Renal involvement in Rosai Dorfman disease, Rare case with diagnostic conundrum

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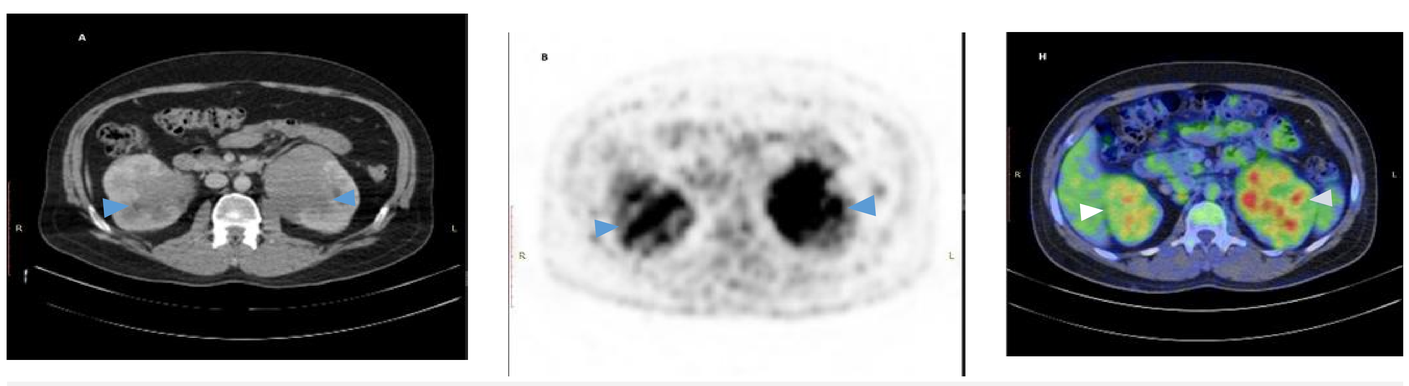
Introduction:

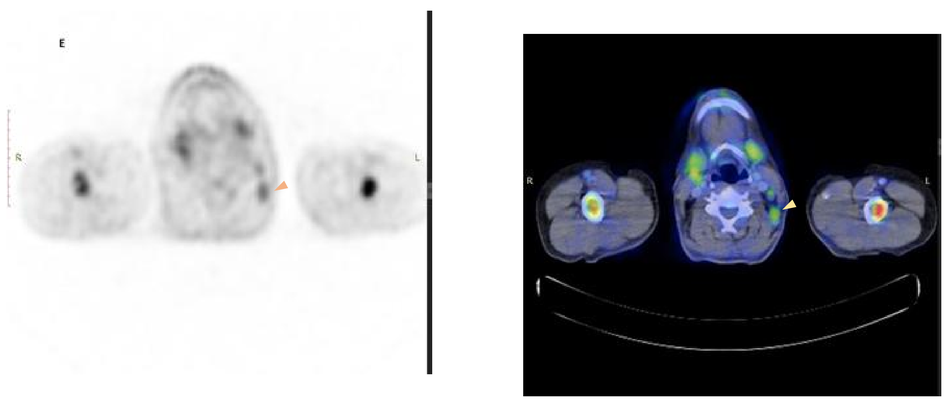
Rosai-Dorfman disease (RDD) is a rare, benign histiocytic proliferative disorder commonly present with painless cervical lymphadenopathy. Extra nodal involvement occurs in few cases, with renal manifestations being exceedingly rare serious manifestation of Rosai-Dorfman disease (RDD), occurring in about 4% of cases and associated with a poor prognosis.RDD. Renal RDD poses significant diagnostic challenges due to its nonspecific clinical and radiologic features. This case report is intended to improve the understanding of clinical radiologist and physicians on a rare RDD disease of the kidneys with lymphadenopathy and describing the imaging and histological features of the disease.

Presentation and imaging:

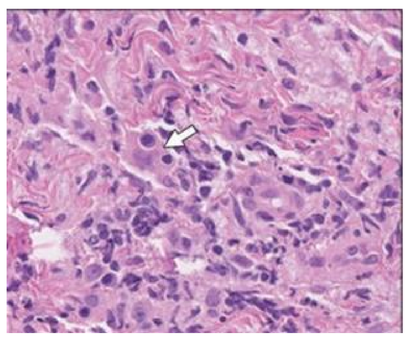
51-year-old male who presented with progressive fatigue and generalized weakness for six months.
No history of fever,hematuria,flank pain or hypertension.No past medical history of TB.
On examination bilateral enlarged lymph nodes in cervical chains were noted.
Initial imaging CECT and PET CT revealed bilateral renal masses and abdominal lymphadenopathy.



