemically, these tumors are positive for one or more histiocytic markers such as the cluster of differentiation (CD) 68 (KP1, PGM1), CD163, and lysozyme, but negative for CD1a, CD21, CD35, CD30, and T cell, B cell, and myeloid lineage markers. S100 can be positive but is usually weak or focal. Ki67 is variable. With the development of immunohistochemical (IHC) techniques, most previous reported cases of HS are now generally recognized to be misdiagnosed examples of non-Hodgkin lymphomas, predominantly diffuse large B cell lymphoma or anaplastic cell lymphoma.

Thus, histiocytic sarcoma is a frequently misdiagnosed and underreported entity. This case report aims at reporting a rare incidence of histiocytic sarcoma at an atypical site, i.e, tibial condyles.

Case Report

A 70-year-old female patient presented with pain and swelling in the left knee joint since 2 months, with a tender hard swelling fixed to the bone. The X-ray of the left knee joint showed a lytic lesion in the medial condyle of the left tibia. MRI showed a well-defined, lytic, eccentric, expansile lesion in epimetaphysis of the proximal tibia (Fig 1a, b). Biopsy was performed and histological features were suggestive of high-grade sarcoma with possibilities of histiocytic sarcoma and epitheloid sarcoma. IHC was suggestive of EMA S100 SMA locally positive, strongly positive for vimentin, CD 68 diffusely positive, LCA, HLA DR lysozyme positive, suggestive of high-grade sarcoma consistent with histiocytic sarcoma. Blood investigations showed red blood cell (RBC) counts as 4.15 x 10^12/μL, white blood cell (WBC) count: 11,000/μL, and platelet count: 2.92 x 10^5/mm^3. Bone marrow biopsy did not reveal any malignant infiltration. PET CT showed localized increased metabolic uptake but no evidence of distant spread (Fig. 2a, b).

The case was discussed by the multidisciplinary tumor board. In view of the localized disease, a plan for surgical treatment was made. Because excision would have led to joint morbidity, curettage of bone marrow was done followed by local radiation therapy (45 Gy in 25 fractions, 5 days a week over 5 weeks) with the IMRT: Intensity Modulated Radiotherapy technique. Until the recent follow-up after 1.5 years of diagnosis, the patient is disease-free.

Discussion

The term histiocytic sarcoma was used by Mathe in 1970 for an entity earlier known by terms such as “reticulosarcoma” (Oberling), “recticulum cell lymphosarcoma” (Silhol), and “retothelsarcom” (Roulet). However, its description at that time was purely based on morphological similarity to macrophages. Since then, attempts to establish its identity in histiocytic cell lineages by immunohistochemical and cytochemical methods is going on. At present, several markers are being used to establish the diagnosis of histiocytic malignancies such as the cluster of differentiation (CD) 68 (KP1, PGM1), CD163, and lysozyme. CD1 63, a hemoglobin
scavenger receptor protein, is a novel marker used for identifying histiocytic cells with a greater degree of specificity and is a promising marker in the diagnosis of true histiocytic malignancies. The World Health Organization (WHO) defines histiocytic sarcoma as a malignancy with morphologic and immunophenotypic features that resemble those of mature tissue histiocytes.

The role of cytogenetics in diagnosis by studying TCR rearrangement or the absence of the Igh gene has been studied and verified. In 2001, the WHO stated that the absence of these genetic features is mandatory for the establishment of the diagnosis of histiocytic sarcoma.

This being a rare malignancy, a standard treatment regime is lacking. Treatment is mostly on the lines of large cell lymphoma. Studies have shown a relation of histiocytic sarcoma with B cell lymphoma lineage. Further studies have also shown two subtypes M1 and M2 macrophages, whose role in the diagnosis and treatment planning is yet to be established. Biological closeness of histiocytic sarcoma with lymphoma indicates similar prognostic values and treatment lines. For localized disease, surgical resection with or without radiotherapy is indicated, while the systemic disease is treated with chemotherapy regimens such as CHOP, CHOEP, and ICE. Evidence exists for other systemic treatment options with targeted therapies such as alemtuzumab in histiocytic sarcoma with lymphoma indicates similar prognostic values and treatment lines. For localized disease, surgical resection with or without radiotherapy is indicated, while the systemic disease is treated with chemotherapy regimens such as CHOP, CHOEP, and ICE. Evidence exists for other systemic treatment options with targeted therapies such as alemtuzumab in histiocytic sarcoma.

Histiocytic sarcoma primarily affecting bone is a rare entity. Although secondary bone involvement as a part of systemic disease is a well-known entity, the primary bone involvement by histiocytic sarcoma is sparsely reported in the literature. Lage et al. reported disseminated primary bone disease that was confused with multiple myeloma. They treated the patient with localized radiotherapy and systemic therapy. Bhalla et al. described a primary bone histiocytic sarcoma in the head and neck. The patient was being treated with systemic therapy. Our patient had a lesion in the tibial bone marrow. Because it was a completely localized disease and was completely excised, systemic therapy was not advocated.

Conflict of Interest
None declared.

References