

Case Report

Imaging findings of head and neck and intracranial sinus histiocytosis with massive lymphadenopathy (Rosai–dorfman disease): A rare case report

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Abstract:

This case report describes a multifocal involvement of Rosai–Dorfman disease (RDD), which is a benign lymphohistiocytosis, with emphasis on imaging findings in a 19 yrs old Ethiopian female patient who was sent to radiology department at Tikur Anbessa Specialized Hospital (TASH) for head and neck and brain imaging evaluations for an indication of bilateral painless neck swelling of 2 years duration. The head and neck computed tomography (CT) and brain magnetic resonance imaging (MRI) revealed numerous enlarged cervical lymph nodes and soft tissue infiltration of paranasal sinuses with associated lytic osseous changes as well as multiple intracranial extra-axial dural-based masses. Smears of fine needle aspiration biopsy from cervical lymph nodes confirmed the diagnosis of Rosai–Dorfman disease (RDD).

Keywords: Rosai- Dorfman disease, cervical lymph nodes, computed tomography, magnetic resonance imaging

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Introduction

Rosai-Dorfman disease (RDD) is a benign lymphohistiocytosis characterized by multiple enlarged cervical lymph nodes. RDD was first described by Desombes in 1965 and was first reported by Juan Rosai and Roland Dorfman as sinus histiocytosis with massive lymphadenopathy in 1969 (1). It is a very rare disease, with a prevalence of 1:200,000 with unknown etiology (2).

RDD tends to affect children and young people (1-3). Most of the patients (83%–95%) present with enlarged cervical lymph nodes (2). Up to 43% of patients show extranodal involvement including the soft tissues of the head and neck, paranasal sinuses, and nasal cavity (3). The central nervous system is involved in less than 5% of the cases, predominantly with extra-axial dural-based masses (1, 4).

The histopathologic findings of RDD include large pale histiocytes with eosinophilic cytoplasm in the background of plasma cells and lymphocytes with emperipolesis (lymphocytophagocytosis) seen in hematoxylin and eosin stain (4). We report imaging

findings of a rare case of RDD with head and neck and intracranial involvement.

Case Presentation

A 19-years-old Ethiopian female patient was referred to TASH for a complaint of bilateral painless neck swelling of 2 years duration for evaluation of head and neck computed tomography (CT) and brain magnetic resonance imaging (MRI). The patient also had multiple nodular facial skin lesions on presentations.

The head and neck CT imaging revealed numerous, bilateral, different sized, discrete and homogeneously enhancing enlarged cervical lymph nodes which involved all cervical lymph node levels. There was also a diffuse soft tissue infiltrative component seen involving the paranasal sinuses (Figure 1). Multiple dural-based intracranial extra-axial masses were also observed on the CT images.

