Texas Society of Neuroradiology (TSNR)
Educational Abstract
2026 Annual Meeting – Dallas, TX
February 21–22, 2026

Extranodal Rosi-Dorfman Disease of the Head and Neck: A Pictorial Review of Your Friendly Tumor Mimic

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Summary

Extranodal Rosai-Dorfman disease (RDD) of the head and neck (H&N) is an uncommon, benign histioproliferative disorder that frequently mimics aggressive neoplasms on imaging, leading to diagnostic uncertainty. This pictorial review presents a large, multi-institutional series of pathologically confirmed extranodal RDD cases involving the H&N, illustrating their CT, MRI, and 18F-FDG PET/CT imaging appearances across a wide range of anatomic subsites. Through a case-based, image-rich approach, we highlight characteristic imaging patterns, common sites of involvement, and key features that overlap with malignant processes. Emphasis is placed on recognizing imaging clues, understanding typical disease distribution, and incorporating RDD into the differential diagnosis to facilitate accurate diagnosis and appropriate clinical management.

Educational Objectives

After reviewing this exhibit, the learner will be able to:

1. *Recognize the common anatomic locations and multimodality imaging features of extranodal RDD in the head and neck on CT, MRI, and 18F-FDG PET/CT.*
2. *Differentiate extranodal RDD from common malignant and inflammatory mimickers of the head and neck based on key imaging characteristics and disease distribution.*
3. *Apply imaging findings, including the role of PET/CT, to improve diagnostic confidence, assess disease extent, and guide appropriate clinical workup.*

Purpose

RDD is a rare, idiopathic, and non-neoplastic histioproliferative disease. It can affect many organ systems, including nodal (70%) or extranodal (30%) sites. Extranodal involvement can occur anywhere in the body, including the H&N. It can exhibit imaging features that mimic aggressive neoplasms, creating a diagnostic

challenge. Here, we do an image-rich review to present the CT, MRI, and 18F-FDG PET/CT imaging characteristics of extranodal RDD in the H&N, and comprehensively discuss differential diagnoses and key teaching points.

1. To review and illustrate the imaging characteristics of extranodal RDD in the H&N, which can present

with features mimicking aggressive neoplasms.

2. To demonstrate various extranodal RDD masses of the H&N by specific anatomic location in a case-based approach.
3. To discuss the differential diagnoses and emphasize the key imaging findings leading to correct diagnosis.

Materials and Methods

We retrospectively reviewed pathologically confirmed RDD cases seen at two academic institutions from 2006 to 2025. Only extra nodal RDD cases were analyzed for anatomical locations and radiologic appearance. Also, the mimickers were recorded.

Results

35 cases of extranodal RDD in the H&N (22 female and 13 male, mean age 46.5 years [range 4-81]) were included. Specific sites of involvement included the following: pituitary, n = 1; lacrimal gland, n = 2; orbit, n=5; paranasal sinus, n = 12; salivary gland, n = 3; vocal cord, n=3; epiglottis and bilateral aryepiglottic fold, n=1; skin, n = 2; nasal cavity/septum, n = 13; skull-base, n=1; nasopharynx, n=2, Meckel cave, n=1; hard palate, n=2; maxillary alveolar ridge and gingiva, n=1; and clavicle, n = 1. Sixteen patients had isolated extranodal RDD disease, the others also had lymph nodes and other organ involvements outside the H&N. The most affected site was the nasal cavity, followed by the paranasal sinuses and orbit, respectively, in extranodal RDD of the H&N.

Conclusion

The clinical presentation and radiological imaging findings of extranodal RDD of the H&N overlap significantly with other neoplastic processes of the H&N. Although the diagnosis is challenging to make prospectively, including RDD in the differential when appropriate may help guide workup. PET/CT, in particular, is very helpful to evaluate the extent of the disease, and when there is involvement of multiple subsites, it can help increase the specificity of diagnosis.

