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507 - ROSAI-DORFMAN DISEASE OF THE BREAST: A CASE REPORT

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Rosai-Dorfman disease (RDD) is a rare histiocytic disorder that was first described by Destombes in 1965 and subsequently classified by Rosai and Dorfman in 1969 as a distinct clinical and pathological condition. It is a self-limiting disease process characterized by a non-Langerhans cell benign proliferation that usually involves the lymph nodes, but can affect extranodal sites. Classic RDD most commonly presents as massive bilateral painless cervical lymphadenopathy, with associated fever and loss of weight in children and young adults. Despite the predilection for the head and neck lymph nodes, RDD may present extranodal case in 43% of cases and in rare instances (around 23% of extranodal cases) only extranodal involvement can be described. The more common extranodal sites include the skin, nasal cavity, bone, orbital tissue, and the central nervous system. RDD involving the breast tissue is extremely rare, with fewer than 50 reported cases and it is important to recognize because it can mimic malignancy. Patients with involvement of the breast can present with painless palpable mass and ill-defined sensation or abnormal mammogram. The radiologic presentation of RDD lesions can have a notable diversity, but commonly have an appearance that is indistinguishable from breast carcinoma on mammogram and US. Therefore, the main diagnostic modality is histopathologic evaluation, with the features of marked dilatation of sinuses due to accumulation of activated histiocytes that demonstrate variable degrees of emperipolesis, along with the immunohistochemical characteristics like S100+, CD68+, and CD1a–. Management of RDD depends on the symptoms and it can be conservative with observation, as long as some cases have spontaneous regression, especially in the classic nodal disease. Surgical excision may be indicated in unifocal extranodal disease, especially for symptomatic cases. A 14-year-old female adolescent visited a breast surgeon with a self-detected painless palpable mass in the left breast for 3 months, without systemic symptoms or significant family history. On examination, there was a firm 3.5-cm mass in the upper inner quadrant of the left breast. The mammogram revealed a focal distortion on axillary tail in the left breast, designated as BIRADS 4. US demonstrated two masses in the left breast, including a 3.4×2.6×1.5 cm hypoechoic, irregular mass at 11:00 and a 1×0.9×0.7 cm hypoechoic, irregular mass at 11:00, both near the infraclavicular region, accompanied by a 2.8×1 cm atypical lymph node in the left axillary region, designated BIRADS 4C. US-guided core biopsies of both masses were obtained, which showed a nonspecific chronic inflammatory process. A new core biopsy was performed but once more with nonspecific histopathological findings, as well as negative results from culture examinations. Following these indeterminate findings, the patient underwent excisional biopsy, which histopathologic conclusion finally elucidated the extranodal RDD diagnosis.