CASE REPORT

27. A Case of Rosai-Dorfman Disease

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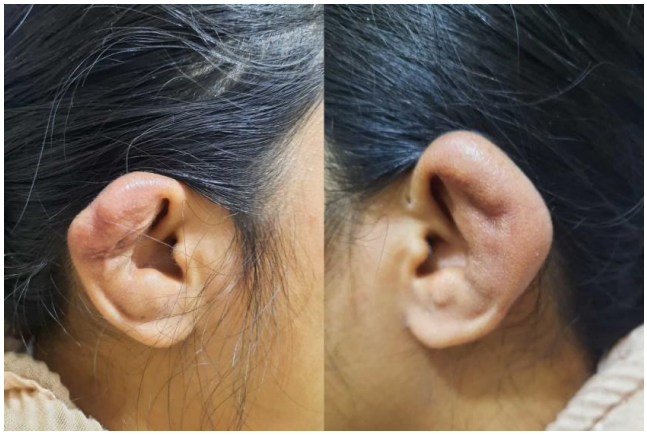
https://www.youtube.com/watch?v=hJIcIJ1w8oM&list=PLhqNq3xJClbafO0Y5bvBcgMmXpgzJxd44&index=5&t=15180s

Background: Rosai-Dorfman Disease (RDD) is a rare form of histiocytosis. We have observed extralymphatic involvement in about 43% of RDD cases in the literature, while dermatological symptoms manifest in only about 10% of cases. We present a case of RDD characterized by both nodal and extranodal involvement, initially presenting with bilateral ear nodules and dyspnea that closely resembled the clinical features of relapsing polychondritis; unfortunately, the diagnosis was missed at first. This case highlights the necessity for greater awareness and education among healthcare professionals regarding RDD to do diagnosis and treatment.

Case Report: A 25-year-old lady had a one-year history of discomfort and swelling in the cartilaginous part of the ears, accompanied by dyspnea (MRC dyspnea scale 2). A clinical diagnosis of relapsing polychondritis was made by an ENT surgeon, and oral steroids and NSAIDs were given. An inhaler containing salbutamol was also prescribed. The symptoms progressively worsened, her left ear became more swollen, dyspnea worsened (MRC dyspnea scale 3), she also developed bilateral cervical lymphadenopathy, and she presented to the medicine OPD. The examination revealed bilateral enlargement of the ear helices, resembling a cauliflower-like appearance (Figure 1). This time, FNAC from the left ear and a biopsy of the cervical lymph node were done, and they showed proliferation of histiocytes. Most of the histiocytes have abundant foamy cytoplasm and prominent nucleoli. These cells were found admixed with plasma cells with Russell bodies, lymphocytes, and neutrophils. Histiocytes showed features of emperipolesis. Fibrosis with a vague storiform pattern is present. These morphologic features are compatible with RDD. Routine laboratory investigations were normal. Serologies for antinuclear antibody (ANA) were positive (16.5 U/mL), and anti-dsDNA was negative. Spirometry revealed an obstructive pattern with a forced expiratory volume in one second (FEV1) of 3.2 L (50% of the predicted value). Computed tomography of the chest revealed bilateral hilar lymphadenopathy. Investigations concerning sarcoidosis and tuberculosis were negative. A watchful waiting strategy was taken as a management plan with medications of oral methotrexate and a salbutamol inhaler.

Conclusion: RDD is a rare non-Langerhans cell histiocytosis that commonly presents with significant lymphadenopathy and may or may not have systemic symptoms. The disease may affect almost every organ system, mimicking different clinical conditions. Frequently afflicted areas include the nasal cavity and sinuses, gastrointestinal, pulmonary, renal, musculoskeletal, and genitourinary systems, and, less frequently, the skin. FNAC or tissue biopsies are needed to diagnose RDD, and they commonly show extensive, pale histiocyte infiltrates with intact intracytoplasmic lymphocytes, which is known as emperipolesis. Histological features help diagnose this condition and separate it from other histiocytic illnesses. Medical literature inadequately describes the prognosis of RDD. In certain cases, a lengthy clinical course might lead to spontaneous remission. Multifocal illness, vital organ involvement, and a big mass worsen prognoses. No guideline on RDD management exists due to its rarity, making standardized therapy difficult. Knowing the clinical and histological presentations of RDD is crucial for accurate diagnosis, as it often mimics other disease conditions. Research is needed to determine the potential treatments.

Figure 1: Bilateral Cauliflower-Like Appearance of the Ears.



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