Case 15497

Eurorad

Rosai–Dorfman disease: Nasal and cervical manifestation

Published on 09.02.2018

DOI: 10.1594/EURORAD/CASE.15497
ISSN: 1563-4086
Section: Neuroradiology
Area of Interest: Head and neck
Procedure: Computer Applications-Detection, diagnosis
Imaging Technique: CT
Special Focus: Inflammation Case Type: Clinical Cases
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Patient: 20 years, male

Clinical History:

A 20-year-old male patient presented with history of nasal obstruction and loss of smell symptoms. The patient presents nasal respiratory insufficiency and bilateral rhinorrhoea, as well as the presence of multiple cervical lymphadenopathies. Marked oedema of the nasal mucosa was observed in fibroscopy as well as the presence of nasal polyps.

Imaging Findings:

Computed tomography of paranasal sinuses without intravenous contrast with axial and coronal reconstructions and bone a soft tissue windows, show nasal cavity occupation with an irregular surface of the mucosa compatible with polyposis. There is also a maxillary occupation of the left sinus and a mucosal thickening of ethmoidal cells.

Cervical computed tomography with intravenous contrast shows multiple bilateral adenopathies in laterocervical spaces that predominate at level Ib, IIa and IIb. All of them show homogeneous uptake contrast, without necrosis nor extracapsular extension.

Discussion:

Rosai–Dorfman disease (RDD) is a rare histocytic proliferative disorder. The aetiology and natural history of RDD remain uncertain. Possible causes include altered immune responses and infections [1]. The natural history and outcome of this disease are variable [2]. The disease has a slight predilection for males and primarily affects children, adolescents, and young adults, 80% of cases occurring in individuals under 20 years of age [3, 4].
Its symptomatology is non-specific, with a variable degree of systemic involvement (fever, weight loss, weakness, anaemia, leukocytosis), with the presence of bilateral and non-painful cervical adenopathies being the initial manifestation in more than 90% of cases. Patients show coarse, painless neck lymphadenopathy, which is usually bilateral. An extranodal manifestation has been described in 43% of patients; in 75% of these patients, the region of the head and neck was involved. In the case of extranodal involvement of the head and neck, about 50% have a manifestation in more than one region. In 73% of cases, the upper airways are affected, in 50% the orbits and in 25% the salivary glands [5, 6]. Sinonasal RDD is an uncommon clinical entity, of the cases described in the literature to date, approximately 11-16% show nasosinusal disease [6-8]. The presentation can be as a submucosal infiltrative lesion or more frequently in pseudotumoral form without bone destruction, simulating a banal polyposis.
The diagnosis is established by biopsy of lymph nodes, and/or other organs affected in each case. Histology of

lymph nodes shows dilated sinuses with numerous histiocytes that exhibit emperipolesis. Immunohistochemical features include positivity for S100 protein and CD68, and negativity for CD1a. In extragnaglionic regions, there is greater fibrosis and less emperipolesis [9].
The natural history of this disease is generally described as benign. However, recurrence and fatal outcomes have been reported [7, 10]. The factors that determine the recurrence or progression of RDD are not well understood . Patients with extranodal involvement (multisystemic extension, based on liver, kidney, pancreatic, respiratory tract, heart and immune involvement, or anaemia) have worse prognosis. [7, 10-14].
No ideal treatment for sinonasal RDD nor for patients with RDD in general has yet to be established. Radiotherapy, chemotherapy, and steroids, used alone or in combination, have been reported with variable responses. Although resection is often successful at relieving symptoms, local recurrence or disease progression after surgery may occur [14, 15]. However, the factors that determine recurrence and the frequency of sinonasal RDD are not well understood. [16]
Differential Diagnosis List: Rosai-Dorfman disease with nodal and extranodal extension affecting nasal cavities, Lymphoma, Tuberculosis, Castleman disease, Kaposi sarcoma, Metastatic thyroid carcinoma

