Case Report

Beware of the Coated Aorta in Fluorodeoxyglucose Positron Emission Tomography–Computed Tomography: A Specific Clue to the Diagnosis of Erdheim–Chester Disease in a Case of Brain and Orbital Lesions with Unknown Primary

Abstract

Erdheim–Chester disease (ECD) is a systemic histiocytosis that can involve several organs, with severity ranging from occult to life-threatening. The disease was first described by William Chester in 1930 after working with the Austrian pathologist Jakob Erdheim. Even today, a correct diagnosis of ECD often takes years, given the rarity and variable manifestations of ECD. We present a case of a 63-year-old female presenting with multiple brain lesions, sent for fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography to find primary, and it showed hypermetabolic right occipital brain lesion, right orbital lesion, and soft tissue around the arch of the aorta (coated aorta), and final histopathology of the brain lesion confirmed histiocytosis ECD.

Keywords: *Brain lesions, coated aorta, Erdheim–Chester disease, fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography*

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Introduction

Erdheim–Chester disease (ECD) is a systemic non-Langerhans cell histiocytosis characterized by multi-organ accumulation of foamy histiocytes. The disease was first described by William Chester in 1930 after working with the Austrian pathologist Jakob Erdheim.[1] Owing to its rarity and varied presentation, ECD presents diagnostic challenges to the clinician and imageologist. Fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography (F-18 FDG PET/CT) plays a vital role in the diagnosis, detection of disease extent and severity, assessment for an appropriate biopsy, and treatment response.[2] Exophthalmos (retro-orbital space infiltration), "coated aorta" (circumferential soft-tissue sheathing of the thoracic aorta), and "hairy kidney" (soft-tissue rind of perirenal infiltration) are typical manifestations in patients with ECD.[3] We describe here the image findings of FDG PET/CT in a case of brain and orbital lesions, which showed hypermetabolic coated aorta which is a specific clue to the diagnosis of ECD.

Case Report

A 63-year-old female presented with altered speech in 2021 post-COVID and magnetic resonance imaging (MRI) of the brain suggestive of diffuse meningitis and suspicious of vasculitis and started on steroids. She recently in 2023 complained of right eye pain, blurring of vision, vomiting, giddiness, and repeated falls. MRI of the brain showed a right occipital lesion with edema. She was referred for a whole-body FDG PET/CT to find primary. The whole-body images showed hypermetabolic dural-based extra-axial enhancing lesion in the right occipital lobe Figure 1a and b], a right retro-orbital lesion encasing the optic nerve [Figure 1c and d], and soft-tissue thickening around the arch of the aorta [Figure 2]. She underwent a brain lesion biopsy, which showed a histiocyte-rich lesion with fibrosis and inflammation and no malignancy; with this, a final diagnosis of ECD was made and sent for BRAF mutation analysis.

Discussion

ECD is a systemic non-Langerhans cell

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histiocytosis characterized by multi-organ accumulation of foamy histiocytes. The disease was first described by William Chester in 1930 after working with the Austrian pathologist Jakob Erdheim.[1] The diagnosis is often delayed due to the rarity of the disease, multi-organ involvement, and varied presentation.[2] Up to 96% of patients show skeletal involvement. 99mTc-methylene diphosphonate bone scan and FDG-PET will show intense, bilaterally symmetric uptake at the end of the long bones centered around the knees with sparing of the epiphysis – "hot-knee pattern," which is a characteristic of ECD. FDG-PET and bone scan uptake can precede radiographic abnormalities. Our patient we did not image below the midthigh to see hot knee pattern.[3,4]

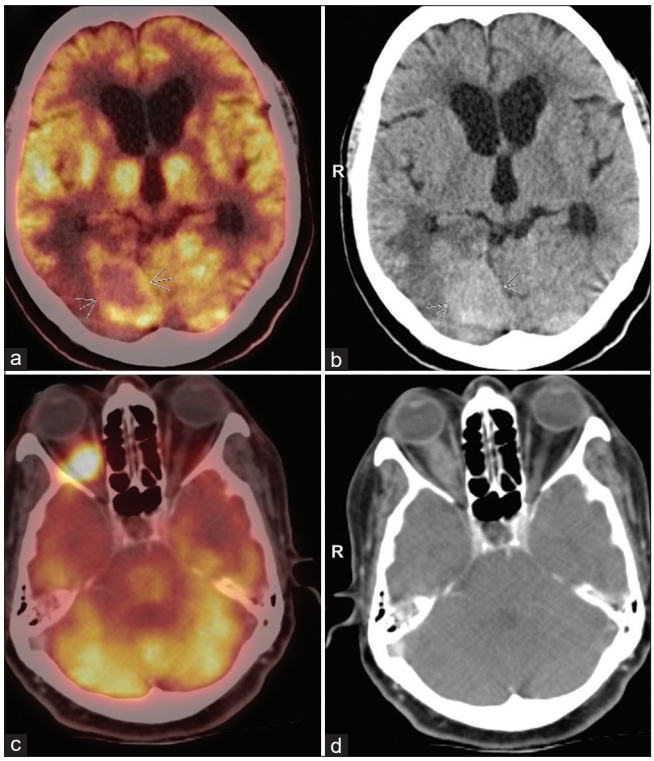

Figure 1: Fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography (PET/CT) axial fused PET/CT (a) axial CT (b) images showing hypermetabolic peripheral enhancing right occipital lesion with edema (arrows) and axial fused PET/CT (c) and axial CT (d) showing hypermetabolic right retro-orbital perioptic nerve lesion

