CASE REPORT

Rosai-Dorfman disease: uncommon meningeal presentation and insights from PET/CT imaging

Bensaid Chaymae[1*], Hasnae Guerrouj[1], Manale Otmani[1], Ayat Mouaden[1], Imad Ghfir[1]

Pakistan Journal of Nuclear Medicine
Volume 15(1):46–49
DOI: 10.24911/PJNMed.175-1721044363

Pakistan Journal of Nuclear Medicine is the official journal of Pakistan Society of Nuclear Medicine

ABSTRACT

Rosai-Dorfman disease (RDD) is a rare, benign histiocytic disorder of unclear etiology, most commonly presenting with lymphadenopathy - particularly in the cervical region - alongside systemic symptoms such as fever and elevated inflammatory markers. While nodal involvement is typical, extranodal manifestations can occur, and neurological involvement - though uncommon - may present as cerebral pachymeningitis or spinal cord abnormalities. 18F-FDG PET/CT has emerged as a valuable tool for early disease assessment and monitoring. We report the case of a 32-year-old patient with RDD presenting predominantly with meningeal involvement. The patient underwent surgical intervention, followed by treatment with corticosteroids and valproic acid (Depakine). Disease prognosis is closely linked to the extent of involvement. In this case, 18F-FDG PET/CT played a critical role in both treatment planning and response monitoring, demonstrating clear advantages over conventional computed tomography (CT) and Magnetic Resonance Imaging in evaluating disease activity.

Keywords: Rosai-Dorfman disease, meningeal involvement, 18F-FDG PET/CT, extranodal histiocytosis.

Received: 15 July 2024 **Revised:** 24 August 2025 **Accepted:** 27 September 2025

Address for correspondence: Bensaid Chaymae
*Department of Nuclear Medicine, Ibn Sina Teaching Hospital, Mohamed V University, Rabat, Morocco.
Email: chamabensaid@gmail.com
Full list of author information is available at the end of the article.

Introduction

Rosai-Dorfman disease (RDD) is a rare, benign non-Langerhans histiocytosis of unknown etiology [1]. The most common clinical presentation involves significant bilateral cervical lymphadenopathy, which typically resolves spontaneously. Neurological involvement occurs in less than 5% of cases [2].

^{18}F-fluorodeoxyglucose (FDG) PET/CT plays a crucial role in capturing the comprehensive features of RDD, especially during initial assessment, treatment planning, and efficacy evaluation. It helps compensate for certain limitations of CT and MRI imaging, which may have difficulty assessing the complete extent of disease activity and distinguishing between active inflammation and fibrosis. Moreover, they do not offer functional information on metabolic activity, a key advantage provided by 18F-FDG PET-CT [3]. We report a rare case of RD where neuromeningeal manifestations predominate, and where ^{18}F-FDG PET-CT revealed the complete extent of disease, including atypical location, suggesting a mixed form of the disease.

Case Report

We report the case of a 32-year-old patient with no notable medical history, who presented a few days before his first consultation with a severe occipital headache, episodic mood swings, behavioral changes, notably anger outbursts, and social withdrawal, all evolving in the context of unquantified weight loss. The patient's condition worsened with the intensification of headaches, leading to increased use of cannabis for pain relief. One month later, the patient presented with sudden blindness. A brain scan revealed bifocal tumor masses in the right frontoparietal and left frontal regions, confirmed by an orbito-cerebral MRI, suggesting a primary or secondary meningeal invasion (Figure 1). The patient underwent surgery, and histopathological and immunohistochemical examination revealed meningeal involvement of Rosai-Dorfman disease (Anti CD68, anti CD163, anti PS100, anti cycline D1 were positive, while anti CD1, anti langérine, anti RP, anti GFAP, anti olig2, anti Melan A, anti HMB45, anti CK, AE1/AE3 were negative). Subsequently, the patient was referred to our department for ^{18}F-FDG PET-CT imaging for disease staging, which revealed cerebral metabolic defects corresponding to postoperative edematous-hemorrhagic changes and hypometabolism in the right parietal lesion area (Figure 2). Additionally, pathological hypermetabolic foci were detected in lymph nodes, musculoskeletal, and cutaneous regions, suggesting a mixed and

