Egyptian Journal of Radiology and Nuclear Medicine

CASE REPORT

Open Access

An incidental finding of the Lincoln sign on FDG PET-CT in a patient with rectosigmoid adenocarcinoma giving rise to the diagnosis of Paget's disease

Muhammad Adib Abdul Onny[1], Sheila Shazlina Kemis Yahyah[1,2], Kavita Arumugam[1,2], Nor Salita Ali[1], Nashrulhaq Tagiling[3] and Norazlina Mat Nawi[3,4*]

Abstract

Background Paget's disease is a metabolic disorder characterized by disorderly bone remodeling process of excessive osteoclastic and osteoblastic activities leading to a structurally disorganized appearance of the bone. Paget's disease is usually diagnosed based on the clinical presentation of bony pain, raised serum alkaline phosphatase, and typical radiological changes. Bone scintigraphy with ^{99m}Tc-labeled radiotracers is commonly used to assess disease extent, and one of the classical findings of Paget's disease on bone scintigraphy is diffuse radiotracer uptake in the mandible bone, widely described as Lincoln or Black Beard sign. On the other hand, FDG PET-CT is commonly used in cancer imaging and frequently for staging or assessing recurrence in various malignancies. However, its use in evaluating Paget's disease is not established, attributed mainly to the heterogeneity of FDG uptake in Paget's disease and the high false-positive and negative findings.

Case presentation A 57-year-old female with metastatic mucinous rectosigmoid adenocarcinoma underwent multiple surgeries and completed 12 chemotherapy cycles with no evidence of local recurrence in the colon or distant metastasis. In the tenth year of active surveillance, her serum CEA levels were found to be elevated, yet there was no evidence of cancer spread from colonoscopy and contrast-enhanced CT. An ^{18}F-FDG PET-CT was then ordered, to which a unique diffuse FDG uptake pattern in the mandible was seen, resembling the Lincoln or Black Beard sign classically described in bone scintigraphy. This appearance was then verified by congruent uptake during a ^{99m}Tc-MDP scan, thus leading to the diagnosis of Paget's disease.

Conclusion The Lincoln sign is not limited to bone scintigraphy. Hence, we intend to add this FDG PET-CT finding to enrich the literature on the Lincoln sign and when to expect this pattern.

Keywords Paget's disease, FDG PET-CT, Bone scintigraphy, Lincoln sign, Black beard sign

*Correspondence:
Norazlina Mat Nawi
norazlina@usm.my
Full list of author information is available at the end of the article

Background

Paget's disease is a metabolic disorder characterized by disorderly bone remodeling process of excessive osteoclastic and osteoblastic activities leading to the structurally disorganized appearance of the bone [1]. Typically, bone scintigraphy using metastable-99 technetium (^{99m}Tc) labeled radiotracers is widely used to assess the distribution of this disease. Paget's disease is commonly seen as diffusely intense radiotracer uptake in solitary (monostotic) or several (polyostotic) bones. One of the most classical appearances on bone scintigraphy is the Black Beard sign or Lincoln sign, which is diffuse intense radiotracer uptake in the mandible bone, resembling the appearance of the famous 16th President of the United States of America [2]. Visualization of this sign is almost pathognomonic of Paget's disease. However, to the best of our knowledge, this sign has never been described in non-skeletal scintigraphy, and this case would probably be the first case relating the appearance of the Lincoln sign in florine-18 fluorodeoxyglucose positron emission tomography computed tomography (^{18}F-FDG PET-CT).

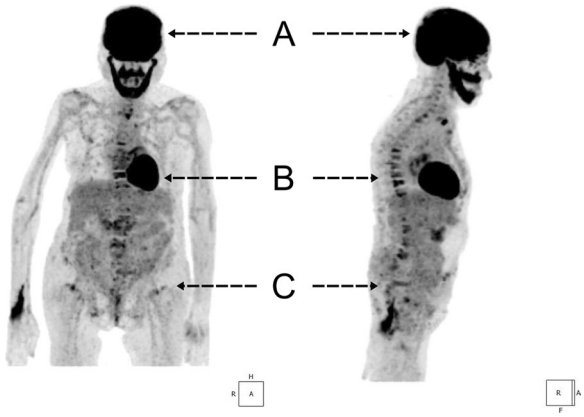

Fig. 1 A 57-year-old female patient with a history of treated metastatic mucinous rectosigmoid adenocarcinoma presented with occasional lethargy and reduced appetite with elevated CEA levels ten years into her active surveillance. The anterior and right lateral MIP of the ^{18}F-FDG PET-CT findings show diffuse increased FDG uptake in the skull and mandible bone (**A**), giving rise to the classical Lincoln or Black Beard sign. Heterogeneously diffuse FDG uptake of varying intensity is also seen along the vertebrae (**B**) and pelvic bones (**C**)

