

Case of the Week

Soft Tissue and Bone Pathology: Rosai Dorfman Disease

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July 24, 2019

History

A 65 year old African American female with a history of hypertension, hypothyroidism, osteoarthritis and presents with right tibial mass x 3 years. The patient reported right leg pain at rest and while walking. An x-ray revealed a 10 cm expansile lesion in the proximal right tibia. A PET/CT showed bilateral hypermetabolic intermedullary nodules in the left tibia and calcaneus along with hypermetabolic bilateral inguinal lymph nodes. The patient then underwent a bone biopsy. Sections of the biopsy are provided in figures 1-5.

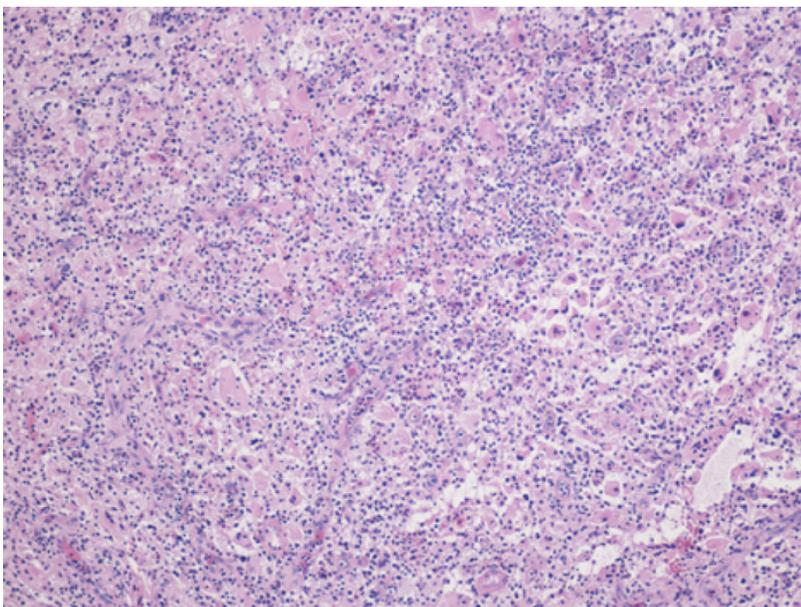


Figure 1: Xanthomatous histiocytic infiltrate with intermixed lymphocytes and plasma cells (H&E, 100x magnification)

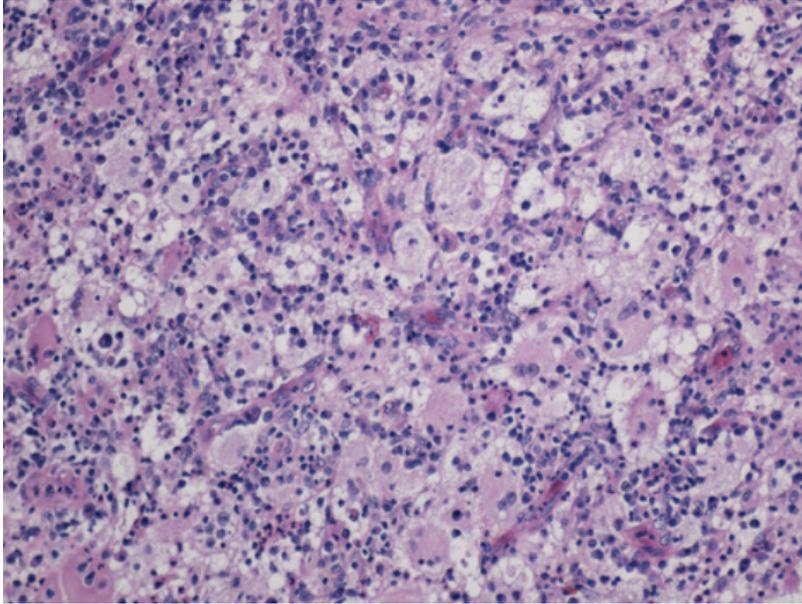


Figure 2: Xanthomatous histiocytic infiltrate with intermixed lymphocytes and plasma cells (H&E, 200x magnification)

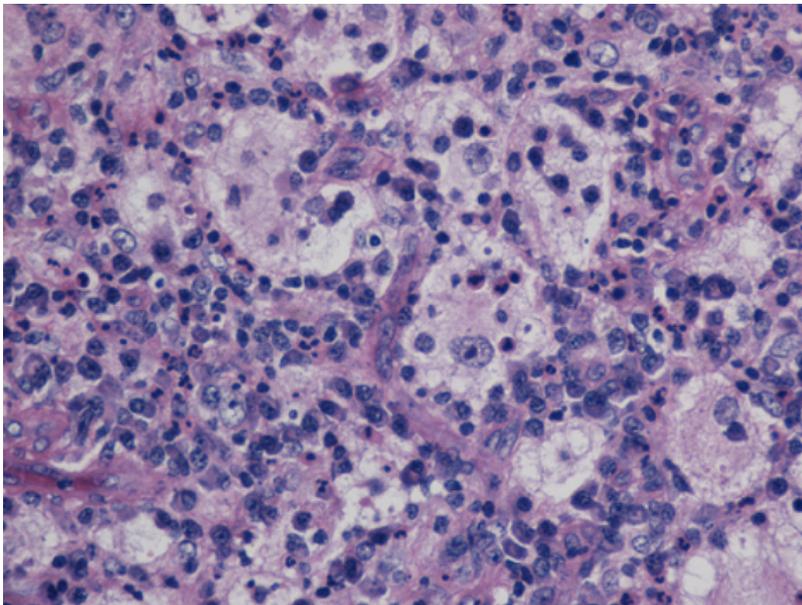


Figure 3: Neutrophils, lymphocytes and plasma cells within cytoplasm of histiocytes (engulfed in cytoplasm via a process called emperipolesis. The histiocytes contain large nuclei with prominent nucleolus (H&E, 400x magnification)