Final Diagnosis: Rosai-Dorfman disease with nodal and extranodal extension affecting nasal cavities

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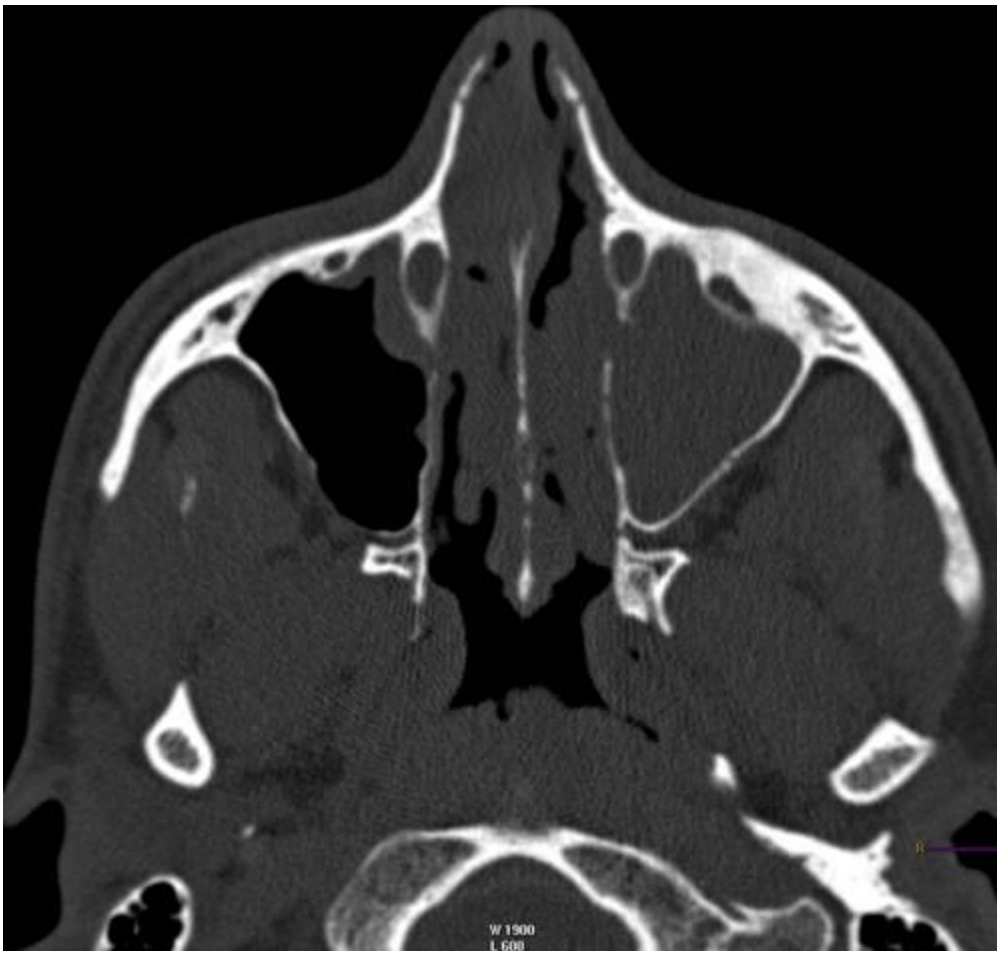
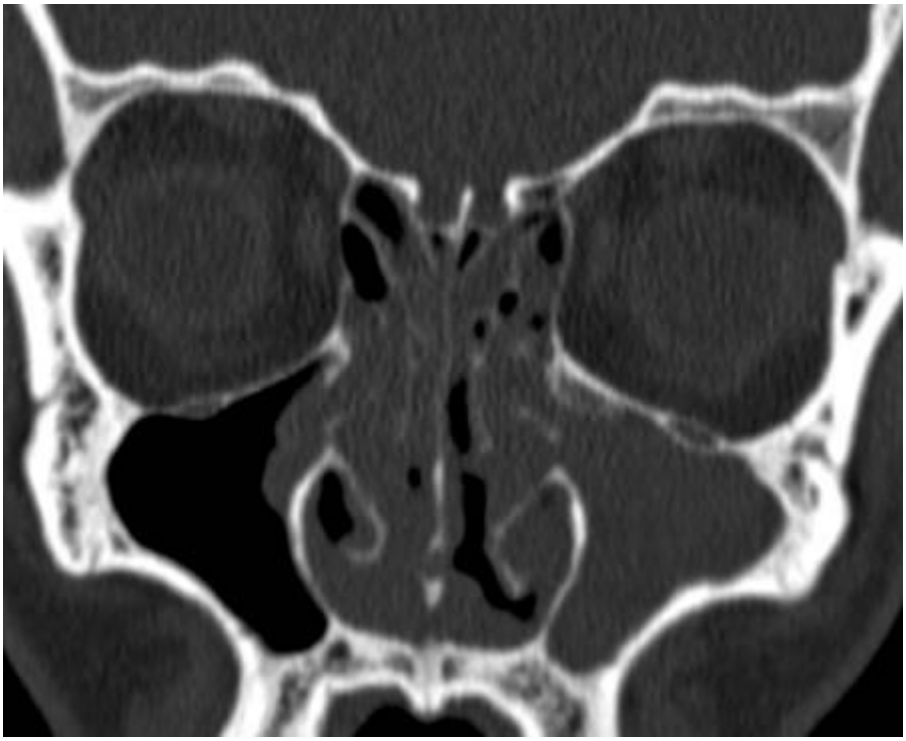
Figure 1