Case presentation

A 57-year-old Malay female from the East Coast of Malaysia (Kelantan) was diagnosed with metastatic mucinous adenocarcinoma of the rectosigmoid and had undergone anterior resection, cholecystectomy, and hysterectomy with excision of the liver nodule. She subsequently completed 12 cycles of chemotherapy, and the post-treatment ^{18}F-FDG PET-CT scan showed no evidence of local recurrence in the colon or any distant metastasis. She was under active surveillance, and in the 10th year, new constitutional symptoms of occasional lethargy and reduced appetite were noted, along with elevated serum carcinoembryonic antigen (CEA) levels from 2.9 µg/L to 5.8 µg/L. However, the colonoscopy examination and the contrast-enhanced computed tomography (CECT) showed no evidence of local recurrence or distant metastasis; hence, she was subjected to an ^{18}F-FDG PET-CT scan. Interestingly, her PET-CT scan showed heterogeneously diffuse FDG hypermetabolism in the skull bones, mandible, and maxilla, along with heterogeneous FDG metabolism involving the vertebrae, ribs, and pelvic bones (Fig. 1), corresponding to mixed lytic-sclerotic bony changes on CT (Fig. 2). In addition, the diffusely intense FDG hypermetabolism in the mandible seen on the maximum intensity projection (MIP) image gives rise to the appearance of the Lincoln or Black Beard sign frequently encountered on bone scintigraphy. Hence, the overall findings of the PET-CT were suggestive of concomitant bone metastasis and metabolic bone disease, with Paget's disease strongly considered. Biochemical examination revealed a significantly raised alkaline phosphatase (ALP) of > 800 IU/L and hypocalcemia of 1.9 mmol/L. She underwent a ^{99m}Tc-methylene diphosphonate (MDP) bone scintigraphy, which showed the presence of diffusely increased tracer uptake in the mandible consistent with the Lincoln sign and bowing of both femurs. The constellation of bone scintigraphy, PET-CT scan, and biochemical findings highly suggested Paget's disease. The patient, however, remained relatively asymptomatic as apart from the occasional lethargy and reduced appetite, she denies any significant bone pain, weakness of the limbs, or numbness. She also denies any difficulty in chewing, swallowing, or talking.

Discussion

Paget's disease (osteitis deformans) is a metabolic bone disease characterized by abnormal osteoclastic and osteoblastic activities characterized by haphazard remodeling of the bone [1]. It was first described in 1877 by Sir James Paget and is more commonly seen among the population but less widely seen among the Asian or Malaysian population [1, 3]. The disease can be divided into three phases: the resorptive/osteolytic phase, the mid-mixed osteoblastic/osteoclastic hyperplastic phase, and the late sclerotic phase [3].

Accurate disease diagnosis depends on the many clinical, biochemical, and radiological findings. Pain and bone deformity, e.g., bowing of the long bones, are the

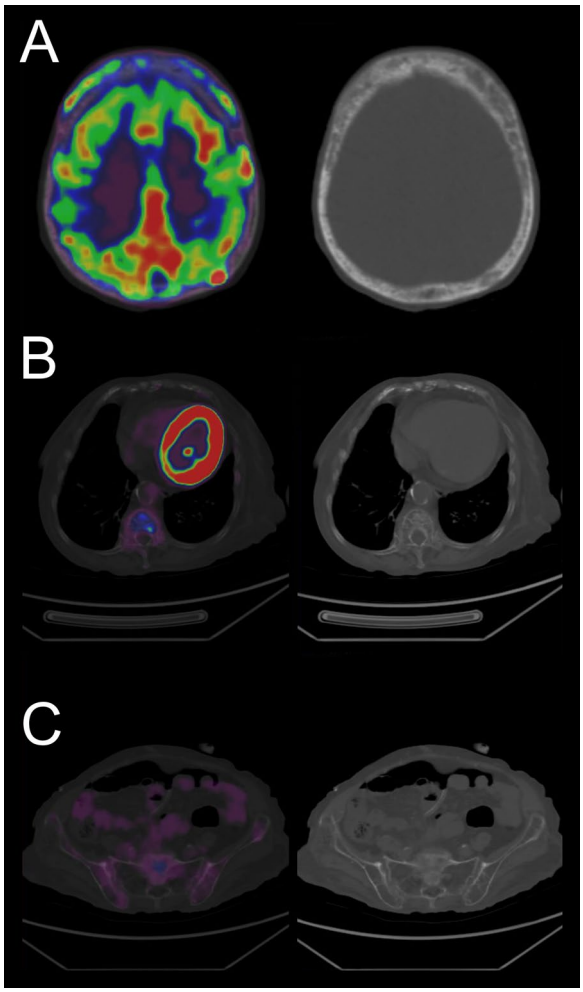

Fig. 2 Axial fused PET-CT and CT images of the skull (**A**), thoracic vertebra (**B**), and pelvic bones (**C**) showing background to mild increased FDG uptake in the visualized bones corresponding to mixed lytic-sclerotic lesions on CT