References

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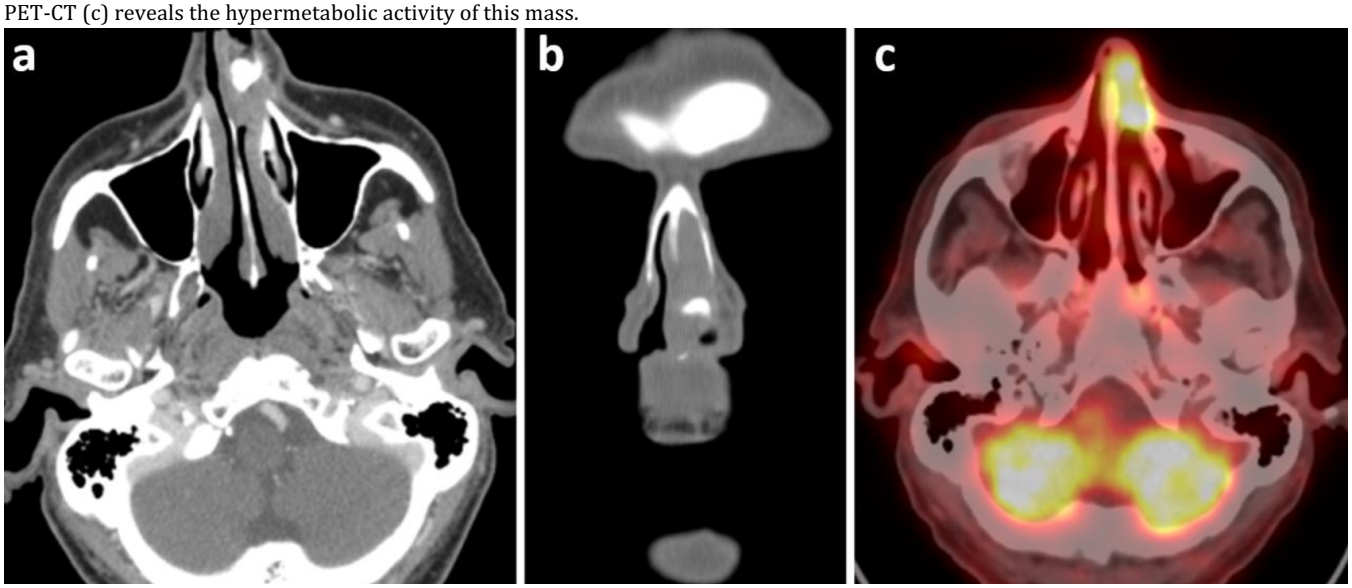
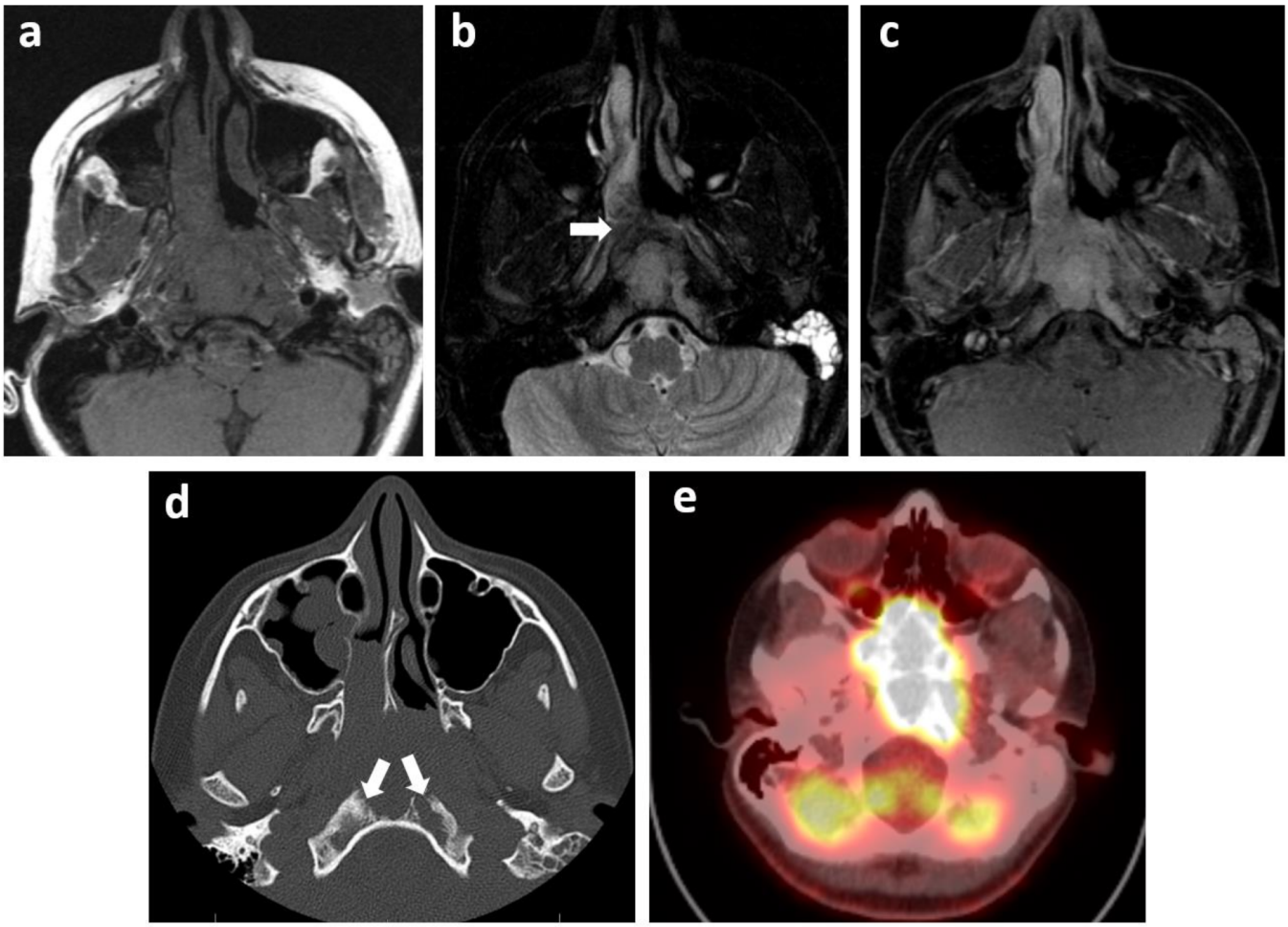
Figures

Figures 1: Axial (a) and coronal (b) CT images of the sinuses demonstrate a soft tissue mass that occupies much of the lower left nasal cavity and nasal aperture, with more inferiorly, a calcific tumor component. PET-CT (c) reveals the hypermetabolic activity of this mass.



Figures 2: Axial T1-weighted (a), T2-weighted (b), and postcontrast T1-weighted MRIs demonstrate T1 isointense, T2 iso to hyperintense contrast-enhanced an extensive mass lesion involving the roof of the nasopharynx and central skull base. This mass lesion extends through the choana into the right posterior nasal cavity. Also, there are hypointense areas in the lesion on T2 WI (b; arrow). CT demonstrates bone erosion of the clivus (d; arrows). PET-CT (e) reveals the hypermetabolic activity of this mass.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