Rosai-Dorfman Disease (RDD) is a rare non–Langerhans cell histiocytosis characterized by accumulation of activated histiocytes within affected tissues. This is an idiopathic, non-neoplastic process that typically affects lymph nodes. Extranodal disease may occur in the skin, central nervous system and nasal cavity, however very rarely affects the breast. We present a case of Rosai-Dorfman Disease presenting as a right breast mass.

18 year old female with no significant medical history who initially presented to her primary care physician with a lump in her right breast and was referred to surgical oncology for biopsy. She had a complete ultrasound of the right breast which revealed a heterogeneous mixed echogenicity at 5 o clock 3.4 x 2.5 x 0.8cm. The differential diagnosis included phlegmonous inflammation, cyst, abscess and malignancy. She underwent an open core biopsy of right breast mass, and a complex indurated mass measuring approximately 3.5 x 3 x 3 cm in greatest dimension was noted. Pathology revealed Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy). This was sent to John's Hopkins pathology for second opinion who confirmed the diagnosis. At her post biopsy follow up visit, she was noted to have inflammatory inferior skin findings of right breast for which she was referred to hematology clinic for consideration of steroids. Baseline CT scans of the chest, abdomen and pelvis were obtained to assess for bulky lymphadenopathy and were negative. Upon subsequent follow up visit, the inflammation and skin changes around biopsy site had improved and no treatment was warranted. Decision was made to monitor with observation. At the 6 month follow up visit, breast lesion continues to regress and patient remains asymptomatic.

Rosai-Dorfman disease is a rare disorder characterized by proliferation and accumulation of histiocytes in the lymph nodes. Extranodal accumulation of histiocytes can also affect the skin, central nervous system and nasal cavity. In <1% of cases, can affect the breast and gastrointestinal tract. This is not characterized as a neoplastic process and the exact cause is unknown. Treatment is indicated only in symptomatic disease which may be from bulky adenopathy or related to extranodal site of disease. Most patients will often regress and spontaneously go into remission over time. If treatment is indicated, steroids are typically recommended as initial treatment. If unifocal disease, surgical resection may be considered. Disseminated and recurrent disease require more complex management including radiation therapy, immune modulating agents such as rituximab and lenolidamide and chemotherapy such as cladribine, vinka alkaloids and methotrexate.

REFERENCES


Radiopedia. Image contributed by G. Carbo, M.D. 6/2021