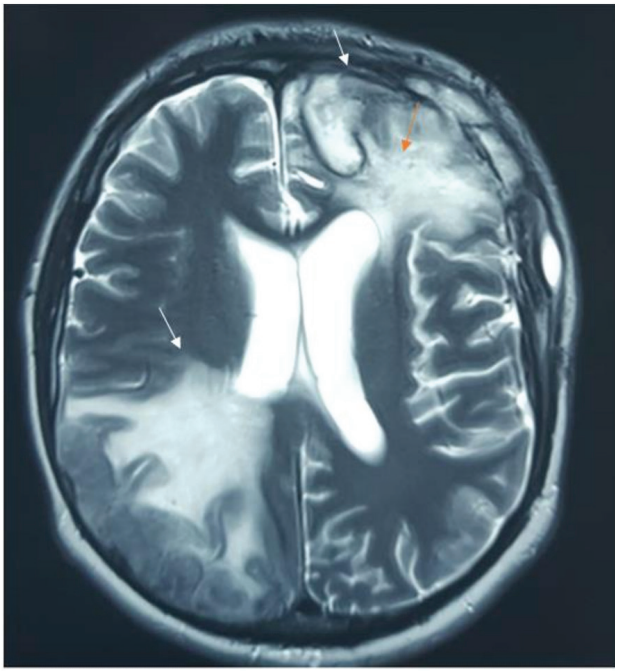

Figure 1. *Axial T2-weighted magnetic resonance imaging showing an extra-axial mass-like process in the left frontal and right parietal regions, appearing isointense on T2 sequences (white arrows). There is associated perilesional edema in the adjacent white matter, appearing hyperintense on T2 (orange arrow), which is causing a slight mass effect on the ventricular structures.*

atypical form of the disease (Figure 3). The patient was treated with corticosteroid therapy and valproic acid. During the course of treatment, the disease progression was favorable. Clinically, there was a notable improvement in neurological symptoms, and imaging studies revealed significant regression of previously noted disease. This was demonstrated by the follow-up PET-CT scan, which showed no pathological hypermetabolic foci in the lymph nodes, viscera, or musculoskeletal system, indicating a significant reduction in disease activity.

Discussion

Rosai-Dorfman disease is a rare, benign histiocytic proliferation characterized by the overgrowth of histiocytic cells within the lymph node sinuses or, in cases of visceral involvement, within the lymphatic vessels. Its etiopathogenesis remains poorly understood, although an infectious cause (bacterial, Epstein-Barr virus, and human herpesvirus 6) is suspected but not yet proven [1,4].

Classically, it manifests as bilateral cervical lymphadenopathy with fever, accelerated erythrocyte sedimentation rate, leukocytosis, and hypergammaglobulinemia [5]. In addition to lymph nodes, other areas can also be affected. Extra-nodal involvement occurs in about 50% of cases. The most commonly affected extra-nodal sites include the

