Nuclear Medicine and Molecular Imaging (2019) 53:436–441
https://doi.org/10.1007/s13139-019-00616-8

INTERESTING IMAGE

ISSN (print) 1869-3482
ISSN (online) 1869-3474

Intramastoid Phosphaturic Mesenchymal Tumor Causing Hypophosphatemic Osteomalacia Detected on ^{68}Ga-DOTATATE PET/CT But Not on ^{99m}Tc-Sestamibi and ^{18}F-FDG Scans

Carlo Scognamiglio Renner Araujo[1] · Luciana Parente Costa Seguro[1] · Paulo Schiavom Duarte[2,3] · Carlos Alberto Buchpiguel[2] · Rosa Maria Rodrigues Pereira[1]

Received: 5 July 2019 / Revised: 2 September 2019 / Accepted: 24 September 2019 / Published online: 14 November 2019
© Korean Society of Nuclear Medicine 2019

Abstract

^{68}Ga-DOTATATE uptake in mesenchymal tumors causing hypophosphatemic osteomalacia has been recently described. Herein, we present a case of ^{68}Ga-DOTATATE uptake in an intramastoid phosphaturic mesenchymal tumor that had not been depicted in previous ^{99m}Tc-Sestamibi and ^{18}F-FDG scans. The lesion was surgically removed and the phosphorus level increased to the normal range.

Keywords Hypophosphatemic · Osteomalacia · Mastoid · Gallium-68 · DOTA · PET/CT

✉ Paulo Schiavom Duarte
psduarte@hotmail.com

[1] Rheumatology Division, Hospital das Clínicas HCFMUSP, Faculdade de Medicina, Universidade de São Paulo, São Paulo, SP, Brazil

[2] Nuclear Medicine Division, Hospital das Clínicas HCFMUSP, Faculdade de Medicina, Universidade de São Paulo, São Paulo, SP, Brazil

[3] Nuclear Medicine Section, São Paulo Cancer Institute (ICESP), Av. Dr. Arnaldo, 251, 4° SS Cerqueira César, São Paulo, SP 01246-000, Brazil

