

Case Report

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Management of a patient with Rosai–Dorfman syndrome and large adrenal pheochromocytoma

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Abstract:

Sinus histiocytosis with massive lymphadenopathy that known as Rosai Dorfman syndrome (RDS) is a rare histiocytic disease which includes benign lymph nodes (LN) enlargement with extra-lymphatic soft tissue involvement. The concomitant presentation of RDS with an adrenal tumor is rarely encountered among our patients. Here in we describe a middle-aged man, presented with a chronic history of repeated LN enlargement. The nodes had mild local discomfort and tenderness with associated fever. LN biopsy showed sinus histiocytosis with lymphophagocytosis characterized as emperipolesis and diagnosis of RDS was decided. After one year, the patient presented with high blood pressure, raised blood sugar, obesity, and abdominal pain. Computerized tomography showed a large adrenal mass, and the patient was treated urgently for his malignant blood pressure followed by surgical excision of the left adrenal mass. The biopsy subsequently revealed non-malignant adrenal tumor (pheochromocytoma). We are reporting successful treatment of rare case with RDS concomitantly presented with large adrenal pheochromocytoma. He was treated medically and surgically, the patient got recovery. RDS is a rare disease but we should consider it in patients with repeated benign lymphadenopathy. we are emphasizing that knowledge about RDS and its disease associations may be informative to the general medical community.

Keywords:

Adrenal pheochromocytoma, Rosai–Dorfman syndrome, sinus histiocytosis

Introduction

Sinus histiocytosis with massive lymph node (LN) enlargement was first described in 1969 by Rosai and Dorfman as a distinct clinicopathological entity.^[1] It is characterized by nonmalignant proliferation of histiocytes (non-Langerhans histiocytes) and phagocytes in the LNs and extralymphatic sites.^[2] Rosai–Dorfman syndrome (RDS) patients are usually asymptomatic or presented with mild symptoms, the median age of 39 years at presentation and more frequently found in male gender and African American than Asian backgrounds.^[1–3] Clinical features include fever, painless LN enlargement, and increased leukocytes. The LN enlargement

is usually found in the head, neck, cervical, axillary, and inguinal regions. Extralymphatic involvements are reported also, and the most common sites are a central nervous system, orbit, eyelids, skin (mainly in females), and gastrointestinal system and upper respiratory tract.^[4,5] Very rarely, some patients present with a painless maculopapular rash and bone infiltration with hepatosplenomegaly.^[3,6] The etiology of RDS is unknown, but many factors are described as immune dysfunction and viral infections such as parvovirus B19, EBV, and HHV-6 or may be associated with IgG4-related disorders and T-regulatory cell dysfunction. Genetic abnormality (SLC29A3) had been reported in a family with RDS. Patients with RDS are usually better to be sent to imaging a brain MRI to exclude CNS involvement.

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LN biopsy reveals matted enlarged LNs with thick capsules, and in RDS, you may see LNs contain histiocytes that have an abundant cytoplasm which contains lymphocytes and plasma cells and this process called emperipolesis.^[1,2] Immunohistochemistry shows positive for CD68, CD163, and S100 and negative for CD1a and BRAF-V60E.^[7,8] The common differential diagnoses include tuberculosis, sarcoidosis, IgG4-related diseases,^[9-14] juvenile xanthogranuloma, Gaucher disease, histiocytic disorders, Hodgkin's/non-Hodgkin's lymphoma, and melanoma.

The treatments of RDS are usually variable which include surgical excision, or chemotherapy like alkylating agents, anthracyclines, Methotrexate, cladribine, and lenalidomide, or biological agents such as rituximab, retinoids, or steroid, and interferon, lastly topical cryotherapy, and radiotherapy also have been mentioned.^[15-19]

The prognosis is like that one-fifth of patients got spontaneous regression, while two-thirds of them may have frequent relapses. Here in we describe successful treatment of rare case with RDS who presented with the large adrenal tumor. we are emphasizing that knowledge about RDS and its disease associations may be informative to the general medical community.