FDG PET/CT may be the most useful imaging modality for differentiating ECD from other histiocytoses. Given multi-organ involvement, ECD often elicits a broad differential diagnosis, including retroperitoneal fibrosis, IgG4-related disease, sarcoidosis, and other types of histiocytoses. Although lymph node involvement in ECD has been reported, it is rare. A BRAF1 status essentially excludes other forms of non-Langerhans cell histiocytoses.[5]

In patients with ECD, neurologic morbidity is common and contributes significantly to disability. Since neurologic symptoms can be the presenting feature of ECD and given the mean delay in ECD diagnosis is 4.2 years, it is critical that neurologists consider ECD and other histiocytosis in patients with inflammatory, infectious, or neoplastic-appearing lesions. Our patient presented with neurological first manifestation.[6] Brain parenchymal lesions will mimic glioma, meningioma, or metastases.[7]

Orbital involvement, which can occur in 25%–37% of patients, presents with retrobulbar masses that can cause visual impairment, motility disturbance, proptosis, and optic nerve edema.[8] The most characteristic vascular

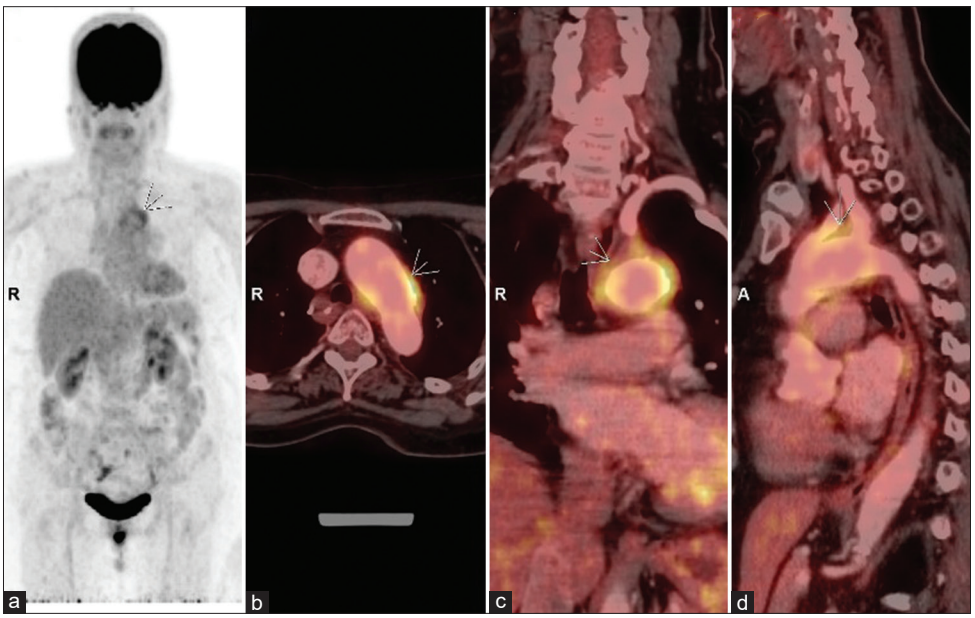

Figure 2: Fluorodeoxyglucose positron emission tomography/computed tomography (PET/CT) maximum intesnity projection (MIP) image (a), axial (b), coronal (c), sagittal (d), fused PET/CT images showing soft tissue surrounding the arch of the aorta and branches (arrows), suggesting a coated aorta sign, specific for Erdheim–Chester disease

presentation was FDG-avid soft tissue "coating" the aorta, involving the adventitial layer. CT typically shows periaortic tissue infiltration, extending from the ascending aorta to the iliac junction and creating the appearance of a "coated aorta."[5,9,10]

Our patient presented with neurological manifestations like stroke, then after 2 years, she presented with right eye vision problem, MRI showed brain lesion and FDG PET/CT showed brain and orbital lesions, which mimicked malignancy, but coated aorta which wont be seen in malignancy gave clue to diagnosis of ECD. A brain biopsy was performed as there was no accessible site that confirmed histiocytosis. Hence, in a known case of brain and orbital lesions in FDG PET/CT, the coated aorta is a specific clue to the diagnosis of ECD. BRAF mutation analysis was sent, and if positive, the patient will be started on vemurafenib.

Conclusion

ECD can manifest initially with neurological symptoms such as stroke, meningitis, and brain lesions and with vision symptoms and retro-orbital masses. Skeletal manifestation is more common, where bone scan is very useful and will show characteristic finding in ECD. FDG PET/CT shows that brain and orbital lesions can occur in malignancy. The coated aorta, which is a characteristic finding in ECD, if seen in those cases, we should raise the possibility of ECD.

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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Conflicts of interest

There are no conflicts of interest.

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