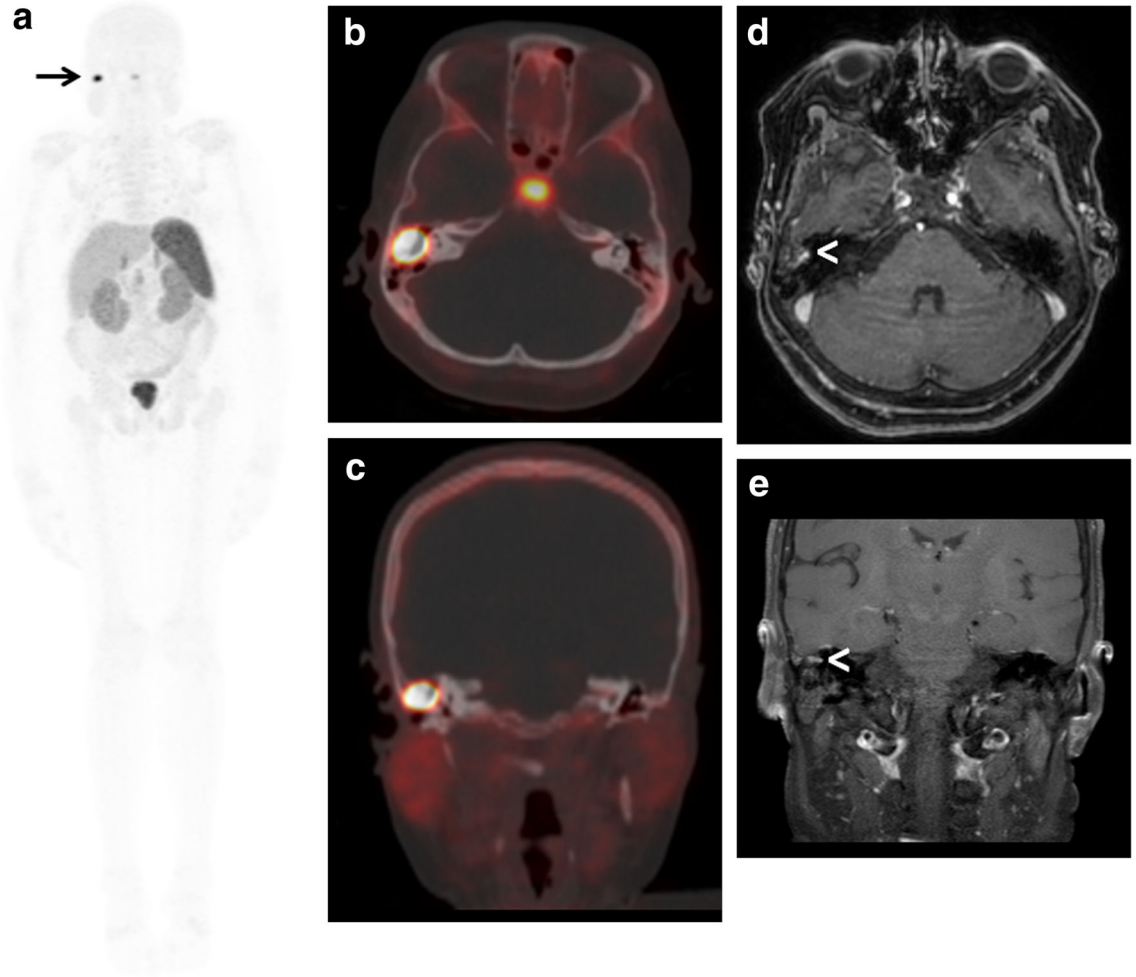

Fig. 1 A 44-year-old female with a few years of history of hypophosphatemic osteomalacia (HO) suspected to be caused by an unknown primary tumor—tumor-induced osteomalacia (TIO)—that had not been detected by other imaging studies was scanned with ^{68}Ga-DOTATATE whole-body positron emission tomography/computed tomography (PET/CT) in an attempt to localize the lesion. The ^{68}Ga-DOTATATE PET coronal maximum-intensity-projection (MIP) image (**a**) demonstrated an intense focal uptake in the right temporal region (arrow). The transversal (**b**) and coronal (**c**) ^{68}Ga-DOTATATE PET/CT images localize the region of uptake inside the right mastoid. The transversal (**d**) and coronal (**e**) images of a magnetic resonance study of the mastoid performed a few days later confirmed a millimetric lesion inside the right mastoid air cells (arrowhead)

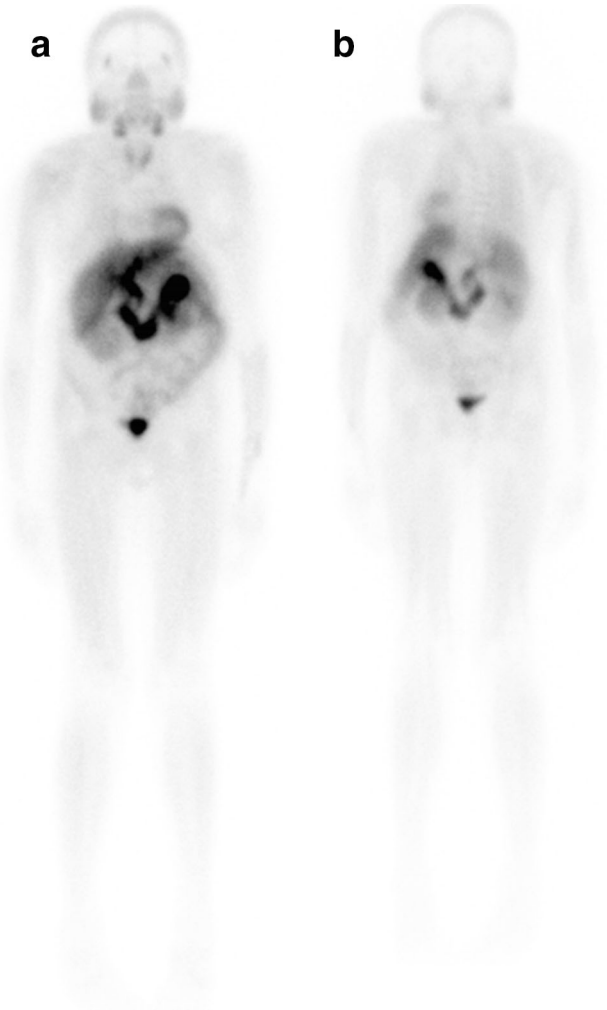

Fig. 2 The ^{99m}Tc-Sestamibi anterior (**a**) and posterior (**b**) whole-body images performed 2 days before the ^{68}Ga-DOTATATE PET/CT did not show any abnormal uptake