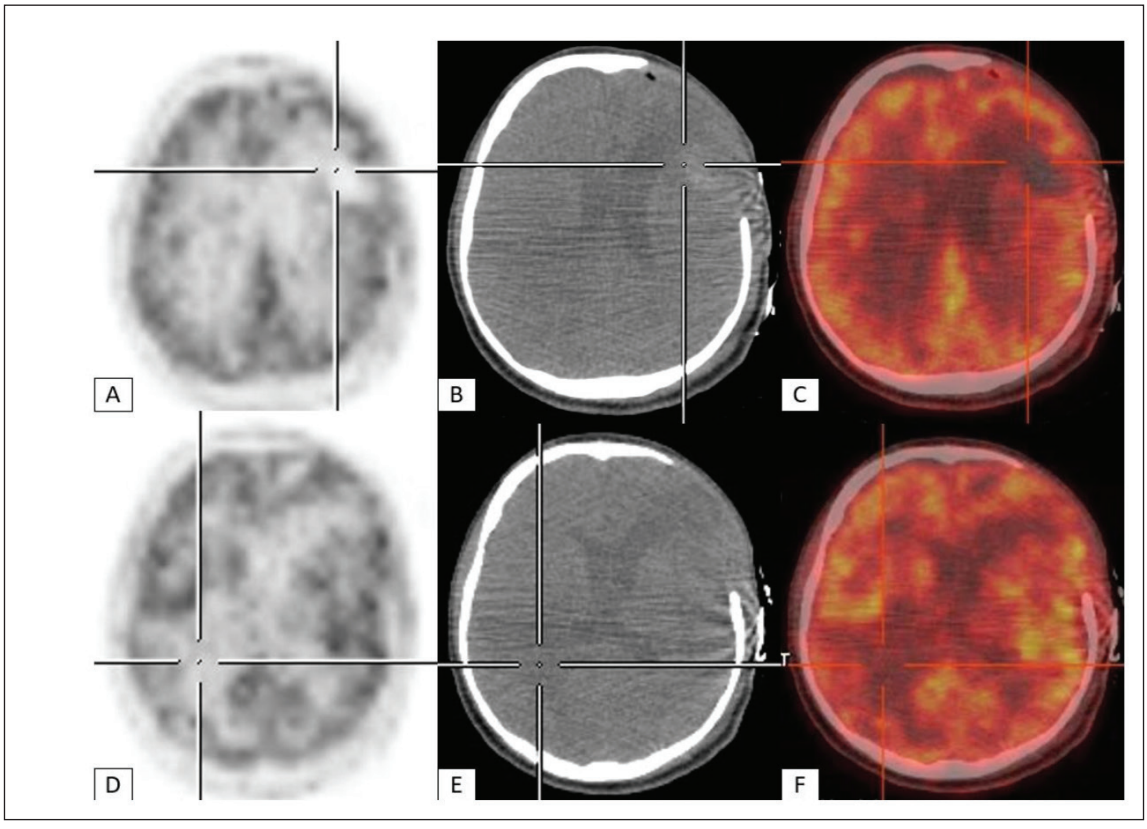

Figure 2. *Axial 18F-FDG PET, CT, and Fused PET/CT brain images revealing a metabolic defect in the postoperative intra-parenchymal edema-hemorrhagic alterations in the left fronto-parietal region and a non-hypermetabolic subgaleal collection in the left frontoparietal area (A, B, C), associated with slight hypometabolism in the right parietal expansive lesion process (D, E, F).*

