Isolated Extranodal Rosai-Dorfman Disease on 18F-FDG PET-CT Scan

Abstract

Rosai–Dorfman disease (RDD) is an uncommon proliferative histiocytic disorder. Patients usually present with painless massive cervical lymphadenopathy with fever and leukocytosis. Isolated extranodal disease is rare and more severe fibrosis, fewer histiocytosis in lesions make diagnosis more difficult as compared to nodal disease. Here, we report a case of isolated extranodal RDD on fluorodeoxyglucose (FDG) positron emission tomography-computed tomography (PET-CT) scan. FDG-avidity of RDD lesions is attributable to the intense glucose dependence of the proliferating histiocytes. PET-CT scan not only demonstrates the complete staging of the disease but also provide functional information about the disease activity to guide biopsy.

Keywords: *Extranodal, fluorodeoxyglucose positron emission tomography-computed tomography scan, Rosai–Dorfman disease*

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A 52-year-old female presented with nasal mass and difficulty in breathing. In view of suspicion of mass being neoplastic, the patient was referred for fluorodeoxyglucose (FDG) positron emission tomography-computed tomography (PET-CT) scan. Maximum intensity projection image [Figure 1a] showed FDG avid lesion in the nasal region only. Axial CT and fused axial PET-CT images [Figure 1b-e] revealed FDG avid lesion involving bilateral nasal cavity, causing erosion of nasal septum, laterally extending and involving the bilateral maxillary sinus. The corresponding coronal and sagittal CT and fused PET-CT images [Figure 1f-i] showed FDG avid lesion causing erosion of hard palate and adjacent alveolar margins. The whole-body survey showed the absence of any FDG avid visible lymph node or FDG avid visible disease elsewhere in the regions of the body surveyed. Biopsy of the FDG avid nasal mass was compatible with Rosai–Dorfman disease (RDD).

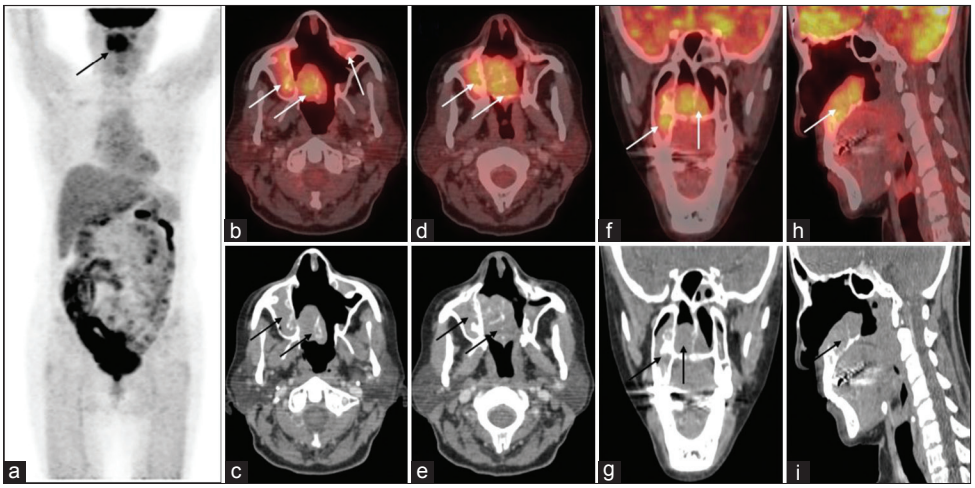

Figure 1: Maximum intensity projection image (a) showing fluorodeoxyglucose avid lesion in nasal region. Axial computed tomography (c and e) and fused axial positron emission tomography-computed tomography images (b and d) showing fluorodeoxyglucose avid lesion involving bilateral nasal cavity and corresponding coronal and sagittal computed tomography (g and i) and fused positron emission tomography-computed tomography coronal and sagittal images (f and h) showing fluorodeoxyglucose avid lesion causing erosion of hard palate and adjacent alveolar margins

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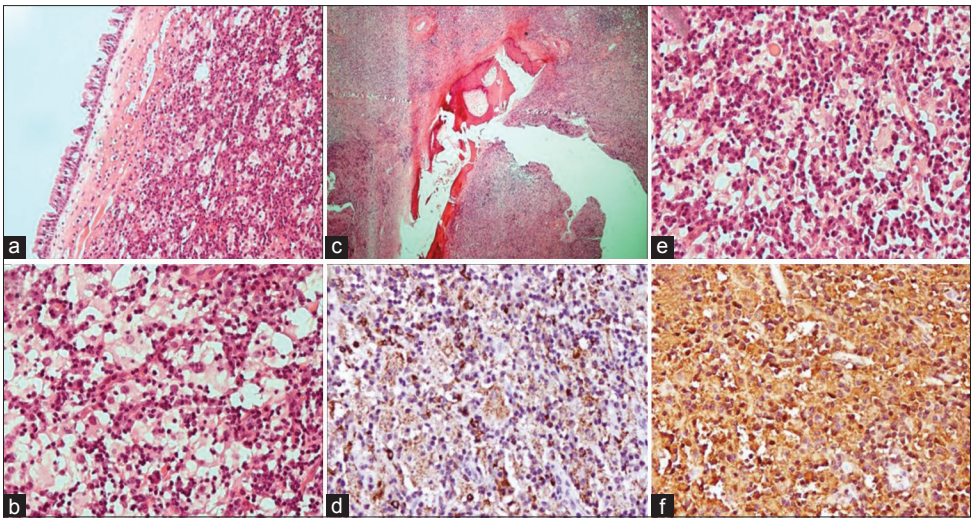

Figure 2: Histopathology and immunohistochemistry images of the patient. Section (a) shows tissue fragments capped with respiratory mucosa (H and E, ×4). The subepithelium section (b) shows a cellular lesion which is infiltrating the bone (H and E, ×4). The lesion (c) is composed of sheets of histiocytes, admixed with many lymphocytes and plasma cells. Some of the histiocytes are foamy (H and E, ×20). Some of the histiocytes (d) show emperipolesis (arrow) (H and E, ×40). Immunohistochemistry images show histiocytic cells positive for CD68 (e) and diffusely positive for S100 (x20) (f)

RDD, which was first described by Rosai and Dorfman in 1969, is a rare, benign lymphoproliferative disease.[1] Concurrent nodal and extranodal disease are seen in approximately 40% of cases.[2] The sole extranodal disease is seen in approximately 20%–25%.[3] Patients with involvement of extranodal sites tend to have a fulminant course. The most common extranodal sites are skin, nasal cavity, eyes, and bone.[4] Definitive diagnosis can be performed by histopathological examination [Figure 2] which reveals intense histiocytic infiltration, emperipolesis, and positivity for S100 and CD68. Differential diagnosis are lymphoma, tuberculosis, sarcoidosis, reactive hyperplasia, and nasopharyngeal carcinoma. Few case reports of extranodal RDD, from nasal septal mucosa,[5] osseous involvement,[6] and extranodal sites[7] have been reported previously. F18-FDG PET-CT scan is a valuable imaging technique for evaluating the extent of disease, demonstrating the complete staging of the disease, and guiding the biopsy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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