Radiologically, intracranial RDD mimics a meningioma.

The histopathologic differentials include lymphoplasmacyte-rich form of meningioma, inflammatory pseudotumor, schwannoma, Langerhans cell histiocytosis [LCH], Erdheim-Chester disease [ECD], metastatic carcinoma, hematolymphoid neoplasm, granular cell tumor and melanoma.

Immunohistochemical work up is useful to arrive at a definite diagnosis.

Surgery is the primary modality of treatment. The highest rate of complete remission (near 90%) has been achieved by surgery. The benefit of radiotherapy alone seems rather limited (30%), while chemotherapy is in general ineffective.

A definitive diagnosis of extranodal RDD largely depends on the awareness of this entity. The histomorphology of a nodal RDD is fairly straightforward in most of the cases as the sinus histiocytosis and emperipolesis are readily appreciated.

However, in extranodal RDDs, emperipolesis may be subtle, and most importantly the sinuses are not present. Furthermore, brisk lymphoplasmacytic infiltrate and/or atypia in the histiocytes complicate the histopathologic features. In such scenarios a broad IHC panel is necessary.

The index case represents a meningeal RDD radiologically and clinically mimicking a meningioma. The morphology and IHC clinched the diagnosis, after exclusion of other meningeal histiocytic disorders. Additionally, this case highlights the importance of immunostains and awareness of this entity in an extranodal location in order to establish a correct and timely diagnosis.