Case Report

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Surgery Section

Rosai-Dorfman Disease- A Surprise diagnosis in Follow up of SCC Pyriform Sinus -Post Surgery and CTRT

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ABSTRACT

Rosai-Dorfman disease also known as Sinus Histiocytosis with massive Lymphadenopathy (SHML). SHML was first described by Rosai and Dorfman in 1969 [1] and is now considered a non malignant inflammatory disorder in which the precise origin of pathologic cells is still controversial [2]. SHML is a rare self-limited pseudolymphomatous disorder of unknown etiology usually present with cervical lymphadenopathy, fever, elevated ESR and hematological abnormalities [3].There is a higher prevalence in males, and among Afro-Caribbeans as compared to Caucasians and Asians. Also, a familial association has been observed in some cases [4]. We present a case of 60-year-old male patient with a case of Rosai-Dorfman Disease in Follow up of SCC Pyriform Sinus-Post Surgery and CTRT

Key Words: Lymphadenopathy, Emperipolesis, Rosai-Dorfman disease

CASE REPORT

Sixty years old male patient, with a case of Squamous cell carcinoma of Right pyriform Sinus with known history of Diabetes mellitus, Hypertension, Chronic renal dysfunction & Hypothyroidism. Status post Total laryngectomy and Bilateral Neck Dissection, concurrent Chemoradiation completed. The patient came with complaints of cervical and inguinal lymphadenopathy after 5 years of treatment. No other complaints. On examination, he was moderately built and nourished. The cervical and inguinal lymph nodes were enlarged, 2-3cm in size, firm, non-tender and not-matted.

We evaluated the patient and went ahead with lymph node biopsy for the tissue dignosis. Histopathology of Lymph Node specimen showed, cut surface pale brown. Microscopically there was lymph node with partially effaced architecture showing lymphoid follicles and expanded interfollicular zone. The interfollicular area showed many plasma cells and macrophages. Many macrophages show emperipolesis suggestive of Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease).

DISCUSSION

SHML (also known as Rosai-Dorfman syndrome) is a rare, benign and self-limited disorder (433 cases were described in 1990 by Foucar et al.) that occurs most frequently in the first two decades of life. Its etiology is largely unknown. Sporadic cases have been associated with EBV or herpes virus infections and with malignant lymphoma [2]. More than 90% of the patients with SHML present with massive bilateral, mobile, and non tender cervical lymphadenopathy [5]. These nodes may at times be matted and prominent by periportal fibrosis. Fever, elevated ESR, neutrophilia, polyclonal gammopathy is other common associations [6]. Forty percent of the cases may show extra nodal involvement. Soft tissues, generally eyelids, orbits and ocular adnexa, skin and subcutaneous tissue, gastro-intestinal tract, upper respiratory tract and central nervous system can also be involved [7]. The cutaneous manifestations, which develop in about 10% of patients, are asymptomatic xanthoma-like, yellowish or reddish brown papules, nodules and plaques which may ulcerate [6].

On histological examination, there is a progressive filling up of the Lymph Node sinuses with normal histiocytes and lymphocytes. The histiocytes in SHML are S100 positive and CD1 negative [5]. The constant feature on histology, ‘emperipolesis’ or lymphophagocytosis’, is the presence of intact lymphocytes within the cytoplasm of histiocytes. This is of great diagnostic significance. Pericapsular fibrosis and inflammation are also seen. Ultrastructurally, histiocytes in SHML lack Birbeck granules and viral particles [4].

The prognosis is excellent in most cases. Complete spontaneous regression is known to occur. Only two cases of progression, one to malignant lymphoma and another to amyloidosis have been documented. Complications are mostly due to the pressure effects exerted by the enlarged

cervical Lymph Node. Extensive disease may lead to complications due to immunological abnormality that may be present [7]. The spread of the disease to vital organs seems to be the direct cause of death in very rare instances, but death is more frequently due to a significant defect in immune function, particularly at the level of the T-cell network [2]. There is no specific treatment for the Rosai-Dorfman syndrome. Treatment is required when the condition is organ threatening or life threatening. The treatment modes include corticosteroids, chemotherapy with a combination of vinca alkaloids and alkylating agents, low dose interferon, radiation therapy and surgery [8]. The etiology of Rosai-Dorfman Diseaese is unknown though it has been speculated that an occult chronic infection or an aberrant exaggerated immune response to an infectious agent or an antigen causes the initial histiocytic proliferation [9].

A Surgeon comes across a case of lymphadenopathy every now and then with a clinical picture hard to tell from malignant lymphomas or benign proliferative pathology. Histopathology of such conditions may rarely present as a surprise diagnosis of Rosai-Dorfman Disease. At the same time it is a relief to the physician and the patient, as this disease essentially has a benign course.

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