a



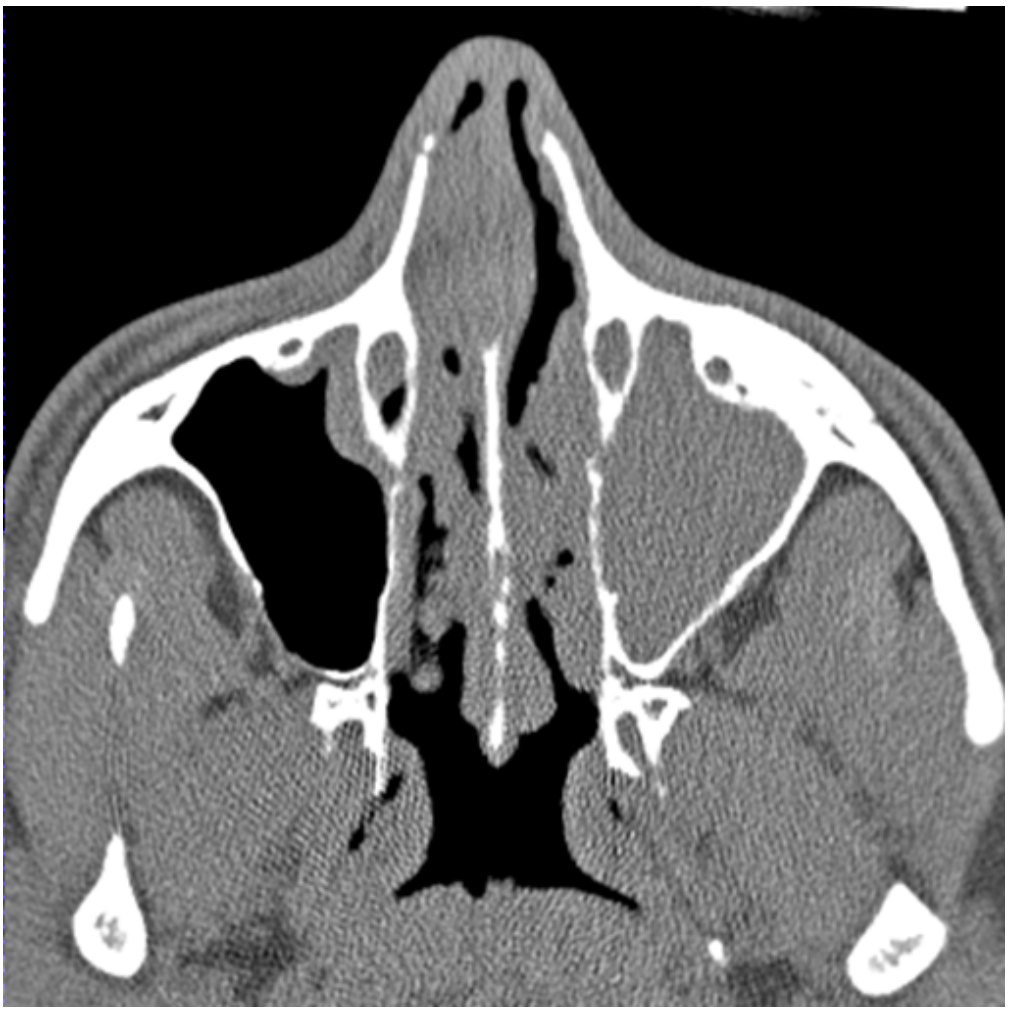
Description: Axial CT images in bone show opacification of left maxillary sinus and bilateral multiple polypoid
masses filling nasal cavities and middle meati **Origin:** Salvador E, Department of Radiology, Madrid, Spain.

