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Rosai-Dorfman disease involving the entire esophagus

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The authors declare that they have no conflict of interest in this study.

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Dear editor:

We present a case of Rosai–Dorfman disease (RDD) occurred in a 6-year-old male child, characterized by extensive involvement of the esophagus. Eight months ago, the child presented with persistent fever and a diffuse dark red rash. MRI revealed a mass occupying the left nasal cavity and septal sinus and biopsy pathology confirmed the diagnosis of RDD. Following dexamethasone and prednisone treatment, the child experienced dysphagia. ^{18}F-FDG PET/CT revealed multiple lesions with increased metabolism in the left nasal sinus, lymph nodes, widespread skin lesions, and the entire esophagus (Fig. 1A–1E). Subsequent biopsies of lymph nodes, abdominal skin, and esophageal lesions was consistent with RDD involvement (Fig. 1F–1G). The child is presently undergoing six cycles of VCR+Ara-c+Dex chemotherapy and the treatment is going well.

RDD is an uncommon myeloproliferative disorder characterized by the abnormal proliferation of histiocytes. RDD primarily affects pediatric and young adult populations, typically manifesting as bilateral cervical lymphadenopathy accompanied by symptoms such as fever, weight loss, and night sweats[1]. In addition to its nodal presentation, RDD can also manifest as extranodal and cutaneous forms, with the most frequently involved sites including the skin, nasal cavity, bones, orbital tissue, and central nervous system[2].

In this case, we describe disseminated RDD involving the entire esophagus, more rarely. Clinical course is variable, ranging from self-limited process to disseminated refractory disease. Histologically, a biopsy of the lesion reveals characteristic features of abnormal S100+, CD68+, and CD1a− histiocytes. The etiology of RDD is unclear. This case report presents a rare occurrence of disseminated RDD involving the entire esophagus. The clinical progression of RDD can vary from a self-limited course to a disseminated refractory disease. Histologically, a biopsy of the lesion reveals distinct features characterized by the presence of abnormal S100+, CD68+, and CD1a− histiocytes. The etiology of RDD remains uncertain. Catón et al [3] reported a case of mesenteric lymph node RDD, raising the possibility that it was secondary to an infectious process.

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RDD lesions demonstrate increased ^{18}F-flurodeoxyglucose (FDG) uptake, making PET a valuable tool for the initial staging of the disease, especially in the identification of non-contiguous sites of involvement. The management of RDD relies mainly on a comprehensive evaluation of the affected organs[4]. Due to the lack of a standardized treatment regimen, the decision-making process should consider factors such as the location, extent of the disease, and involvement of vital organs[5]. This child presents with multifocal, unresectable extranodal disease necessitating systemic therapy. The patient has received treatment consisting of corticosteroids and chemotherapy, resulting in a favorable clinical response.

In summary, RDD is a rare condition, and the utilization of ^{18}F-FDG PET/CT has been proven valuable in accurately delineating the extent of the disease in RDD patients. It is imperative to consider the potential involvement of the esophagus.

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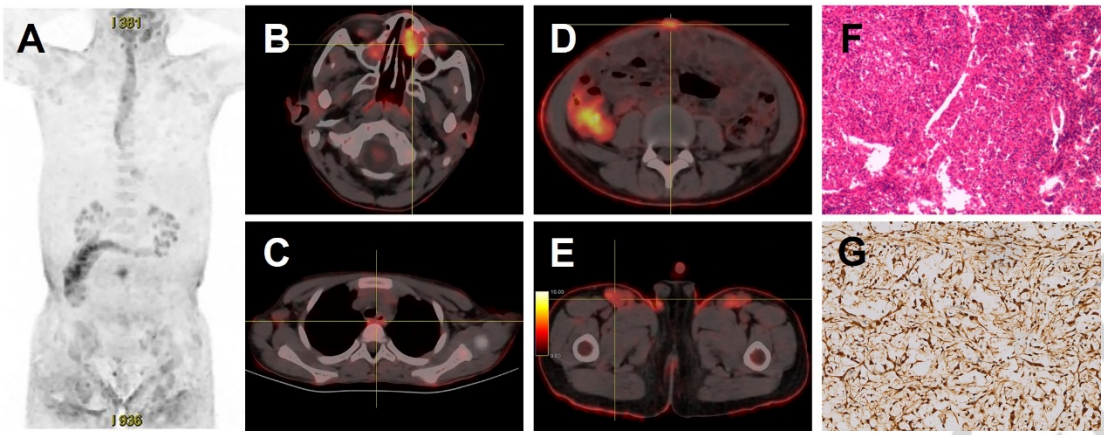

Fig.1. ^{18}F-FDG PET/CT showed multiple FDG-avid lesions in the whole segment of the esophagus, multiple lymph nodes and skin lesion were found by maximum intensity projection image (A). Axial fusion images showed increased metabolism in the left nasal cavity (B, SUVmax = 11.8), esophagus (C, SUVmax = 5.2), anterior abdominal median skin (D, SUVmax = 6.9) and bilateral inguinal lymph nodes (E, SUVmax = 4.7). The pathological examination showed histiocytosis, lymphocytic and plasma cell infiltration (F), with positive expression of S-100 (G)**.**

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