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Potential role of ^{18}F-FDG PET/CT in a case of progressive Rosai Dorfman disease

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ABSTRACT

Rosai Dorfman disease is a rare form of nonlangerhans cell histiocytosis, presenting with extensive lymphadenopathies. Treatment in most cases of nodal disease, involves close observation; however, extranodal involvement requires a more definitive treatment strategy. Herein, we report a case of extensive Rosai Dorfman disease in a 43-year-old woman presenting for evaluation of treatment response by ^{18}F-FDG PET/CT after frequent relapses and disease progression. In addition to extensive lymphadenothapies in cervical, supraclavicular, superior mediastinum, axillary, abdominopelvic and inguinofemoral regions, the patient had metabolically active bone and subcutaneous lesions which were not previously recognized. Following this ^{18}F-FDG PET/CT study, the patient management was changed to sirolimus and prednisolone. To choose the best treatment option for Rosai Dorfman patients, knowledge of the full extent of disease is important. Compared with conventional imaging, ^{18}F-FDG PET/CT has the advantage of being a whole-body imaging modality and can recognize disease involvement prior to any anatomical changes.

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Introduction

Rosai Dorfman disease (RDD) is a nonmalignant disease of the histiocytic cells, most commonly presenting as massive lymph nodes (1). Extranodal findings are not unusual (2). Despite a benign course in most cases and possibility of spontaneous remission, advanced cases have also been reported, warranting more definitive management (2). ^{18}F-FDG PET/CT is valuable in demonstrating the extent of involvement in these patients (3, 4). Recognition of the exact sites of involvement is crucial, to opt for the best treatment option (3). Among extranodal sites, involvement of the skeleton, harbors a more protracted course and frequently requires therapy (5). Herein, we present a patient with RDD referred for evaluation of treatment response with ^{18}F-FDG PET/CT after frequent relapses.

Case presentation

A 43-year-old woman with established diagnosis of Rosai Dorfman disease was referred to our center, for evaluation of treatment response. Three years prior, she had developed a mass on the left side of the neck and the CT-scan performed for further assessment revealed multiple lobulated lymph nodes on anterior cervical zones, bilaterally. The tissue sampling from a cervical lymph node revealed massive sinus histiocytosis. The sinuses were filled with histiocytes, some of which contained emperipolesis. The patient was treated with dexamethasone and antibiotics, which were effective in reducing the size of lymph nodes. However, follwing discontinuation of treatment, within one month, the lymph node masses resurfaced. Consequently, she was further treated with Rituximab every three weeks. This

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treatment was ineffective and was replaced with Vinblastin which was able to hinder disease progression; however, upon discontinuation, there was further enlargement in the size of lymph nodes. The last CT scan, dated August 2018, showed extensive cervical lymphadenopathy in almost all cervical compartments. Few lymph nodes were noted in bilateral axillary regions. Also, there were lymphadenopathies in the paraaortic, left common and external iliac chains (SAD=25 mm).

At the time of presentation, the patient had protrusion of the right eye and extensive swelling of the neck. She underwent PET/CT study which demonstrated extensive hypermetabolic lymphadenopathies in bilateral parapharyngeal, bilateral periparotid, bilateral supraclavicular and bilateral cervical zone I-V (SUV_{max} up to 26) (Figure 1), superior mediastinum and bilateral axillary regions (SUV_{max} up to 18.82). Additionally, there were highly active lymphadenopathies in the retro and intraperitoneal as well as bilateral inguinofemoral regions (SUV_{max} up to 16.24). Also, lytic bone lesions with high FDG avidity were noted in the right femur, left iliac and bilateral humeri (SUV_{max} up to 8.7) (Figure 2). Subcutaneous nodules were seen in upper posterior chest wall and medial aspect of left thigh (SUV_{max} up to 3.21) (Figure 3). Following these findings, the patient's therapeutic management was changed to sirolimus and prednisolone.