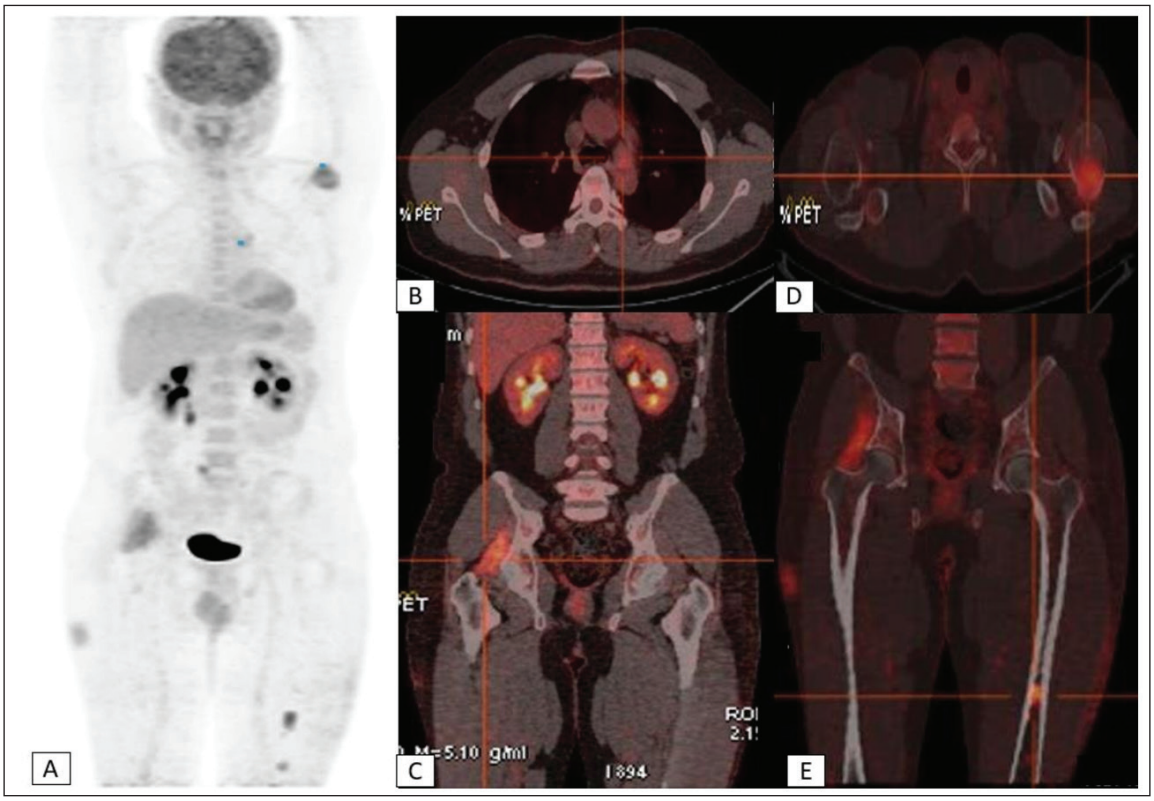

Figure 3. *(A): Maximum intensity projection (MIP) from the 18F-FDG PET-CT scan displays both the physiological and pathological distribution of the radiopharmaceutical throughout the entire body, providing a comprehensive overview of metabolic activity. (B): The axial fusion image highlights a slightly hypermetabolic mediastinal lymph node located in the aortopulmonary window, suggesting pathological involvement. (C): The coronal fusion image demonstrates hypermetabolism in the right gluteus minimus muscle, indicating abnormal metabolic activity in this region. (D) and (E): Axial and coronal fusion images reveal pathological hypermetabolic bone foci in the left humeral head and left femoral diaphysis, indicative of a mixed and atypical presentation of Rosai-Dorfman disease.*

skin [16%], (ear, nose, and throat) [16%], bones [11%], and orbits [11%]. Typically, these extra-nodal manifestations are found alongside lymph node involvement, even if isolated extra-nodal locations are possible, but rare [1]. Neurological involvement in RDD does not exceed 5%, mainly characterized by pachymeningitis and spinal cord localization. The meningiomatous form is even rarer [6], as is the case in our patient.

Confirmation of diagnosis relies on histopathological examination. Histological analysis reveals enlarged cells with abundant eosinophilic or occasionally xanthomatous cytoplasm, commonly known as "Destombes cells," accompanied by large, often nucleolated nuclei. A hallmark feature is the presence of numerous intact lymphocytes within the histiocytic cell cytoplasm, termed "emperipolesis," indicating active lymphocyte penetration rather than phagocytosis. Immunohistochemical analysis further supports the diagnosis, with RDD histiocytes expressing all pan-macrophage markers (CD68, CD14, CD64, and CD15) and the S100 protein, while CD1a is negative [1]. This aligns with the findings observed in our patient.

The specific contribution of imaging to the management of RDD remains uncertain. While several case reports have highlighted imaging findings in RDD, the variability in the disease's nature and progression complicates defining the exact role of imaging. Among conventional imaging methods, MRI stands out in certain instances, offering benefits in assessing intracranial, spinal, or perineural involvement [7]. Furthermore, ^{18}F-FDG PET/CT helped reveal the comprehensive features of RDD, particularly during initial assessment, adjustment of treatment strategies, or evaluating efficacy. Indeed, lesions of Rosai-Dorfman disease are known to exhibit FDG uptake, making PET a valuable tool for initial staging, especially in detecting non-contiguous sites of involvement [7,8]. According to Mahajan et al. [7] ^{18}F-FDG PET/CT would accurately characterize the disease foci, reveal foci not detectable by anatomical imaging, and prompt modifications in RDD treatment strategies, as demonstrated in their study where PET/CT led to a change in management in 41% of patients [7]. In our context, ^{18}F-FDG PET/CT facilitated the extension assessment for a mixed and atypical form of RDD by confirming the neuromeningeal involvement and revealing other lymph node, musculoskeletal, and skin locations.

^{18}F-FDG PET/CT also offers the advantage of early evaluation and disease extent in addition to treatment response evaluation, as was the case in our patient.

