

A Rarest of Rare Presentation of ROSAI - DORFMAN Disease Involving Para Nasal Sinuses with Massive Lymphadenopathy and Renal Mass

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Abstract: *Introduction: Lampert and Lennert in 1961 were the first to describe what is now known as Rosai-Dorfman disease (RDD). Subsequently in 1969 Rosai and Dorfman described 4 cases of a disease they called sinus histiocytosis with massive lymphadenopathy (better known as RDD). Case: A 23-year-old man presented with loss of appetite, significant weight loss (10 kg in last 6 month) and dull left flank pain for 6 month. There was past history of chronic nasal polyps and sinusitis for which he underwent nasal sinus surgery (Three times in last six years). Physical examination revealed averagely nourished individual without peripheral lymphadenopathy and organomegaly. Conclusion: Involvement of the paranasal sinuses, kidney and para aortic reteroperitoneal lymph node as encountered by us is an extremely rare presentation even not reported so far in literature of Rosai-Dorfman disease. The diagnosis is primarily based on histopathological one and confirmation by IHC- CD68, S-100 positivity and CD1a negitivity.*

Keywords: ROSAI-DORFMAN Disease, Histiocytosis, Lymphadenopathy, Nephrectomy, Paranasal Sinuses

1. Introduction

Lampert and Lennert in 1961 were the first to describe what is now known as Rosai-Dorfman disease (RDD). Subsequently in 1969, Rosai and Dorfman described 4 cases of a disease they called sinus histiocytosis with massive lymphadenopathy (better known as RDD) [1].

RDD generally manifests in children or young adults with massive cervical lymphadenopathy, fever, leukocytosis, and increased erythrocyte sedimentation rate [2] other lymphatic groups such as mediastinal, axillary and inguinal lymph nodes can also be affected.[3] Extranodal RDD lesions may occur with or without lymphadenopathy and may be solitary or multiple.[4]

2. Case

A 23-year-old man presented with loss of appetite, significant weight loss (10 kg in last 6 month) and dull left flank pain for 6 month. There was a past history of chronic nasal polyps and sinusitis for which he underwent nasal sinus surgery three times in last four years. On biopsy sinus histiocytosis was reported. Physical examination revealed averagely nourished individual without peripheral lymphadenopathy and organomegaly.

Laboratory examination revealed, Hemoglobin was 9.2 g/dL, Leucocyte count of 9, 800, ESR was significantly increased up to 112 mm/hour, Serum creatinine 1.18 mg/dl, mean corpuscular volume was 69.5 fL, mean corpuscular hemoglobin was 26.2 pg and a peripheral blood smear showed microcytic hypochromic red blood cells. All other laboratory studies were normal.

CECT PNS: shows complete soft tissue opacification of bilateral frontal, ethmoid, sphenoid, and right maxillary sinus. polypoidal mucosal thickening noted in left maxillary sinus and bilateral nasal cavities reaching posteriorly upto choana.

CECT Thorax, Abdomen & Pelvis: A large well defined homogenous mass of density 30 to 40 HU and size of 84x73mm in axial plane and cranio caudal extension of 94x82 mm arises from left renal mid and lower pole an, interpole region.

With the presumed diagnosis of renal cell carcinoma, left open radical nephrectomy was done.

3. On HPE Biopsy Report

Macroscopic 1. Lt radical nephrectomy specimen shows a 10.0x 8.5x 5.5 cm tumour involved lower and middle pole of kidney without extra renal involvement.(Figure-3)**2.** Para aortic lymph node.

Microscopic: 1. Section from mass shows infiltration by histiocyte cells, lymphocyte and plasma cells. there are presence of cells showing lymphocytrophagocytosis {emperipolesis}. perinephric fat, ureter, renal vessels and adrenal glands are not affected by these cells. total two perinephric lymph node are identified which are showing sinus histiocytosis.(Figure-4)**2.** Total four lymph nodes are identified. All are showing sinus histiocytosis. **3.** Biopsy from maxillary sinus polyps shows marked lymphoplasmacytic infiltrate and large histiocyte with engulfed lymphocyte and plasma cells { Emperipolesis} suggestive of Rosai – Dorfman's disease.

Immunohistochemical staining with S100 protein CD 68 revealed strongly stained histiocytes cells.

4. Discussion

Sinus histiocytosis is a diffuse, lymphoproliferative disorder involving numerous organs that occurs most often in children or young adults, although patients in their 7th decade have also been described. Associated symptoms and signs may be caused by specific organ involvement or may be constitutional, such as fever and weight loss.

Laboratory findings include anemia, leukocytosis and serum polyclonal hypergammaglobulinemia. Although early descriptions concluded that nearly every case was marked exclusively by cervical lymph node involvement, other organ systems may be affected including the eye and eyelid, bone, central nervous system, ear, nose, throat, upper respiratory tract, liver, skin, salivary gland and testis. [5]

Kidney involvement is very uncommon, and therefore sinus histiocytosis is not frequently considered in the differential diagnosis of an infiltrative renal mass. Five cases of RDD in kidney cases have been reported with two of them diagnosed with adenocarcinoma of the prostate..