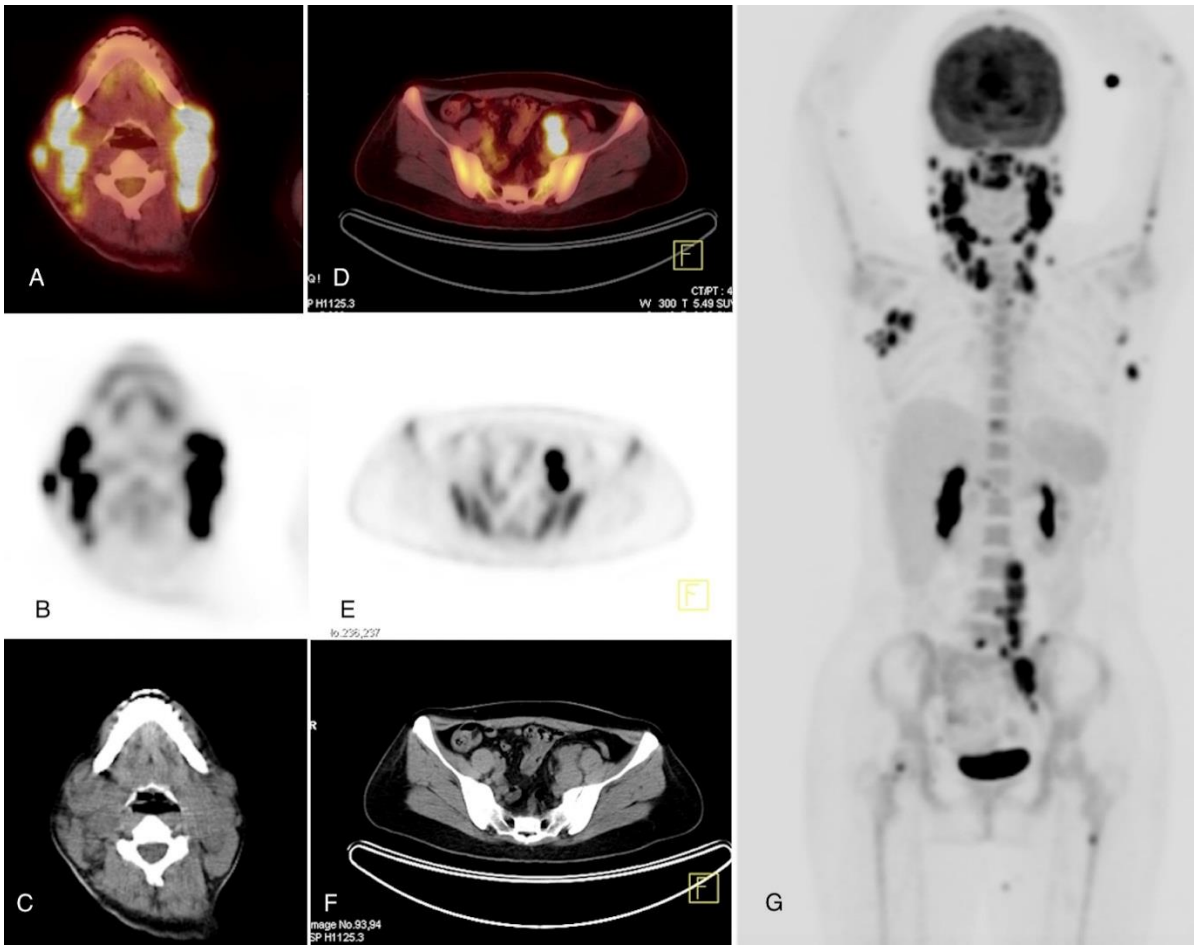

Figure 1. PET/CT fusion (A), PET (B) and CT (C) images demonstrated hypermetabolic lymphadenopathies in the bilateral periparotid and bilateral cervical zone II. PET/CT fusion (D), PET (E) and CT (F) images revealed highly active lymph nodes in the left external and internal iliac regions. MIP (G) image revealed extensive lymphadenopathies involving multiple anatomic sites