Case Report

A 43-year-old male presented with bilateral inguinal swelling for a 3-year duration. The condition associated with intermittent mild fever, mild inguinal pain, and tenderness. Frequent fine-needle aspiration has been done, and the excision biopsy of the LNs revealed reactive hyperplasia; finally, one of the biopsies showed marked sinus histiocytosis with lymphophagocytosis (emperipolesis), with lymphoplasmacytic infiltration

with S100 positivity [Figure 1a-c]. One year later, he developed headache, sweating, and attacks of raised blood pressure reaching >200/100 mmHg, hyperglycemia, and irritability, with weight gain and obesity. The patient investigated by ultrasonography and computerized tomography, revealing a well-defined mass between upper pole of the left kidney and spleen (79 mm × 83 mm) [Figure 2a] and right side inguinal LN enlargement of about 42 mm in diameter; the sonography of the neck showed enlarged LNs with reactive changes.

On routine blood tests, there was high blood sugar and high alanine aminotransferase, while other blood tests were normal. The patient had been admitted to the medical unit for the control of his blood pressure and blood sugar for few days before preparation for surgical excision of the left adrenal mass on September 2015. The excised mass grossly looked like an encapsulated oval mass of 8.5 cm × 7 cm × 6 cm, weighed 200 g, adherent to the normal-looking adrenal gland of 6 cm. Microscopically, it consists of pleomorphic large polygonal cells arranged in nests bounded by delicate fibrovascular stroma. The cells had granular eosinophilic cytoplasm and moderate-to-severe nuclear pleomorphism, and mitosis cannot be found. Special stains such as AE1/AE3 and chromogranin A were positive with focal moderate positivity for synaptophysin. Finally, it was diagnosed as adrenal neoplasm (oncocyctic pheochromocytoma with none malignant features). Postoperatively, blood pressure settled and his blood sugar stabilized over few months with diet regimen and using metformin as outpatient treatment [Figure 2b].

Discussion

RDS is rare disease and usually involves lymph-nodes, but may involve extra-nodular lesions.^[1] The main way of treatment is surgical excision of the mass, steroid, chemotherapy, radiotherapy and others.^[15-19] The severity of the high blood pressure and blood sugar before surgical removal of the adrenal mass indicate the high activity of sympathetic neuroendocrine activity of the

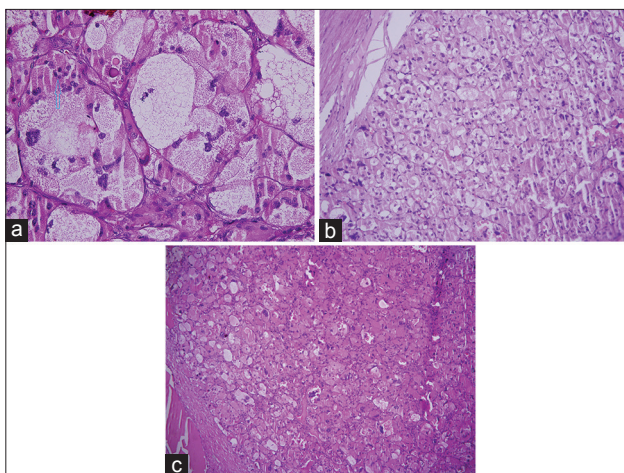


Figure 1: Right inguinal lymph node excision biopsy, stained with hematoxylin and eosin, under power (panel a) ×100, (panel b) ×40, and (panel c) ×20 magnification, showing sinus histiocytosis and characteristic emperipolesis

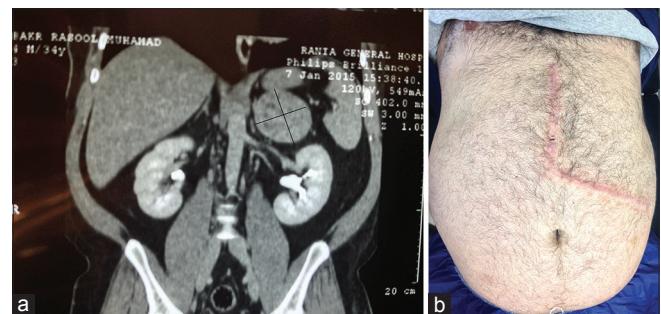


Figure 2: (a) Computerized tomography of a 43-year-old male, with mass between spleen and left kidney showing an adrenal mass. (b) Panel B: Showing an abdominal scar

pheochromocytoma. The response was dramatic after surgery; the patient still remained on Metformin and diet regime. We reviewed literature but we couldn't find significant relation between these two diseases. This condition needs more investigation to prove the relation, or it may be a coincidental presentation.

Conclusion

Here, we are reporting a very rare case of RDS in a middle-aged Kurdish male who presented with recurrent LN enlargement and concomitant presentation with large adrenal mass with features of pheochromocytoma; the patient is treated medically and surgically with a very good response.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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