Role of ^{18}F-Fluorodeoxyglucose Positron Emission Tomography/Computed Tomography Scan in Castleman's Disease

Abstract

Castleman's disease (CD) is a rare benign lymphoproliferative disorder. We are presenting three cases of CD of which one is unicentric CD, and the other two are an idiopathic multicentric CD. One of the two multicentric cases is associated with POEMS syndrome. The whole body ^{18}F-Fluorodeoxyglucose positron-emission tomography–computed tomography scan plays a significant role in identifying the centricity, distribution of disease, response to therapy, and in early detection of remission.

Keywords: *Castleman's disease, fluorodeoxyglucose, positron-emission tomography–computed tomography*

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Introduction

Castleman's disease (CD) was first described in 1954 by Benjamin Castleman, a pathologist at Massachusetts General Hospital.[1] It was clinically classified into unicentric CD (UCD) and multicentric CD (MCD) and pathologically into hyaline vascular variant (HVV), plasma-cell variant (PCV), and mixed variants. HVV was more commonly unicentric and less aggressive whereas PCV was more commonly multicentric and more aggressive. Whole body ^{18}F-Fluorodeoxyglucose positron-emission tomography–computed tomography (WB FDG PET/CT) scan plays a significant role in identifying the centricity, distribution of disease, response to therapy, and in early detection of remission.

Case Reports

Case 1

A 32-year-old male patient presented with progressive neck swelling for 6 months. There was no history of fever, pain, night sweats, or swellings elsewhere in the body. Ultrasound sonography of neck showed the right level II, III, and IV cervical lymph nodes (largest measuring 4 cm × 1.9 cm) and few small nodes (6 mm × 5 mm) in left side of the neck. Core needle biopsy of the right cervical LN was taken. The histopathological examination (HPE) and immunohistochemistry (IHC) were suggestive of CD (HVV type). WB FDG PET/CT scan [Figure 1] showed mild FDG-avid right level II-IV cervical lymph nodes (SUVmax-4.3) and few nonFDG-avid subcentimetric left cervical and mediastinal nodes. There were no other abnormal hypermetabolic lesions elsewhere in rest of the body suggesting UCD. The patient was treated with four cycles of rituximab and hydrocortisone. A follow-up of WB FDG PET/CT scan was performed to assess the response. The scan showed a mild decrease in uptake of right cervical lymph nodes (SUVmax-3.7) with no significant change in the size [Figure 1]. Subsequently, the patient was given local radiotherapy to the right cervical lymph nodes. There was a clinical regression of cervical lymph nodes and was on follow-up until now with no progression.

Case 2

A 56-year-old male patient presented with progressive multifocal peripheral neuropathy for 1 year. The patient also had bilateral cervical lymphadenopathy. HPE and IHC of left supraclavicular lymph node were suggestive of CD (PCV type). Ultrasonography of abdomen showed hepatosplenomegaly. WB FDG PET/CT scan [Figure 2] showed mild FDG-avid enlarged lymph nodes in cervical, mediastinal, abdominal, and pelvic regions (SUVmax-4.3). In addition, mild FDG-avid multiple sclerotic bone

How to cite this article: Reddy Akepati NK, Abubakar ZA, Bikkina P. Role of ^{18}F-Fluorodeoxyglucose positron emission tomography/computed tomography scan in castleman's disease. Indian J Nucl Med 2018;33:224-6.

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Access this article online

Website: www.ijnm.in

DOI: 10.4103/ijnm.IJNM_26_18

Quick Response Code:

lesions were also seen. Serum electrophoresis showed an M-spike (IgA Lambda). Bone marrow biopsy was reported as normal. Based on these findings, a diagnosis of idiopathic MCD (iMCD) with POEMS syndrome was made. The patient was treated with steroids and melphalan for 6 months. There was no further clinical progression of the disease until now.

Case 3

A 41-year-old female patient presented with chest pain for 4 months and was diagnosed outside with a mediastinal mass. She was referred to our hospital for further management. WB FDG PET/CT scan [Figure 3] showed FDG-avid multiple mediastinal (largest measuring 52 mm × 33 mm, SUVmax-6.3) and pelvic lymph nodes (SUVmax-6.5) along with multiple FDG-avid sclerotic bone lesions (SUVmax-4.7). HPE and IHC are suggestive of CD (PCV type). The patient was treated with steroids for 3 months. A follow-up of WB FDG PET/CT scan for response assessment showed no significant difference in number, size, and metabolic activity of the existing lesions. The patient became clinically asymptomatic after steroid treatment and was on follow-up since then.