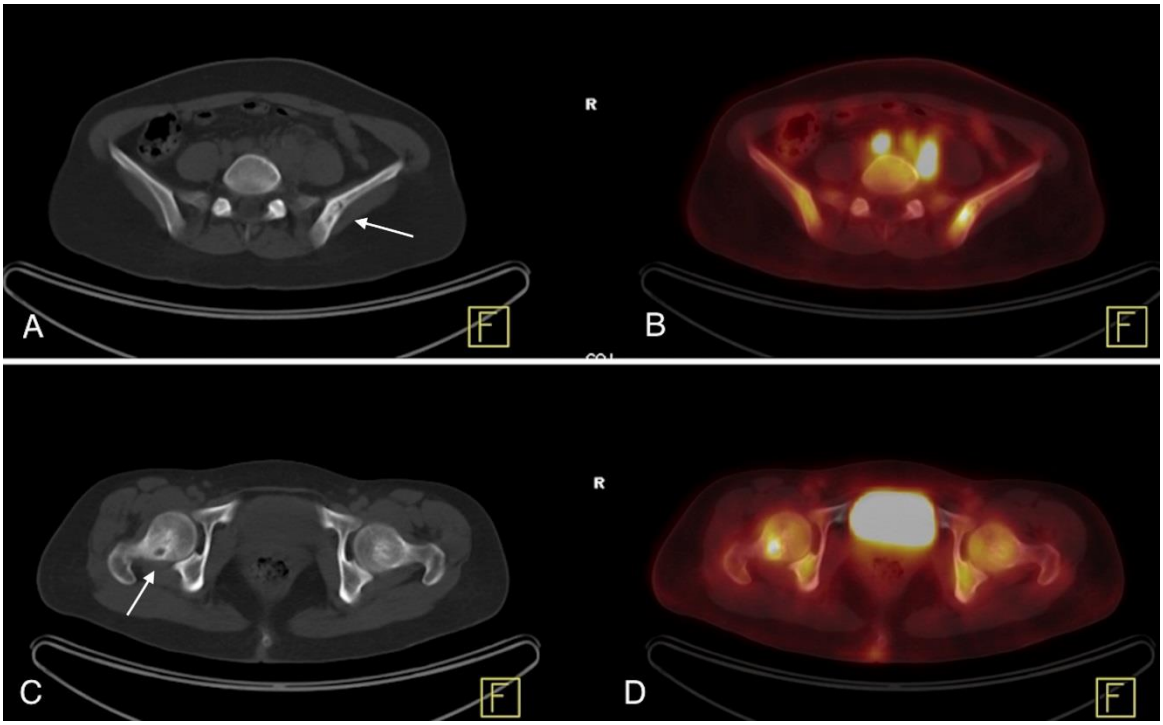

Figure 2. CT (A, C) and PET/CT fusion (B, D) images revealed FDG avid lytic lesions in the left iliac bone and right femur head, respectively

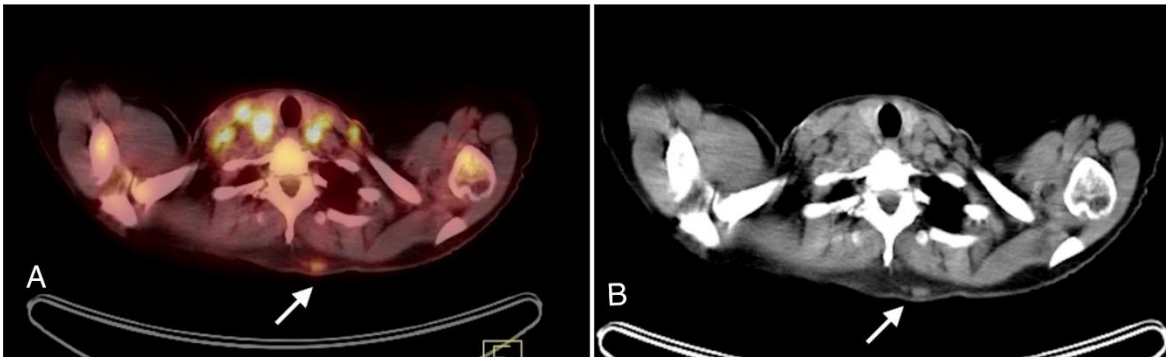

Figure 3. PET/CT fusion (A) and CT (B) images demonstrated a metabolically active subcutaneous lesion in upper posterior chest wall

Disucussion

Rosai Dorfman disease is a rare nonneoplastic entity, which presents most commonly with massive cervical lymphadenopathies (5). In this condition, the lymph node sinuses dilate and extensive histiocytes are seen within these sinuses accompanied by emperipolesis (intact lymphocytes in the cytoplasm of histiocytes). On microbiology evaluation, the cells stain positive for S100, CD68 and are negative for CD1a which makes this entity a non-langerhans cell histiocytosis (2, 6). Extranodal involvement is seen in about 40% of cases (5). Various sites of extranodal involvement, including skin, bone, respiratory tract, pituitary, hepatic and pancreatic regions have been reported (2, 7-9). There have been few case reports of isolated extranodal involvement. kong et al. reported isolated bone and pituitary involvement in a 10-year- old boy (9). Zhao et al. reported three cases of isolated extranodal involvement in nasopharynx, internal auditory canal, and facial sinus (10). The prognosis and treatment depend on the sites of involvement. Treatment options vary depending on the patient status of disease. In most cases without extranodal involvement, observation seems reasonable; however, with extranodal disease ie. Liver, pleura, osseous lesions, different therapeutic options have been employed (2). The treatment options include surgery, radiotherapy, chemotherapy and glucocorticoids (2, 11).