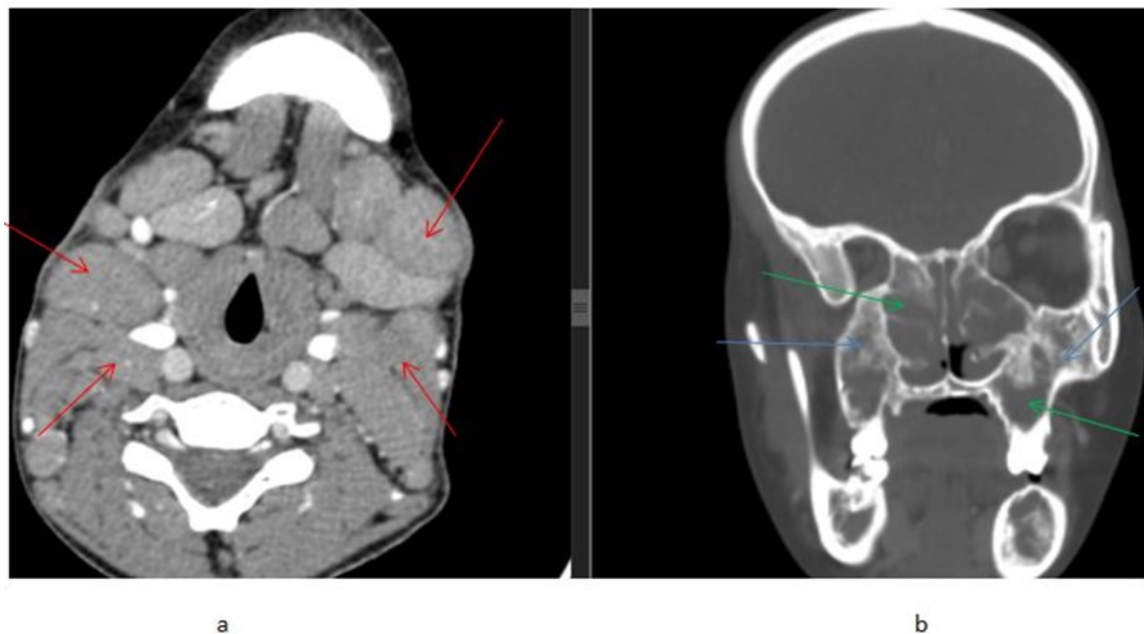


Figure 1 Head and Neck CT (a) axial soft tissue window show bilateral different sized numerous enlarged cervical lymph nodes (red arrows). (b) Coronal plane bone window show opacification of maxillary and ethmoid sinuses (green arrows) and lytic change of maxillary bone (blue arrows).

The head MRI with gadolinium-based intravenous contrast agent revealed multiple intracranial extra-axial well-defined dural-based masses which involve multiple intracranial compartments such as middle and posterior fossa. They involved the right cerebellopontine angle and parasellar region, extending to the right side of cavernous sinus, and the left side of middle cranial fossa. The extra-axial masses showed T1 isointense and T2 hypointense signal (Figure 2) relative to the cortical signal.

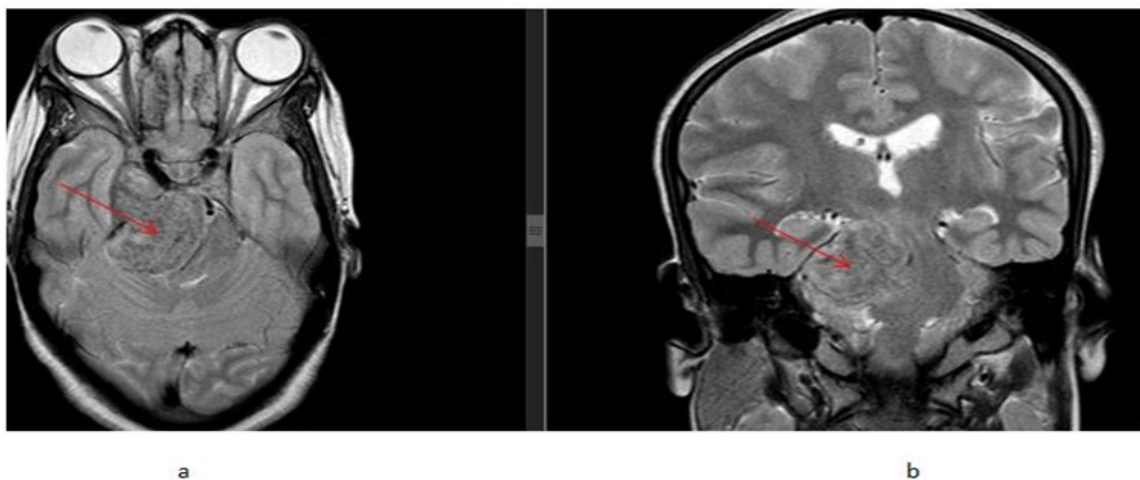


Figure 2: T2 brain MRI, (a) axial and (b) coronal planes showing right side multicompartmental extraaxial dural based hypointense masses (red arrows) in the right prepontine cistern, Meckel's cave and parasellar regions

The intracranial masses showed diffuse avid contrast enhancement on the post-contrast study. There was compression of the adjacent brain parenchyma but no obvious brain parenchymal infiltration or vasogenic edema seen. The intracranial masses also showed extracranial extension through the widened skull base

foramen (Figure 3). Chest x-ray and abdominopelvic ultrasound studies were done for this patient and showed no remarkable findings.