In a rare case reported by Buchino et al, the kidney was only focally involved with a single small mass in the lower pole that contained an admixture of histiocytes, lymphocyte and plasma cells. [6] In another case reported by Bechtold et al, a lobular irregularly enlarged kidney with distorted calyces associated with large matted para-aortic lymph nodes was described and the diagnosis [8] Grossly the masses are matted together by prominent perilesional fibrosis. Their cut surface varies from gray to golden yellow, depending on the amount of fat present. Microscopically there is an accumulation of lymphocytes, plasma cells (some containing Russell bodies), and most notably numerous cells of histiocytic appearance with a large vesicular nucleus and abundant clear cytoplasm that may contain large amounts of neutral lipids. Many of these histiocytes have within their cytoplasm numerous intact lymphocytes, a feature that has been designated as emperipolesis or lymphocyte phagocytosis.[6, 7]

The differential diagnosis of RDD in kidney includes malignant fibrous histiocytomas and histiocytic proliferations of infectious etiology (the presence of S100 is useful in discriminating these lesions), leukemia or lymphoma, especially when accompanied by lymphadenopathy (absent of emperipolesis and IHC profiles help to correct diagnosis). Other possible differential diagnoses include storage disease, tuberculosis or even renal cell carcinoma, a metastatic tumor such as malignant melanoma. RDD generally has a favorable prognosis, but involvement of a greater number of nodal groups and associated extranodal systems worsens the prognosis. No intervention is necessary in most cases, but some patients may undergo surgery. In disseminated aggressive cases, chemotherapy and external beam radiotherapy may be used.[8, 9].

This is first case so far according to literature in which simultaneous involvement of kidney, lymph node and paranasal sinuses detected.

5. Conclusion

Involvement of the paranasal sinuses, lymph node and kidney as encountered by us is an extremely rare presentation even not reported so far in literature of Rosai-Dorfman disease. The diagnosis is primarily a histopathological one and confirmation by IHC-CD68 S-100 positivity.



Figure 1

Complete soft tissue opacification of bilateral frontal, ethmoid, sphenoid, and right maxillary sinus. polypoidal mucosal thickening noted in left maxillary sinus and bilateral nasal cavities reaching posteriorly upto choana.

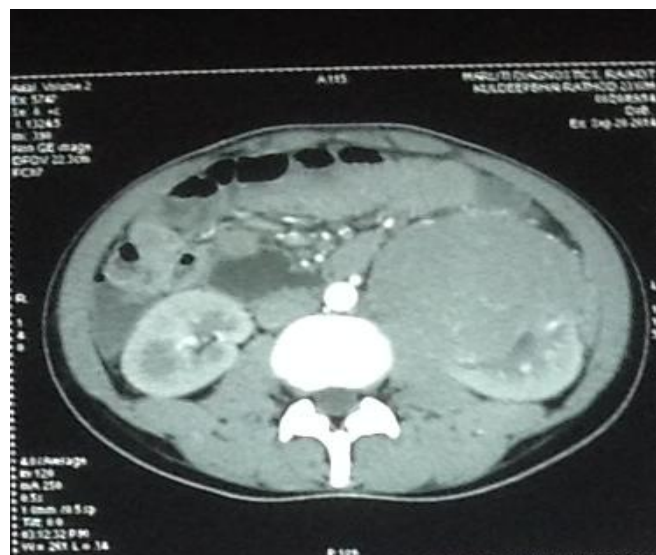


Figure 2

A large well defined homogenous mass of density 30 to 40 HU and size of 84x73mm in axial plane and cranio caudal extension of 94x82 mm arises from left renal mid and lower pole an, interpole region.

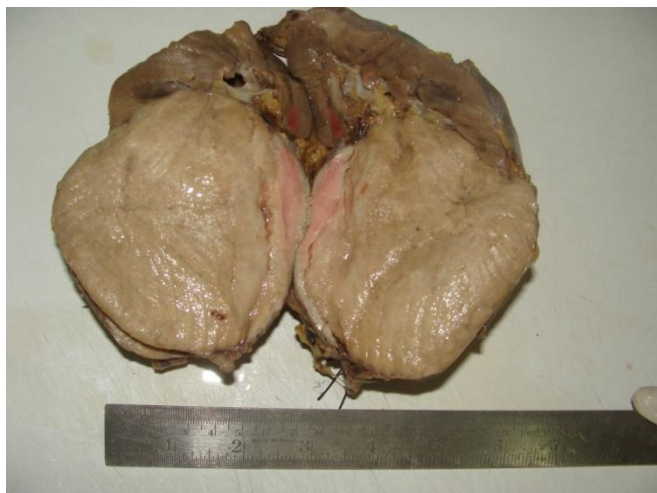


Figure 3

Lt radical nephrectomy specimen shows a 10.0x 8.5x 5.5 cm tumour involved lower and middle pole of kidney without extra renal involvement

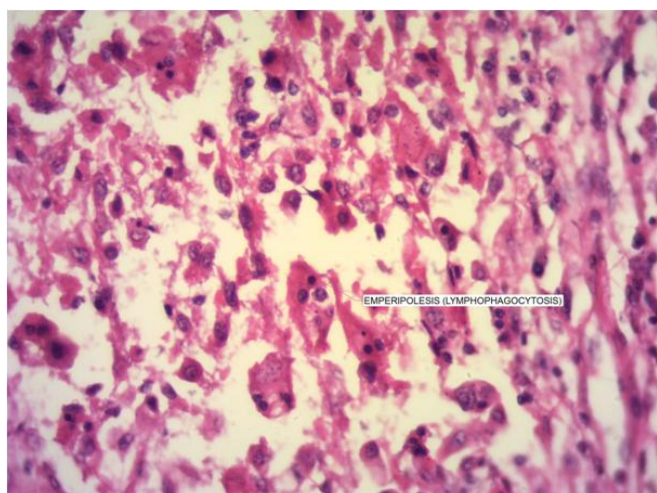


Figure 4

Section from mass shows infiltration by histiocyte cells, lymphocyte and plasma cells. there are presence of cells showing lymphocytophagocytosis {emperipolesis}.

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