b



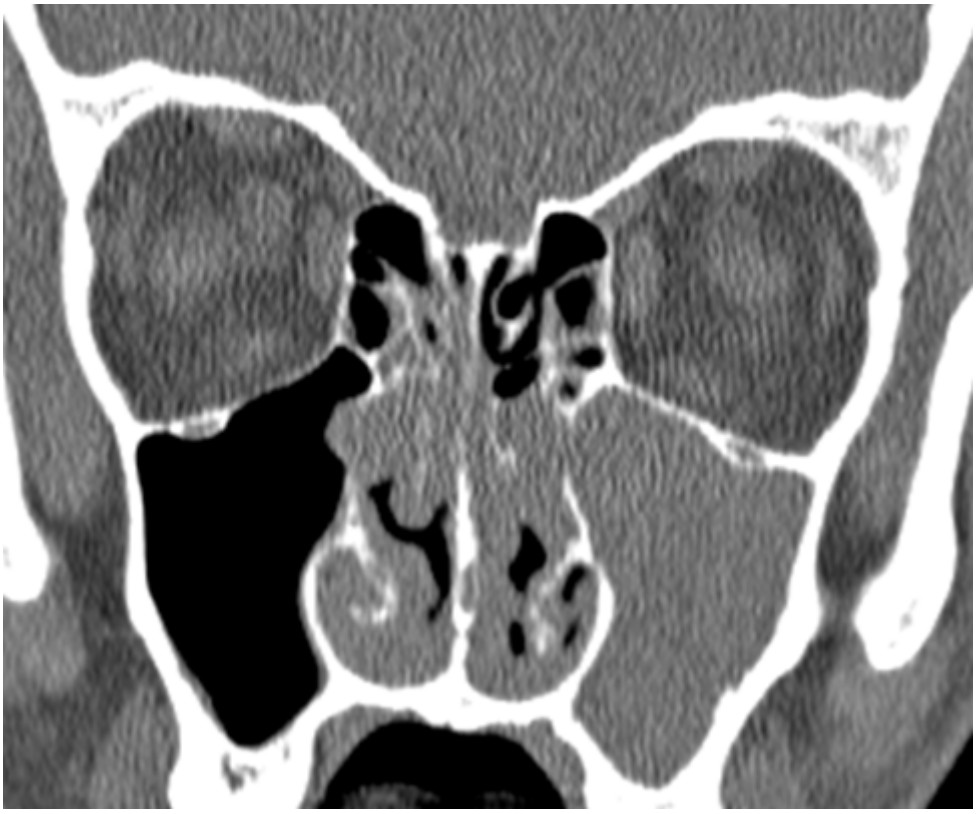
Description: Coronal CT bone images show sinonasal polyposis with complete opacification of the left maxillary sinus with deossification of the ethmoid trabeculae and widening of the ethmoid infundibuli and osteomeatal units. **Origin:** Salvador E, Department of Radiology, Madrid, Spain.

c



Description: Axial CT images with the soft tissue algorithm show nasal polyposis and opacification with secretions in the left maxillary sinus. **Origin:** Salvador E, Department of Radiology, Madrid, Spain.