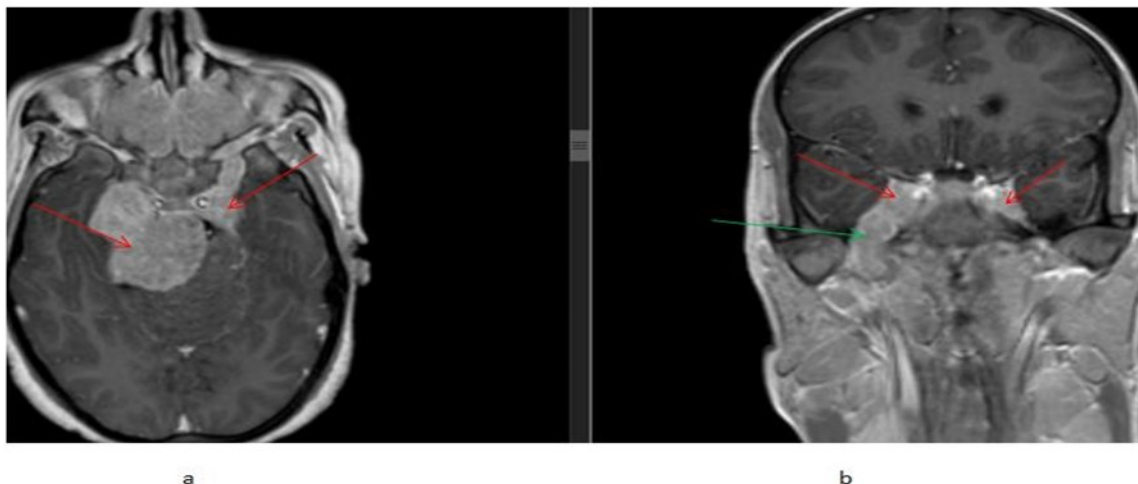


Figure 3 T1 post-contrast brain MRI, (a) axial and (b) coronal planes show multiple well-defined extra-axial dural based diffusely enhancing masses (red arrows) with extracranial extension through right foramen ovale (green arrow).

Fine needle aspiration cytology (FNAC) was done from bilateral neck lymph nodes and smear showed lots of macrophages containing numerous lymphocytes in their cytoplasm. The large histiocytic cells contain engulfed lymphocytes (emperipolesis¹⁸). The findings confirm the diagnosis of RDD.

Discussion

Our case revealed multifocal imaging findings of head and neck and intracranial involvement of a very rare benign disease. A young female patient with diagnosis of RDD after she presented with painless neck swelling and imaging revealed multiple enlarged cervical lymph nodes and intracranial dural based extraaxial masses. Differential diagnosis of lymphoma, metastases and multiple meningiomas were considered based on imaging findings and diagnosis was made only after FNAC was done.

The most common finding in patients with RDD is an enlargement of multiple cervical lymph nodes. This is similar to our case and well described in previous literature and case reports (1-5). This is also well described in previous two retrospective case review studies done by Vaidya et al and La Barge et al (2, 3). The soft tissue infiltration of paranasal sinuses and lytic osseous changes seen in our case, were also reported in previous case reviews (4, 5). The involvement of sinonasal regions is previously mentioned as the second most common site of involvement in RDD (2).

Intracranial involvement of RDD is less common. The common findings in intracranial involvement are multiple extra-axial enhancing masses. This is well described by H. Zhu *et al* and Raslan *et al* in their case reviews (2, 4). The intracranial masses in RDD as described by H. Zhu *et al* tend to be extra-axial, well-circumscribed, and dural-based with T2 iso- to hypointense signal with avid and homogeneous contrast enhancement on post-contrast study similar to our case (4).

The limitation of this case report is that we were not able to retrieve the pathology slides to include representative pathology slide images for we lost them.

Conclusion

RDD could mimic a number of pathologies on imaging, and one has to consider a differential diagnosis especially in patients with multiple cervical lymphadenopathy and other associated findings such as sinonasal soft tissue infiltrations and intracranial extra-axial dural-based masses.

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Competing Interests: The authors declare that this manuscript was approved by all authors.

Ethical clearance and patient consent: Informed consent was waived by the department's IRB as this case report involves analyzing existing data and poses no more than minimal risk for the subject.

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