Discussion

UCD affects both sexes equally and most commonly present between third and fifth decade.[2] Most of the cases of UCD are detected incidentally. The incidence of MCD is equal in both sexes and is predominantly seen in the age group of fifth to sixth decade. MCD are of two types such as HHV8 associated and idiopathic (iMCD). Both MCD cases presented here are of an idiopathic type. Prevalence of iMCD is five per million in the Asia-Pacific region.[3] Both iMCD cases presented here are of PCV type and UCD is of HVV type. Up to 37% of patients with MCD have associated POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes) and 9%–24% of POEMS syndrome have associated MCD.[4,5] Five-year mortality rate in MCD is about 35% and there is a 3-fold increased risk of developing malignancy.[6] Diagnosis of CD is difficult and often misdiagnosed due to overlap in the clinical, histological, and immunological findings. In addition, other malignant, autoimmune, and infective conditions need to be excluded before making a diagnosis of CD.

WB 18F-FDG PET/CT has an important role in diagnosis and management of CD. Since it is a WB study, it is the investigation of choice to establish the centricity of disease and to know the distribution of disease. In case three, the initial diagnosis of UCD was changed to MCD by WB FDG PET/CT scan. It is also very useful in assessing response and to detect relapse of the CD. It may be helpful in excluding conditions that mimic iMCD.[7] CD tends to have mild FDG uptake compared to many of the lymphomas which have intense uptake. All the three cases, presented here have lesions with mild metabolic activity (SUVmax ranging from 3.7 to 6.5). Few small studies showed that SUV maximum value may serve as a prognostic factor in CD but it needs to be further evaluated.[8]

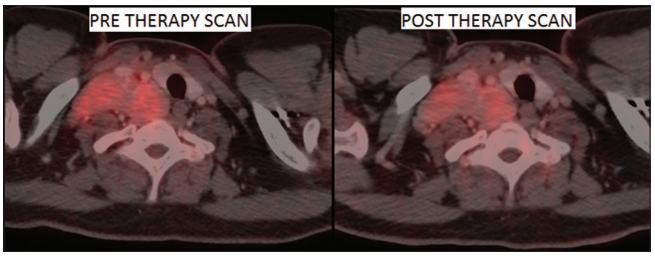

Figure 1: Case 1 – pretherapy and posttherapy positron-emission tomography/computed tomography scan image with mild fluorodeoxyglucose-avid right cervical lymph nodes. After treatment with rituximab and steroids, only a mild decrease in metabolic activity with no significant change in size is noticed

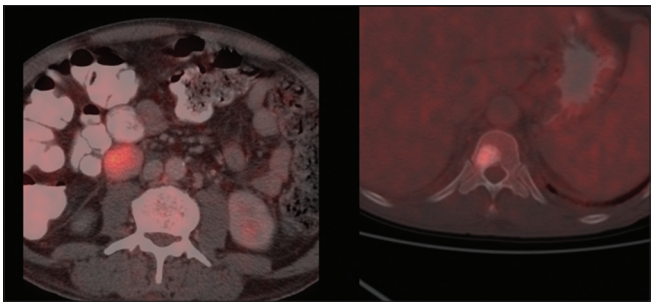

Figure 2: Case 2 – Multicentric Castleman's disease with fluorodeoxyglucose-avid paracaval lymph node and sclerotic bone lesion in vertebrae

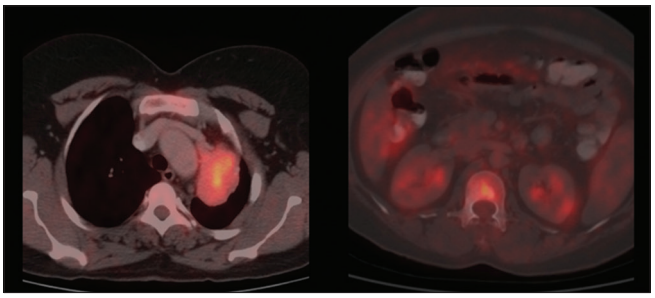

Figure 3: Case 3 – Multicentric Castleman's disease with fluorodeoxyglucose-avid mediastinal lymph node and sclerotic bone lesion in vertebrae

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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