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CASE REPORT **OPEN ACCESS**

An Extremely Rare and Interesting Case of Rosai-Dorfman Syndrome Coexisting with Malignancy: A Case Report

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ABSTRACT
Rosai-Dorfman Syndrome (RDS) is a rare histiocytic disorder known for its distinctive clinicopathological features. Coexistence with malignancy is exceptionally rare and introduces complexities to its clinical presentation and management. This is about a 55-year-old female presented with calf and gluteal swellings, initially suggestive of peripheral nerve sheath tumor. Further investigations revealed normocytic normochromic RBCs with anisopoikilocytosis and neutrophilic leucocytosis. Imaging studies indicated subcentimetric lymph nodes. FNAC results indicated spindle cell sarcoma and metastatic deposits, leading to the diagnosis of Rosai-Dorfman Disease (RDD). However, subsequent lymph node biopsy confirmed peripheral T-cell lymphoma. Treatment involved oral steroids, methotrexate, and folvite for RDD, resulting in symptomatic improvement. Subsequent lymph node biopsy revealed peripheral T-cell lymphoma, prompting chemotherapy with cladribine. Despite a positive response, cessation of steroids led to disease relapse with severe manifestations. High-dose steroids, chemotherapy, and oral chemotherapy with endoxan were initiated, with a gradual improvement in symptoms. The patient responded well to the combined treatment, emphasizing the importance of continuous follow-up. This case underscores the intricate interplay between Rosai-Dorfman Syndrome and malignancy, emphasizing the need for a nuanced diagnostic approach and comprehensive management strategies. Further research is warranted to elucidate the pathogenesis, clinical spectrum, and treatment outcomes of this rare coexistence.
KEYWORDS: Rosai-Dorfman syndrome, Malignancy, Histiocytic disorder, Steroid therapy

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INTRODUCTION

The Rosai-Dorfman Syndrome (RDS), a rare and distinctive histiocytic disorder, seldom fails to captivate the interest of clinicians and pathologists due to its intriguing clinicopathological features. First described by Juan Rosai and Ronald Dorfman in 1969, RDS typically manifests as painless, massive lymphadenopathy and is renowned for its self-limiting course [1-3].

However, the narrative takes an unprecedented turn when RDS coexists with malignancy, an exceedingly rare phenomenon that adds layers of complexity to its clinical presentation and management [4-6].

This case report aims to shed light on an extremely rare and captivating instance – the coexistence of Rosai-Dorfman Syndrome with malignancy. As we delve into the details of this exceptional case, our objective is to provide insights that extend beyond the conventional understanding of RDS and to foster a deeper appreciation for the intricacies of these rare clinical entities.

CASE REPORT:

A 55/F, with no comorbidities, came to OPD in Jan 2020, with complaints of Swellings in Right calf since2months and right gluteal swelling (1.5x2cm). On examination, they are firm nodules with no warmth/tenderness. All routine investigations were normal, except Peripheral smear showed Normocytic normochromic RBCs with anisopoikilocytosis, Neutrophilic leucocytosis with MILD SHIFT TO LEFT and FEW REACTIVE LYMPHOCYTES.

USG of nodes was done which showed features suggestive of Peripheral Nerve Sheath Tumour. CT Chest showed Sub centimetric lymph nodes in Right Paratracheal and Sub cranial region. CT Abdomen showed no nodes. USG Neck also showed Bilateral Sub centimetric Lymph nodes, largest being 13x6mm.

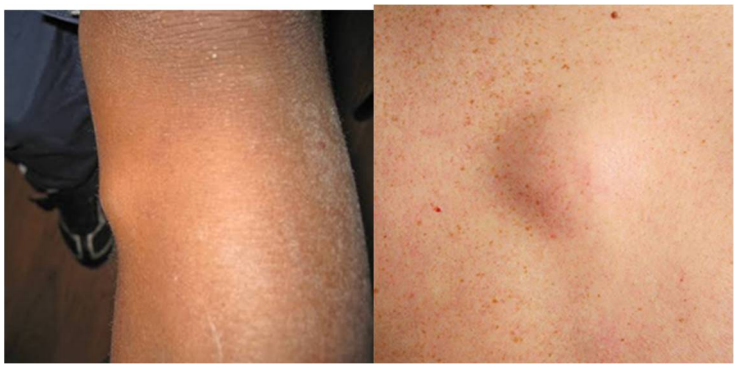

Fig 1: SUBCUTANEOUS SWELLINGS ON RIGHT CALF AND GLUTEUS

Platelets were normal, Serum calcium and Vit D were normal, EBV IgM and Mantoux were negative, ruling out other causes like TB, SARCOIDOSIS, EBV INFECTION.

On next visit, FNAC of swellings is done in which inferior swelling showed features of Spindle Cell Sarcoma and superior swelling showed features suggestive of METASTATIC DEPOSITS IN LYMPH NODE. Hence, HPE was done to confirm the diagnosis which showed MASSIVE PROLIFERATIVE ENLARGED LYMPHOCYTES(HISTIOCYTES) DEMONSTRATING EMPERIPOLESIS. Immunology done for lymphocytes which WERE EXPRESSIVE FOR CD3, CD20, CD68, LCA, S100 BUT NOT FOR CD1a, finally confirming the diagnosis of ROSAI-DORFMAN DISEASE.