Fig. 3 Anterior and posterior planar images of ^{99m}Tc-MDP bone scintigraphy showing diffuse intense radiotracer uptake in the mandible bone in keeping with the Lincoln or Black Beard sign. Other increased tracer uptake is also seen at the lower thoracic vertebrae, similar to the FDG PET-CT findings. The prominent bowing of both femurs is also visualized

most typical clinical presentations, whereas elevated ALP is a useful biomarker of Paget's disease. Imaging plays an important in establishing the diagnosis, assessing the phases of the disease, the extent of the disease (monostotic or polyostotic), and assessing the integrity of the bone, which in turn helps evaluate the risk of complications [1].

Apart from conventional radiological examinations such as X-rays, nuclear medicine imaging plays a significant role in assessing Paget's disease. Bone scintigraphy has become an essential tool as it is sensitive in detecting early and subtle osteoblastic changes, usually not picked up on conventional X-rays. It has also been shown to visualize radiotracer uptake in the affected bones in all three phases [4, 5]. One of the most common findings on bone scintigraphy is the Lincoln or Black Beard sign, characterized by the diffusely intense radiotracer uptake in the body of mandible bone on ^{99m}Tc-MDP bone scintigraphy [2]. Due to the propensity of bone-seeking radiotracer uptake in Paget's disease, bone scintigraphy is commonly performed to establish the diagnosis and, more importantly, to determine the extent of bone involvement (Fig. 3).

On the other hand, ^{18}F-FDG PET-CT is not frequently used to assess Paget's disease [5]. Although indispensable for evaluating malignancy, its role in Paget's disease is still controversial. It was demonstrated that the FDG uptake in most patients with Paget's disease was variable and poor, even in those deemed active through ALP measurements [6]. Moreover, various false-positive FDG PET-CT findings of Paget's disease mimic metastasis, particularly in elderly patients [7].

As highlighted here, extensive mixed osteolytic–osteoblastic lesions in the skeletal system of this patient initially raised the suspicion of bone metastasis. However,

intense and diffuse FDG uptake throughout the mandible bone, giving rise to the Lincoln or Black Beard appearance corresponding to mixed lytic-sclerotic bone changes, prompted the possibility of concomitant metabolic disease, possibly Paget's disease. This led to further assessment, which showed significantly elevated serum ALP and a congruent radiotracer uptake pattern in the mandible on bone scintigraphy—confirming the diagnosis of Paget's disease and obviating an invasive biopsy procedure. To the best of our knowledge and literature review, this is probably the first case to report the concurrent appearance of the Lincoln or Black Beard sign in FDG PET-CT and bone scintigraphy.

Interestingly, despite the extensive mixed lytic-sclerotic bony changes, FDG uptake in these bones was remarkably heterogeneous, with some areas showing increased FDG uptake. In contrast, others showed only mild FDG uptake, consistent with several other publications that showed vast variations in the FDG uptake pattern and intensity in Paget's disease, which may mimic or mask metastatic foci in patients with underlying malignancy [7–13]. In addition, this case also highlights the variation of FDG uptake of Paget's disease, adding to the complexity of assessment of such disorder in patients with underlying malignancy.

Conclusions

In conclusion, FDG PET-CT should not be used solely in assessing Paget's disease but instead could be complementary in determining Paget's disease. The appearance of this finding in a patient with underlying malignancy should prompt clinicians to explore the possibility of Paget's disease, ensuring a holistic approach to patient management.

Abbreviations

^{99m}Tc	Metastable-99 technetium
^{18}F-FDG PET-CT	Florine-18 fluorodeoxyglucose positron emission tomography computed tomography
CEA	Carcinoembryonic antigen
CECT	Contrast-enhanced computed tomography
MIP	Maximum intensity projection
ALP	Alkaline phosphatase
MDP	Methylene diphosphonate

Acknowledgements

The authors thank the patient and her family for providing informed consent to use her de-identified clinical data and images for knowledge sharing and publication.

Author contributions

MAAO, SSKY, NSA, and NMN handled the interpretation of imaging results and initial management of the patient. NT and NMN were responsible for editing and proofreading the manuscript for submission. All authors made substantial writing, read, and approved the final manuscript.

Funding

This study was supported in part by the Ministry of Higher Education Malaysia (Fundamental Research Grant Scheme [FRGS], Ref: FRGS/1/2020/SKK08/USM/02/1).

Availability of data and materials

The authors obtained all the data and materials relevant to this case report from the patient's electronic and medical records from our respective institutions. All data used during this case report are included in this published article.