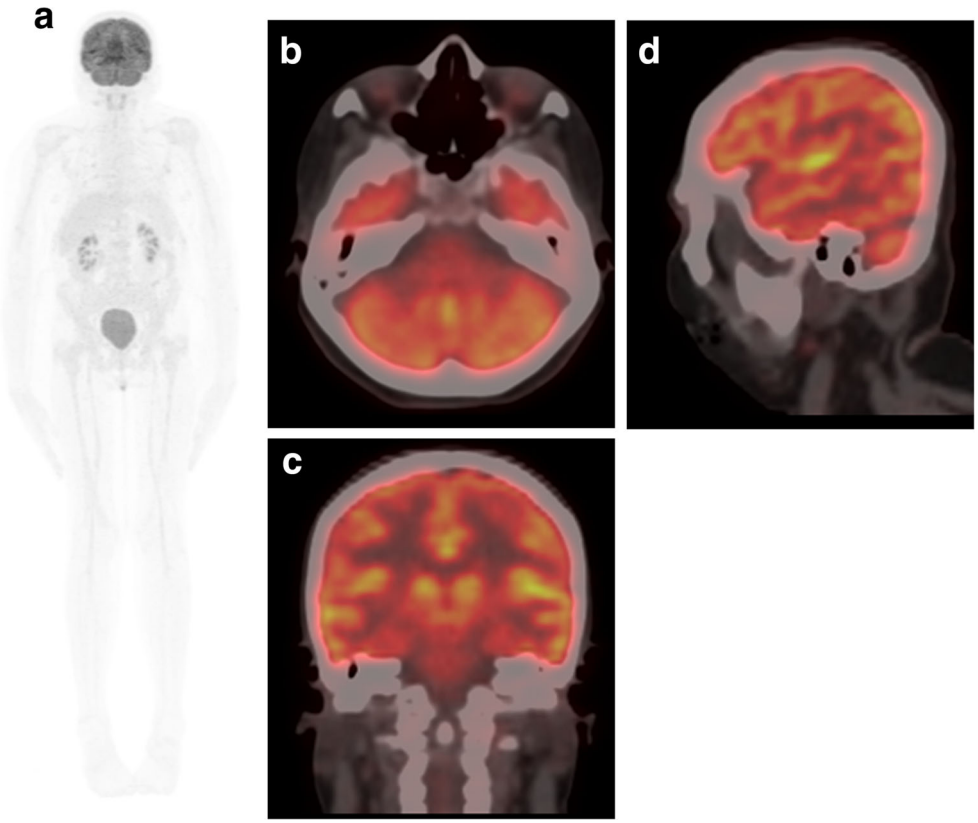

Fig. 3 The images of an ^{18}F-FDG PET/CT study performed previously also did not show any abnormal uptake as demonstrated on MIP coronal images (**a**) and the transversal (**b**), coronal (**c**), and sagittal (**d**) PET/CT images at the right mastoid level