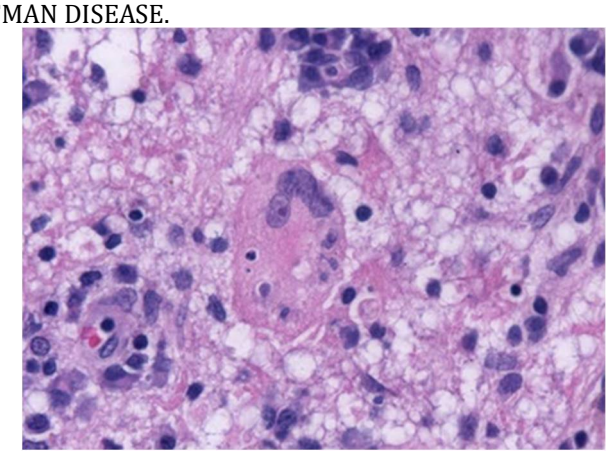

Fig 2: HPE SHOWING EMPERIPOLESIS

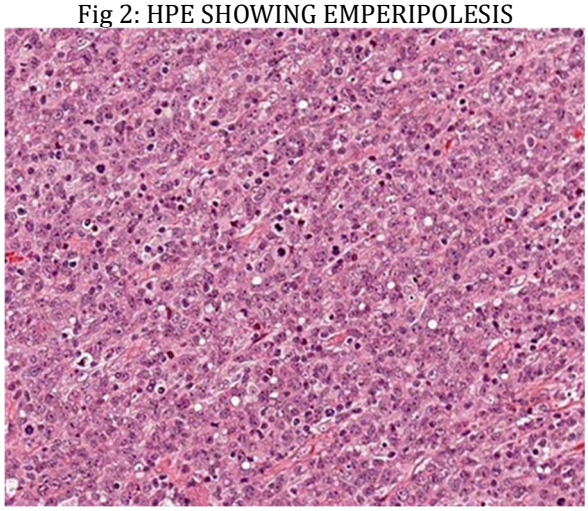

Fig 3: LYMPH NODE BIOPSY SHOWING PERIPHERAL T-CELL LYMPHOMA

FINDINGS AND TREATMENT

The patient was immediately started on Oral Steroids (WYSOLONE), Methotrexate and Folvite, as advised by RHEUMATOLOGY. Swellings gradually showed decrease in size with course of time and doses were tapered accordingly. In view of suspicion of malignant metastasis, Lymph Node Biopsy was done which

showed features diagnostic of PERIPHERAL T-CELL LYMPHOMA, confirming the malignancy. The patient was referred to MEDICAL ONCOLOGY and was started on Chemotherapy with CLADRIBINE (5mg/m/day x 5days every 28days for 3-4cycles). Patient became symptomatically better.

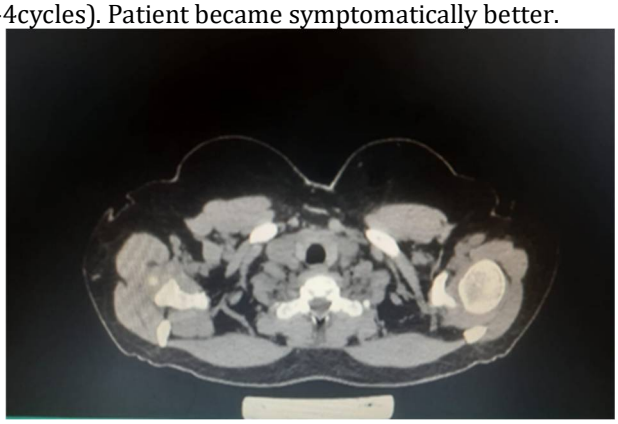

Fig 4: CT CHEST SHOWING MULTIPLE SUBCENTIMETRIC LYMPH NODES

OUTCOME

During this course, patient stopped taking Oral Steroids for 3months on her own decision and New lesions started appearing again all over the body. CT Chest and CT Abdomen both showed MULTIPLE SOFT TISSUE DENSE LESIONS AND LYMPH NODES, this time more severe than the initial stages. Steroids were restarted at HIGHER DOSE and chemotherapy was continued. After the COMPLETED cycles, it was changed to ORAL CHEMOTHERAPY WITH TAB.ENDOXAN 50MG OD for a period of 2weeks.

Then, Steroids were continued further PARALLELLY. Corticosteroids were well tolerated overall, and no major dose limiting toxicities were reported. Patient is continued on oral steroids. Patient improved gradually with decrease in swellings and is on REGULAR FOLLOWUP TILL TO DATE.

CONCLUSION

RDD is a relapsing and remitting disease with 50% spontaneous remissions. Pathogenesis is not well understood, and it is unclear whether to be classified as Neoplastic/Benign. Histopathological diagnosis of RDD can be CHALLENGING due to its rarity and specific histologic findings, especially in extra nodal forms. RDD is syndromic in nature with wide spectrum of manifestations. The cells can be POLYCLONAL in nature. Clinical spectrum and treatment outcomes are also not well defined. Hence, it is important to DIAGNOSE THE DISEASE AT THE EARLIEST, and rule out any neoplastic etiology involved, so that early intervention can be done accordingly. STRICT FOLLOWUP should be advised to the patient to COUNTERACT THE HIGHER REMISSION RATE.

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