The treatment of RDD is still controversial. According to some authors, treatment is not necessary in the majority of cases as this condition typically exhibits a spontaneously favorable course. Treatment is reserved for disseminated forms with extra-nodal involvement causing clinical symptoms, such as in our case, or those with compressive lymphadenopathy. The most commonly used treatment is corticosteroid therapy. Surgery is only indicated in cases with compressive forms. Radiotherapy is reserved for progressive forms or in cases of spinal cord compression or respiratory impairment. The prognosis is often favorable, with mortality not exceeding 4% [1,6].

Conclusion

Rosai-Dorfman disease is characterized by its clinical polymorphism, including rare events such as meningeal involvement. PET/CT imaging, especially with 18F-FDG, has become essential for evaluating and monitoring RDD, particularly for detecting extranodal disease. It provides valuable insights for treatment planning and monitoring the course of the disease. Further research is needed to confirm the effectiveness of PET/CT imaging in optimizing RDD management and patient outcomes.

List of Abbreviations

^{18}F-FDG PET-CT	18F-FDG positron emission tomography–computed tomography
MRI	Magnetic resonance imaging
RDD	Rosai-Dorfman disease
SUVmax	Standardized uptake value

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

Funding

None.

Consent for publication

Permission was obtained from the patient/parents/guardians of the patient to publish the case and the accompanying images.

Author contributions

Conceptualization: Imad Ghfir, Chaymae Bensaid, Manale Otmani
Literature search: Chaymae Bensaid
Methodology: Imad Ghfir, Chaymae Bensaid
Manuscript preparation: Chaymae Bensaid
Manuscript editing and manuscript review: Imad Ghfir, Hasnae Guerrouj, Chaymae Bensaid, Manale Otmani, Ayat Mouaden

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

Author details

Bensaid Chaymae[1], Hasnae Guerrouj[1], Manale Otmani[1], Ayat Mouaden[1], Imad Ghfir[1]
1. Department of Nuclear Medicine, Ibn Sina Teaching Hospital, Mohamed V University, Rabat, Morocco

References

1. Ben Thayer M, Helal I, Khanchel F, Hmidi A, Ben Brahim E, Jouini R, et al. La maladie de Rosai Dorfman : une entité rare encore mal connue. La Rev Méd Interne. 2021;42:491. https://doi.org/10.1016/j.revmed.2021.03.113
2. Foré R, Cohen-Aubart F, Compagnat M, Cussinet L, Palat S, Fauchais AL, et al. Atteinte neurologique isolée de la maladie de Rosai-Dorfman-Destombes. La Rev Méd Interne. 2019;40:A124–5. https://doi.org/10.1016/j.revmed.2019.10.164
3. Lu X, Wang R, Zhu Z. The value of 18F-FDG PET/CT in the systemic evaluation of patients with Rosai-Dorfman disease: a retrospective study and literature review. Orphanet J Rare Dis. 2023;18(1):116.
4. Vanhaecke C, Vignon-Pennamen MD. Maladie de Rosai-Dorfman. Ann Dermatol Vénéréol. 2012;139(11):761–4. https://doi.org/10.1016/j.annder.2012.05.003
5. Bouzaher OMA, Taibi L, Zobiri S, Mameri S, Amma-Khodja A, Benkaidali I. Maladie de Rosai Dorfman cutanée exclusive : à propos d'un cas. Ann Dermatol Vénéréol. 2016;143(12):S393–4. https://doi.org/10.1016/j.annder.2016.09.636
6. Mankai R, Skouri W, Tounsi H, Ben Ammou B, Garbouj W, Chaabene I, et al. Atteinte pseudo-méningiomateuse au cours de la maladie de Rosai-Dorfman. La Rev Méd Interne. 2022;43:A420–1. https://doi.org/10.1016/j.revmed.2022.10.155
7. Mahajan S, Nakajima R, Yabe M, Dogan A, Ulaner GA, Yahalom J, et al. Rosai-dorfman disease—utility of 18F-FDG PET/CT for initial evaluation and follow-up. Clin Nucl Med. 2020;45(6):e260–6. https://doi.org/10.1097/RLU.0000000000003014
8. Xue Q, Miao W. Spontaneous recovery of Rosai-Dorfman disease on FDG PET/CT. Clin Nucl Med. 2017;42(8):608–9.