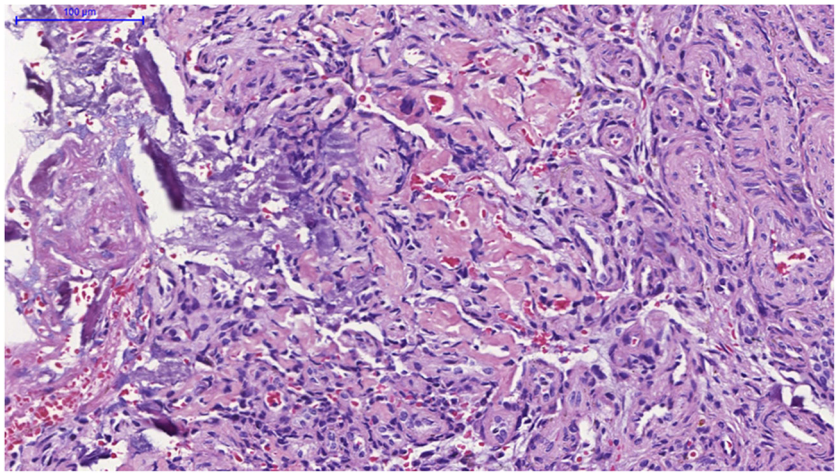

Fig. 4 The microscopic image demonstrated features suggestive of phosphaturic mesenchymal tumor with short strands of slightly atypical fusiform cells with very low mitotic rate, focal stromal hyalinization, and prominent vascularization. TIO is a rare paraneoplastic syndrome characterized by severe hypophosphatemia and osteomalacia that may be associated with benign mesenchymal tumors [1]. In as much as the surgical excision of the tumor can be curative, the localization of the lesion is an important task [2]. However, the tumor can be very small and localized anywhere in the body [2]. After the discovery of somatostatin receptors in mesenchymal tumors [3], the use of radiolabeled somatostatin analogue to find the primary tumor in patients with HO has been described. The ^{111}In-octreotide was the first somatostatin analogue to be used in the localization of the primary lesion in patients with TIO [4, 5]. But, in the last years, the ^{68}Ga-labeled somatostatin analogues have been used in places where PET/CT is available with high accuracy [6]. Thus, nowadays, ^{68}Ga-labeled somatostatin analogues may be the best imaging method to be used in the detection of the culprit lesion in cases of TIO [7]. However, a direct comparison of ^{68}Ga-labeled somatostatin analogue with other radiopharmaceuticals has rarely been performed. In an analysis of 4 patients with TIO, it was described that the ^{68}Ga-DOTATATE was capable to detect the primary lesion in 3 and the ^{18}F-FDG in 2 of them [8], and, in a recent article, it was shown that ^{68}Ga-DOTATOC PET/CT was able to detect the TIO in some of the patients with negative results on ^{111}In-pentetreotide SPECT/CT and on ^{18}F-FDG PET/CT scans [9]. Therefore, the presented case brings two interesting aspects: first, it shows the capability of the ^{68}Ga-DOTATATE PET/CT to detect a phosphaturic mesenchymal tumor in an unusual site and, second, it shows that this radiopharmaceutical is superior to the ^{99m}Tc-Sestamibi and ^{18}F-FDG scans in this detection. This superiority could be explained by the diffuse strong expression of somatostatin receptors in the phosphaturic mesenchymal tumors [10] by the fact that most of them are benign and present low cellularity and low mitotic activity [11], and by the variable number of mitochondria in the tumor cells with few organelles in some cases [12]

Acknowledgments The authors are grateful to Drs. Vitor Gabriel Ribeiro Grossi and Eloiza Wilma Poma Gonzales for providing the histological images.

Compliance with Ethical Standards

Conflict of Interest Carlo Scognamiglio Renner Araujo, Luciana Parente Costa Seguro, Paulo Schiavom Duarte, Carlos Alberto Buchpiguel, and Rosa Maria Rodrigues Pereira declare that they have no conflict of interest.

Ethical Statement All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent The institutional review board of our institute approved this study, and the requirement to obtain informed consent was waived.

References

1. Jan de Beur SM. Tumor-induced osteomalacia. JAMA. 2005;294:1260–7.
2. Weidner N, Bar RS, Weiss D, Strottmann MP. Neoplastic pathology of oncogenic osteomalacia/rickets. Cancer. 1985;55:1691–705.
3. Reubi JC, Waser B, Laissue JA, Gebbers JO. Somatostatin and vasoactive intestinal peptide receptors in human mesenchymal tumors: in vitro identification. Cancer Res. 1996;56:1922–31.
4. Rhee Y, Lee JD, Shin KH, Lee HC, Huh KB. Lim SK Oncogenic osteomalacia associated with mesenchymal tumour detected by indium-111 octreotide scintigraphy. Clin Endocrinol. 2001;54:551–4.
5. Nguyen BD, Wang EA. Indium-111 pentetreotide scintigraphy of mesenchymal tumor with oncogenic osteomalacia. Clin Nucl Med. 1999;24:130–1.
6. Zhang J, Zhu Z, Zhong D, Dang Y, Xing H, Du Y, et al. 68Ga DOTATATE PET/CT is an accurate imaging modality in the

detection of culprit tumors causing osteomalacia. Clin Nucl Med. 2015;40:642–6.
7. Clifton-Bligh RJ, Hofman MS, Duncan E, IeW S, Darnell D, Clarkson A, et al. Improving diagnosis of tumor-induced osteomalacia with gallium-68 DOTATATE PET/CT. J Clin Endocrinol Metab. 2013;98:687–94.
8. Agrawal K, Bhadada S, Mittal BR, Shukla J, Sood A, Bhattacharya A, et al. Comparison of 18F-FDG and 68Ga DOTATATE PET/CT in localization of tumor causing oncogenic osteomalacia. Clin Nucl Med. 2015;40:e6–e10.
9. Paquet M, Gauthé M, Yin JZ, Nataf V, Bélissant O, Orcel P, et al. Diagnostic performance and impact on patient management of 68Ga-DOTA-TOC PET/CT for detecting osteomalacia-associated tumours. Eur J Nucl Med Mol Imaging. 2018;45:1710–20.
10. Houang M, Clarkson A, Sioson L, Elston MS, Clifton-Bligh RJ, Dray M, et al. Phosphaturic mesenchymal tumors show positive staining for somatostatin receptor 2A (SSTR2A). Human Pathol. 2013;44:2711–8.
11. Folpe AL, Fanburg-Smith JC, Billings SD, Bisceglia M, Bertoni F, Cho JY, et al. Most osteomalacia-associated mesenchymal tumors are a single histopathologic entity: an analysis of 32 cases and a comprehensive review of the literature. Am J Surg Pathol. 2004;28:1–30.
12. Weidner N, Cruz DS. Phosphaturic mesenchymal tumors. A polymorphous group causing osteomalacia or rickets. Cancer. 1987;59:1442–54.

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.