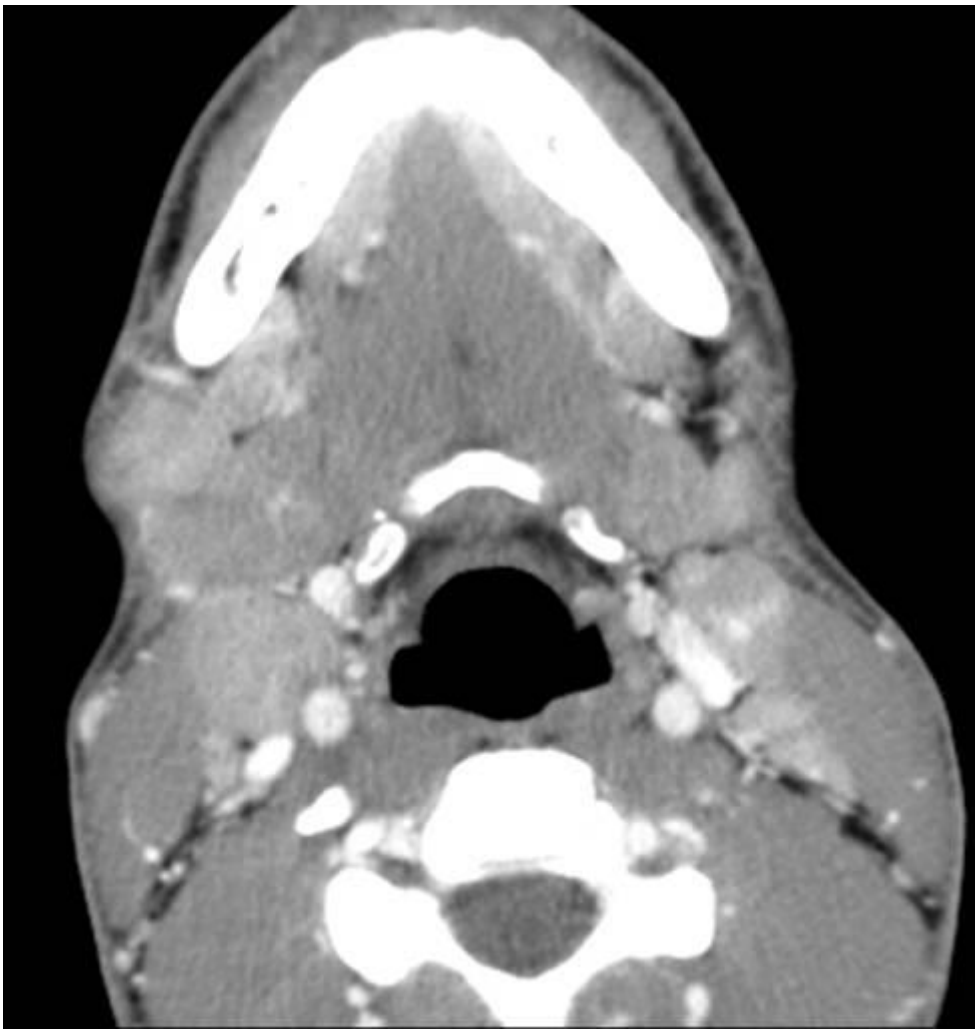
d



Description: Coronal CT images with the soft tissue algorithm show bilateral nasal polyposis and opacification in the left maxillary sinus and widening wiyh obstruction of the left osteomeatal unit.
Origin: Salvador E, Department of Radiology, Madrid, Spain.