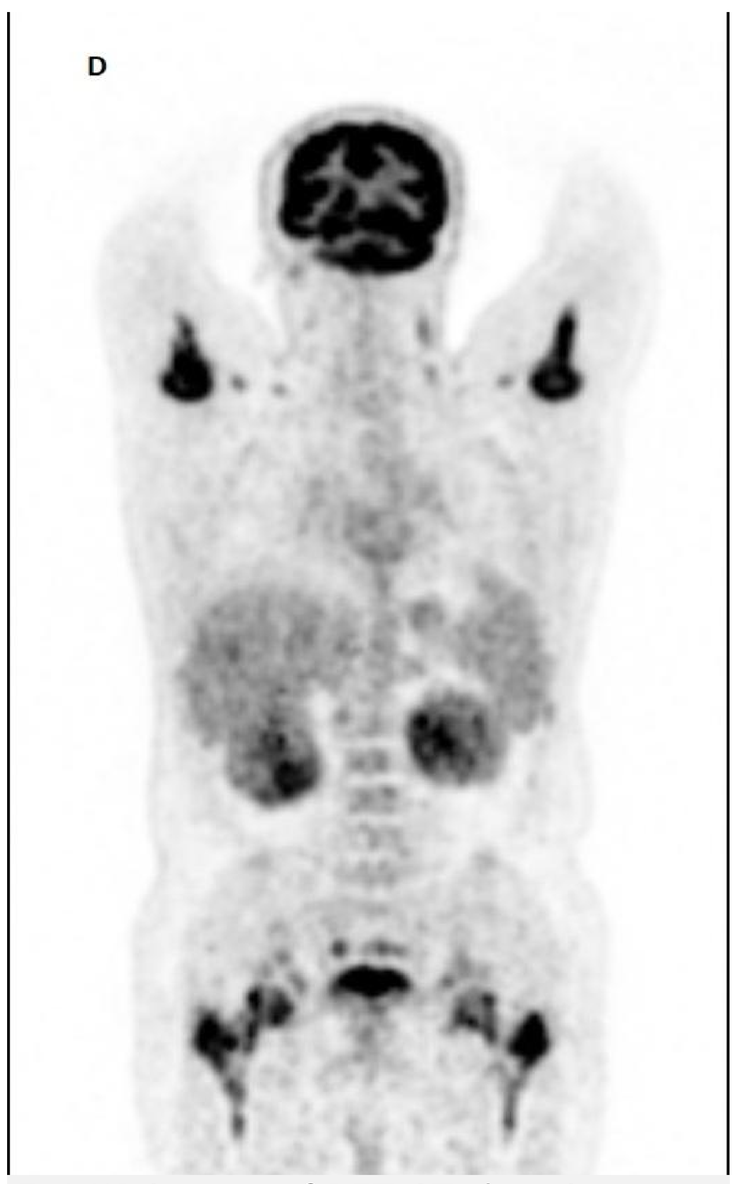
PET CT fused images revealed hyper-metabolic bilateral renal masses with FDG avid para aortic enlarged lymph nodes



D;Fused PET CT images show hyper metabolic enlarged cervical lymph nodes



Photomicrograph (H and E, ×400) shows large histiocyte (arrow) with engulfed lymphocyte and plasma cell, known as emperipolesis, in background of histiocytes, lymphocytes, and plasma cells.



E; MIP image of PET CT showing hyper metabolic renal masses, cervical lymph nodes and multifocal marrow uptake in proximal femora and humeri

Discussion and teaching points:

- Roasi dorfmen disease is also called sinus Histiocytosis with lymphadenopathy is a rare idiopathic non Langerhans cell reactive histiocytic . This heterogeneous syndrome has different various prototypes that include sporadic, familial, and cutaneous.
- Extra nodal RDD has been reported in 43% of cases and usually occurs in older adult patients with nodal involvement. RDD has a predominantly systemic involvement, in which skin, bone, and soft tissue are most frequently involved. Kidney involvement is less common, accounting for 4% of cases
- Rosai Dorfmen disease benign a multi organ disorder, the clinical and the radio logical features depending on the site of involvement.
- RDD is mostly benign and self-limiting, the various clinical manifestations and non-specific imaging results make the diagnosis challenging and difficult to differentiate from RCC, lymphoma, and metastasis. FDG PET/CT is beneficial for initial staging and treatment response in patients with RDD.
- The definite diagnosis is currently on basis of histopathology. The characteristic include numerous enlarged histiocytes with emperipolesis which engulfed intact inflammatory cells in the cytoplasm, a characteristic finding of RDD. On the immunohistochemical study, the histiocytes were positive for S100 protein and CD68
- The diagnostic challenge in this patient was the rarity of presentation of bilateral renal masses with lymphadenopathy with overlapping features of lymphoma made it essential to use a combination of imaging and immunohistological markers to arrive at a correct diagnosis

Conclusion This case highlights an unusual presentation of RDD with bilateral renal involvement and enlarged cervical and abdominal lymphadenopathy, emphasizing the importance of including RDD in the differential diagnosis of renal masses.Recognition of this rare entity is crucial to avoid misdiagnosis and unnecessary aggressive treatment.

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