Bone involvement is rare and is seen in about 10% of cases. Bone lesions have been reported as a sole finding (12) or associated with lymphadenopathies (5). Bone lesions are lytic and frequently involve the skull or extremities. They usually have a protracted course, mandating therapy with glucocorticoids, or other

therapeutic options (5). Cutaneous lesions can occur as an isolated finding or with systemic involvement. Although limited skin involvement can be surgically resected, there is a risk of recurrence, and a conservative approach might be preferred (13).

Our patient, not only had extensive lymphadenopathies in the cervical and abdominopelvic regions that continued to relapse despite therapy for three years, she also suffered from bone and subcutaneous involvement which were not recognized prior to the PET/CT study. RDD lesions are quite FDG avid and PET/CT is extremely valuable in demonstrating the extent of disease, and choosing the best treatment strategies (3). Assessment of the true burden of the disease is not always possible on conventional imaging, either due to underrecognition of lesions or being outside the field of view of these imaging modalities (3). As a result, the best treatment option might not be applied if important extranodal sites of involvement such as bones are missed. On the other hand, not only ^{18}F-FDG PET/CT as a whole-body imaging technique, can be utilized for early staging, it can also be valuable in monitoring treatment response. A decrease in metabolic activity in follow-up ^{18}F-FDG PET/CT, indicates a therapeutic response, while this change might not be easily detected on conventional imaging (3). Mahajan et al. reviewed 109 ^{18}F-FDG PET/CT scans in 27 patients with Rosai Dorfman disease. In six patients additional sites of involvement was recognized in the bone and pleura which were missed on conventional imaging resulting in a change in management in 41% of cases (3). Rosai Dorfman lesions are quite FDG avid, making it possible to recognize lesions not easily detected on conventional imaging, in a study pleural lesions, ear nodules and testicular involvement (3) and in our case, bone and subcutaneous lesions were additionally detected on PET/CT study. Also, Shrestha et al. retrospectively evaluated FDG PET/CT findings of 14 patients with proven RDD and found ^{18}F-FDG PET/CT a valuable modality in extranodal sites (14).

Some limitations may apply, in Rosai Dorfman disease, dural based intracranial lesions are common in CNS involvement; however, spinal and cerebral lesions are rare but have been reported (15,16). PET/CT is traditionally known to be limited for brain lesions, due to the presence of high background brain uptake (17), although Mahajan et al. were able to demonstrate intracranial lesions on PET/CT study due to their high metabolic activity (3) and Deshayes et al. reported a case of RDD of hypothalamic lesion detected on FDG PET/CT (18), this limitation of PET/CT must be considered especially in patients with clinical sign of intracranial involvment. Also, for spinal intramedullary lesions, although with close evaluation, PET/CT might be able to detect most lesions, MRI is considered the superior modality (19).

In conclusion, although the role of ^{18}F-FDG PET/CT in Rosai Dorfman disease has not been fully validated yet, our case in concordance with previous reports (3, 9, 14, 20) emphasizes on its potential value to demonstrate the full extent of disease and choose the best management strategy.