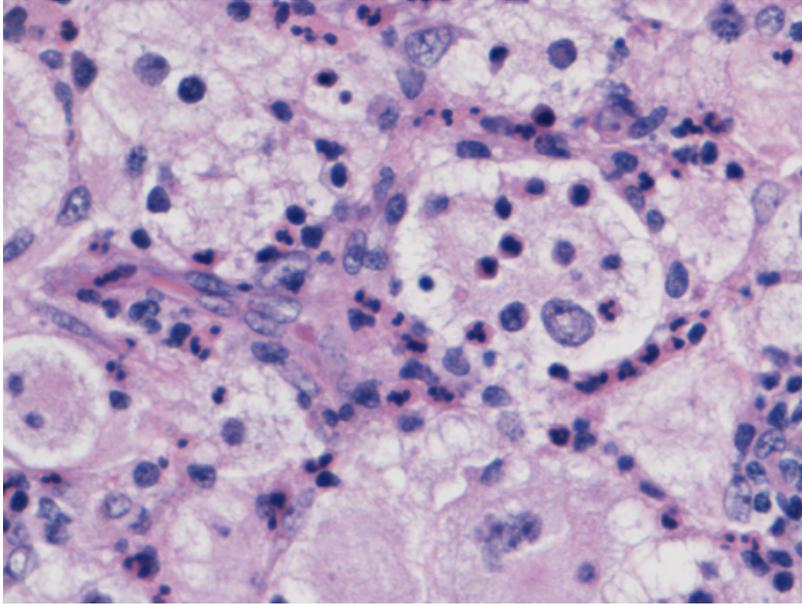


Figure 4: Neutrophils, lymphocytes and plasma cells being engulfed by histiocytes in a process called emperipolesis. The histiocytes contain large nuclei with conspicuous nucleoli and abundant clear xanthomatous cytoplasm (H&E, 600x magnification)

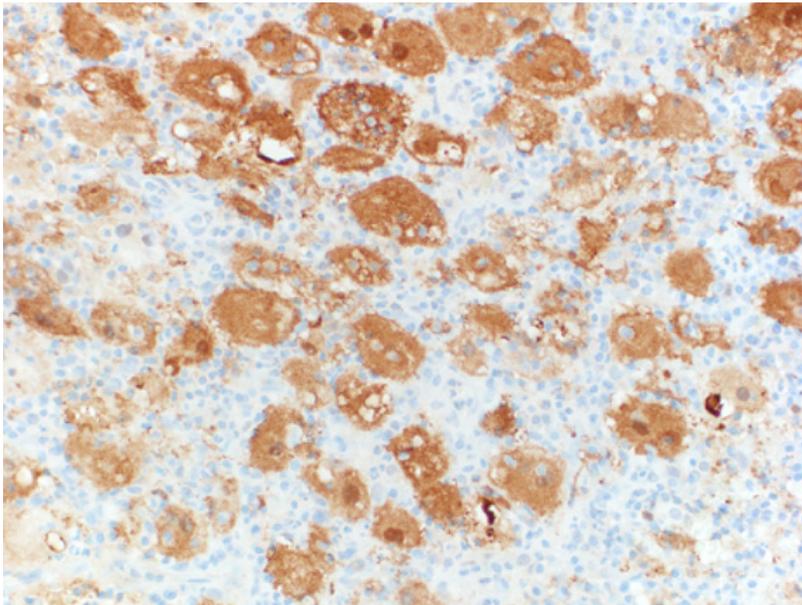


Figure 5: The histiocytes show immunoreactivity for S100 (IHC, 400x magnification)

Other immunohistochemical stains performed (not pictured): The histiocytes show reactivity for CD68 and CD163 and are non-reactive for CD1a and BRAF.

Diagnosis

Rosai-Dorfman Disease

Discussion

Rosai-Dorfman disease or sinus histiocytosis with massive lymphadenopathy is a non-neoplastic disease of unknown etiology characterized by proliferation of histiocytes. It usually presents in the lymph node (with the cervical lymph nodes being involved in up to 90% of cases) but 2-10% of patients present with bone involvement. Rosai-Dorfman disease is rare and fewer than 50 cases of primary Rosai-Dorfman of the bone have been reported in the literature. The mean age of presentation is 28 years with the age range being anywhere from 1.5 to 63 years. Patients typically present with localized pain and/or swelling or they may be asymptomatic. The most common areas affected are the metaphyseal region of long bones and the craniofacial bones. Most lesions are solitary but multiple bones may be affected.

Microscopically, the bone marrow spaces are replaced by numerous large histiocytes with abundant eosinophilic cytoplasm. The histiocytes contain vesicular nuclei and prominent nucleoli and demonstrate a process known as emperipolesis which is characterized by phagocytosis of leukocytes, plasma cells and/or neutrophils. The histiocytes show reactivity for S100, CD68 and CD163 and are negative for CD1a or BRAF. Currently, no genetic alterations have been detected in Rosai-Dorfman disease.

Treatment consists of either surgical curettage or surgical resection. Following local surgical treatment, 60% of patients with primary bone disease do not experience recurrence over a 12- to 18-month period, while 40% of patients develop extraosseous disease.

References

Deyrup AT, Siegal GP. Practical Orthopedic Pathology A Diagnostic Approach. Philadelphia, PA: Elsevier; 2016.

Mills SE, Greenson JK, Hornick JL, Longacre TA, Reuter VE. Sternberg's Diagnostic Surgical Pathology. 6th ed. LWW; 2015.