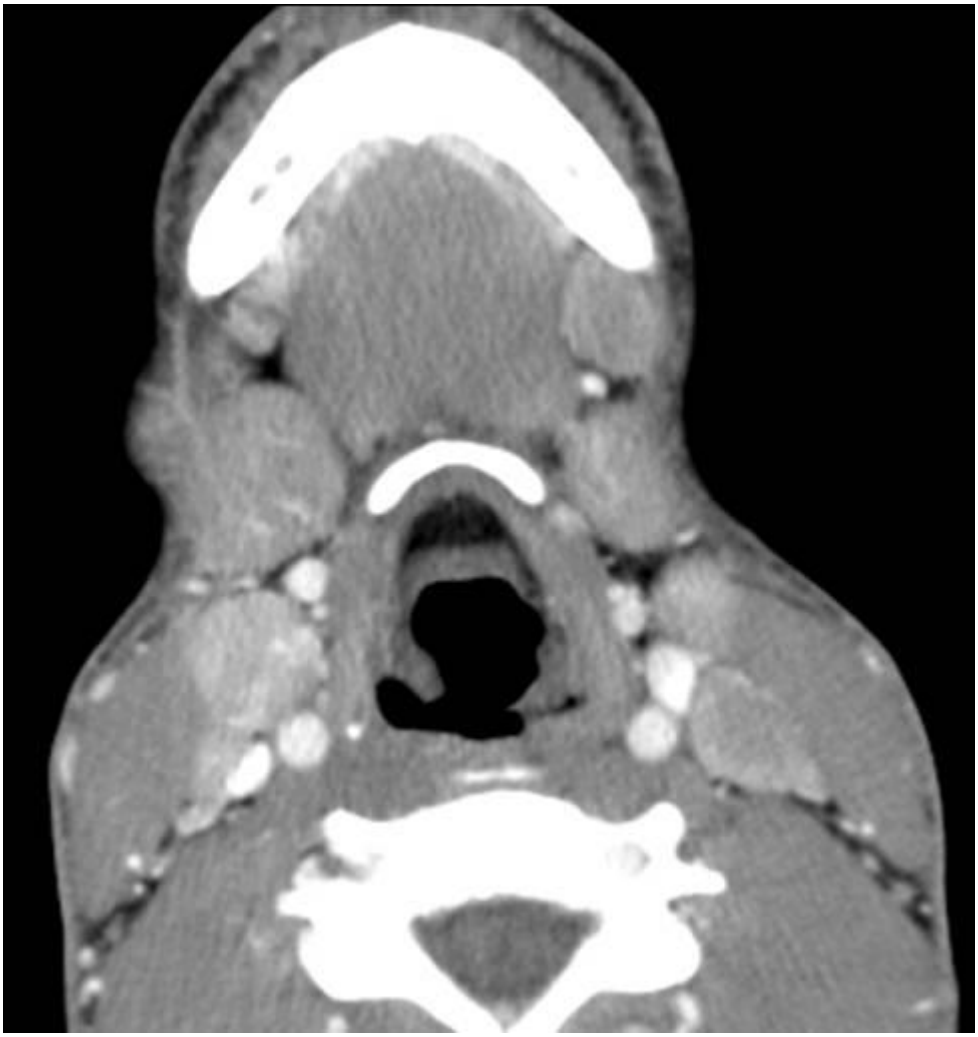
Figure 2

a



Description: Axial contrast-enhanced cervical CT show bilateral level IIB and IB enlarged lymph nodes, all of them show marked homogenous contrast enhancement. **Origin:** Salvador E, Department of Radiology, Madrid, Spain.

b



Description: Axial contrast-enhanced cervical CT show bilateral level IIB and IB enlarged lymph nodes, all of them show marked homogenous contrast enhancement. **Origin:** Salvador E, Department of Radiology, Madrid, Spain.