Declarations

Ethics approval and consent to participate

Verbal informed consent was obtained from the patient to participate in this case report.

Consent for publication

The authors have obtained written informed consent from the patient to publish the case report details and related images.

Competing interests

The authors declare that they have no competing interests.

Author details

[1]Nuclear Medicine Department, Institut Kanser Negara, 62250 Putrajaya, Malaysia. [2]Nuclear Medicine Unit, Radiology and Oncology Cluster, Advanced Medical and Dental Institute, Universiti Sains Malaysia, 13200 Kepala Batas, Pulau Pinang, Malaysia. [3]Department of Nuclear Medicine, Radiotherapy and Oncology, School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kota Bharu, Kelantan, Malaysia. [4]Hospital Universiti Sains Malaysia, Universiti Sains Malaysia, 16150 Kota Bharu, Kelantan, Malaysia.

Received: 19 October 2023 Accepted: 25 December 2023
Published online: 15 January 2024

References

1. Theodorou DJ, Theodorou SJ, Kakitsubata Y (2011) Imaging of Paget disease of bone and its musculoskeletal complications: review. Am J Roentgenol 196(6):S64–S75. https://doi.org/10.2214/ajr.10.7222
2. Bal CS, Sahoo MK, Damle N (2013) Lincoln's sign where should we expect on 99mTc-MDP bone scintigraphy? Clin Nucl Med 38(10):e390–e391. https://doi.org/10.1097/RLU.0b013e318270851c
3. Lim KP, Kok WH, Kamaruddin NA (2018) Metastatic bone disease secondary to bronchial adenocarcinoma in a patient with Paget's disease of the bone. J ASEAN Fed Endocr Soc 33(1):63–68. https://doi.org/10.15605/jafes.033.01.11
4. Kumar AA, Kumar P, Prakash M, Tewari V, Sahni H, Dash A (2013) Paget's disease diagnosed on bone scintigraphy: case report and literature review. Indian J Nucl Med 28(2):121–123. https://doi.org/10.4103/0972-3919.118258
5. Cortis K, Micallef K, Mizzi A (2011) Imaging Paget's disease of bone—from head to toe. Clin Radiol 66(7):662–672. https://doi.org/10.1016/j.crad.2010.12.016
6. Ramírez-Sanabria AD, Valero MA, Mantilla-Hernández RD, Ordóñez-Rubiano EG (2023) The role of 18F-FDG PET/CT for diagnosis of Paget's disease of bone with cranial and spinal compromise: Case report and literature review. Revista Colombiana de Reumatología (English Edition) 30(2):166–172. https://doi.org/10.1016/j.rcreue.2021.05.009
7. Dondi F, Albano D, Treglia G, Bertagna F (2022) Paget disease as common pitfall on PET with different radiopharmaceuticals in oncology: not all that glitters is gold! J Clin Med 11(18):1. https://doi.org/10.3390/jcm11185372
8. Mahmood S, Martinez de Llano SR (2008) Paget disease of the humerus mimicking metastatic disease in a patient with metastatic malignant mesothelioma on whole body F-18 FDG PET/CT. Clin Nucl Med 33(7):510–512. https://doi.org/10.1097/RLU.0b013e318177928a

9. Zhong X, Ye Y, Ou X (2017) Impressive paget disease of the lumbar spine masks the coexisting multiple myeloma. Clin Nucl Med 42(9):e417–e421. https://doi.org/10.1097/rlu.0000000000001733
10. Woo JH, Kim S, Choi SJ, Lee YH, Ji JD, Song GG (2010) Diagnosis of Paget's disease of the pelvis using F-18 FDG PET/CT. Int J Rheumat Dis 13(4):1. https://doi.org/10.1111/j.1756-185X.2010.01538.x
11. Sasikumar A, Joy A, Pillai MRA, Raman V, Vasudevan A, Madhavan J (2016) A rare case of rectal carcinoma and prostate carcinoma with coexistent Paget's disease mimicking bone metastases in both 18F-FDG and 68Ga PSMA PET/CT. Eur J Nucl Med Mol Imaging 44(1):173. https://doi.org/10.1007/s00259-016-3529-8
12. Kamaleshwaran K, Natarajan S, Shibu D, Malaikkal A, Shinto A (2015) Paget´s disease of pelvis mimicking metastasis in a patient with lung cancer evaluated using staging and follow-up imaging with fluorine-18 fluorodeoxyglucose-positron emission tomography/computed tomography. Indian J Nucl Med 30(2):1. https://doi.org/10.4103/0972-3919.152980
13. Park ET, Kim S-E (2010) Radiography, bone scan, and F-18 FDG PET/CT imaging findings in a patient with Paget's disease. Nucl Med Mol Imaging 44(1):87–89. https://doi.org/10.1007/s13139-009-0013-4

Publisher's Note