References

1. McClain KL, Natkunam Y, Swerdlow SH. Atypical cellular disorders. Hematology. 2004; 2004(1):283-96.
2. Averitt AW, Heym K, Akers L, Castro-Silva F, Ray A. Sinus histiocytosis with massive lymphadenopathy (Rosai Dorfman disease): diagnostic and treatment modalities for this rare entity revisited. J Pediatr Hematol Oncol. 2018; 40(4):e198-e202.
3. Mahajan S, Nakajima R, Yabe M, Dogan A, Ulaner GA, Yahalom J, et al. Rosai-Dorfman Disease—Utility of ^{18}F-FDG PET/CT for Initial Evaluation and Follow-up. Clin Nucl Med. 2020; 45(6):e260-e6.
4. Karunanithi S, Singh H, Sharma P, Naswa N, Kumar R. ^{18}F-FDG PET/CT imaging features of Rosai Dorfman disease: a rare cause of massive generalized lymphadenopathy. Clin Nucl Med. 2014; 39(3):268-9.
5. Mosheimer BA, Oppl B, Zandieh S, Fillitz M, Keil F, Klaushofer K, et al. Bone involvement in Rosai-Dorfman disease (RDD): a case report and systematic literature review. Curr Rheumatol Rep. 2017; 19(5):29.
6. Mantilla JG, Goldberg-Stein S, Wang Y. Extranodal Rosai-Dorfman disease: clinicopathologic series of 10 patients with radiologic correlation and review of the literature. Am J Clin Pathol. 2016; 145(2): 211-21.
7. Foucar E, Rosai J, Dorfman R, editors. Sinus histiocytosis with massive lymphadeno-pathy (Rosai-Dorfman disease): review of the entity. Semin Diagn Pathol. 1990; 7(1):19-73.
8. Shaikh F, Awan O, Mohiuddin S, Farooqui S, Khan SA, McCartney W. ^{18}F-FDG PET/CT imaging of extranodal Rosai-Dorfman disease with hepatopancreatic involvement-a pictorial and literature review. Cureus. 2015; 7(12).
9. Kong Z, Wang Y, Ma W, Cheng X. FDG PET/CT Image for a Rosai-Dorfman Disease With

Pituitary and Bone Involvement in a Pediatric Patient. Clin Nucl Med. 2019; 44(11):873-5.
10. Zhao X, Yu D, Jin C. Three Cases of Extranodal Rosai-Dorfman Disease and Literature Review. Open Life Sciences. 2018; 13(1):263-8.
11. Ocheni S, Ibegbulam O, Okafor O, Raveenthiran V, Nzegwu M. Usefulness of oral corticosteroid in Rosai–Dorfman disease. Eur J Cancer Care. 2007; 16(3):286-8.
12. Sundaram C, Chandrashekar P, Prasad V, Umadevi M. Multifocal osseous involvement as the sole manifestation of Rosai-Dorfman disease. Skeletal Radiol. 2005; 34(10):658-64.
13. Hadchiti J, Kamar FG, Abi Ghosn J, Haidar M, Younes A, Obeid A, et al. ^{18}F-fluoro-2-deoxyglucose positron emission-computed tomography in a rare cutaneous form of Rosai-Dorfman disease: A case report. Mol Clin Oncol. 2018; 8(2):236-41.
14. Shrestha R, Jamison DK, Anene D, Roarke M, Yang M. The Role of ^{18}F-FDG PETCT in the Evaluation and Surveillance of Rosai-Dorfman Disease. J Nucl Med. 2020; 61(supplement 1):1158.
15. Sandoval-Sus JD, Sandoval-Leon AC, Chapman JR, Velazquez-Vega J, Borja MJ, Rosenberg S, et al. Rosai-Dorfman disease of the central nervous system: report of 6 cases and review of the literature. Medicine. 2014; 93(3): 165-175
16. Adeleye AO, Amir G, Fraifeld S, Shoshan Y, Umansky F, Spektor S. Diagnosis and management of Rosai–Dorfman disease involving the central nervous system. Neurol Res. 2010; 32(6):572-8.
17. Corrigan AJG, Schleyer PJ, Cook GJ, editors. Pitfalls and artifacts in the use of PET/CT in oncology imaging. Semin Nucl Med; 2015: 481-499.
18. Deshayes E, Le Berre J-P, Jouanneau E, Vasiljevic A, Raverot G, Seve P. ^{18}F-FDG PET/CT findings in a patient with isolated intracranial Rosai-Dorfman disease. Clin Nucl Med. 2013; 38(1):e50-e2.
19. Mostardi P, Diehn FE, Rykken J, Eckel L, Schwartz K, Kaufmann TJ, et al. Intramedullary spinal cord metastases: visibility on PET and correlation with MRI features. AJNR Am J Neuroradiol. 2014; 35(1):196-201.
20. Dhull VS, Passah A, Rana N, Kaur K, Tripathi M, Kumar R. ^{18}F-FDG PET/CT of widespread Rosai-Dorfman disease. Clin Nucl Med. 2016; 41(1):57-9.