Texas Society of Neuroradiology (TSNR)
Educational Abstract
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Pictorial Review of a Surprising Tumor Mimic of the Central Nervous System: Extranodal Rosai-Dorfman Disease

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Summary

Rosai–Dorfman disease (RDD) is a rare, non-neoplastic histiocytic disorder that infrequently involves the central nervous system (CNS). Extranodal CNS RDD can closely mimic neoplastic processes, particularly dural-based tumors, resulting in frequent preoperative misdiagnosis. This educational exhibit provides an image-rich, case-based review of pathologically confirmed extranodal CNS RDD from two academic institutions over an 18-year period. Various anatomic patterns of involvement—including dural, parenchymal, leptomeningeal, spinal, and sellar regions—are illustrated. Key imaging features are highlighted and compared with common disease mimickers to improve diagnostic recognition and confidence.

Educational Objectives

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

1. Recognize the spectrum of imaging appearances and anatomic distributions of extranodal Rosai–Dorfman disease involving the CNS.
2. Differentiate extranodal CNS RDD from common mimickers such as meningioma, lymphoma, inflammatory, and other dural-based or parenchymal neoplasms based on key imaging characteristics.
3. Understand the clinical and diagnostic challenges of isolated CNS RDD and identify imaging clues that may prompt consideration of this rare entity in the differential diagnosis.

Purpose

Rosai Dorfman Disease (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 CNS, which is rare. It can exhibit imaging features that mimic

neoplasms, creating a diagnostic challenge. Here, we do an image-rich review to present the imaging characteristics of extranodal RDD of the CNS, and comprehensively discuss differential diagnoses and key teaching points.To review imaging-based level-based classification of cervical lymph nodes.

1. To review and illustrate the imaging characteristics of extranodal RDD of the central nervous system (CNS).
2. To demonstrate various extranodal RDD masses of the CNS 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 2024. Only CNS extra nodal RDD cases were analyzed for anatomical locations and radiologic appearance. Also, disease mimickers were recorded.

Results

15 cases of extranodal CNS RDD (7 male and 8 females, mean age 51.1 years [range 12-77]) were included. Specific sites of involvement included the following: dural-based intracranial, n = 8; dural-based spinal, n = 1; pituitary gland, n=1; epidural spinal, n= 1; spinal neural foraminal, n=1; cerebellar parenchymal, n = 2; and cerebellar and spinal leptomeningeal, n=1. Only three patients had additional extra CNS involvement, and all the others had isolated CNS disease. The most common CNS manifestation was dural-based lesions. There were only two parenchymal lesions (both with cerebellar involvement) and only one case with leptomeningeal involvement.

Conclusion

Here we present one of the largest series of extranodal RDD of the CNS. The diagnosis of extranodal RDD of the CNS remains challenging, and the condition is often misdiagnosed as other dural based neoplasms such as meningioma preoperatively. Less common presentations include leptomeningeal disease or intraparenchymal masses. Extranodal CNS RDD often presents as isolated disease of the CNS compounding an already challenging diagnosis.

References

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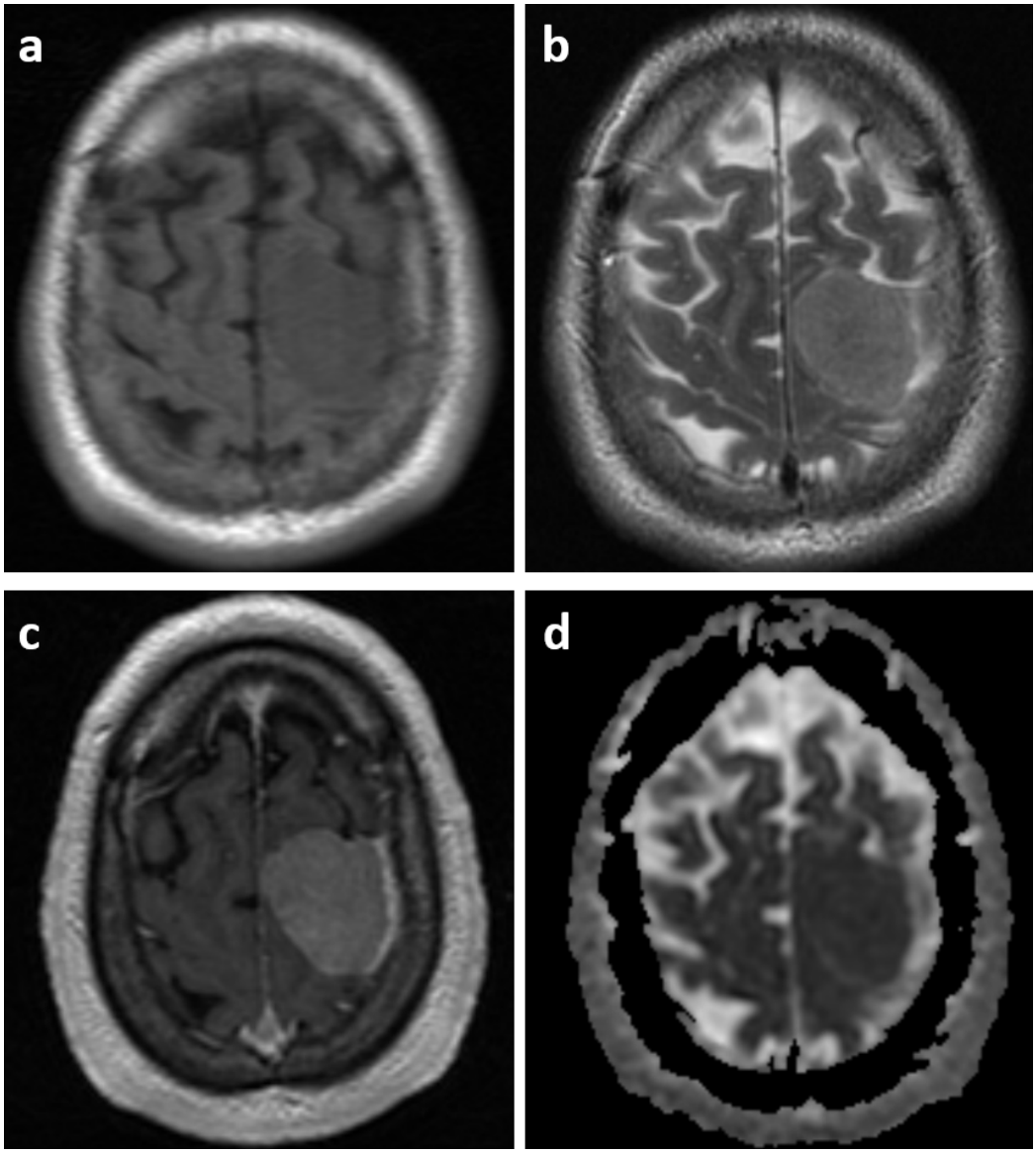
Figures

Figure 1. Axial T1-W (a), T2-W (b), and post-contrast T1-W (c) images demonstrate a T1 and T2 isointense left frontoparietal dural-based lesion with homogenous contrast enhancement mimicking meningioma. There is no obvious diffusion restriction on ADC map (d).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