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Case Report

Clinically occult image revelations of fluorodeoxyglucose positron emission tomography/computed tomography in a peculiar case of sinonasal myeloid sarcoma

ABSTRACT

Myeloid sarcoma (MS) is a sparse association of acute myeloid leukemia (AML) with poor overall survival. Sinonasal involvement in MS is rarer and meagerly studied. Recent prospective studies have underlined the importance of positron emission tomography/computed tomography (PET/CT) in disease assessment and relapse detection in extramedullary (EM) AML. Herein, we report a case of MS with sinonasal, orbital, dural, breast, pleural extent of disease with sacral neural foraminal compression. Initial histopathologic and clinicoradiologic features had pointed toward lymphoma. Immunohistochemistry (IHC) on the contrary revealed MS. Although the role of IHC in asserting the diagnosis of MS is remarkable, PET/CT surpasses other modalities in early detection of EM sites of involvement, overall assessment of disease burden, targeting therapy, prognostication, and relapse recognition. Inclusion of PET/CT as a baseline imaging modality can be beneficial in tailoring patient management to improve survival and overall quality of life.

Keywords: Immunohistochemistry, positron emission tomography in extramedullary acute myeloid leukemia, positron emission tomography in myeloid sarcoma, sinonasal myeloid sarcoma

INTRODUCTION

Myeloid or granulocytic sarcoma constitutes extramedullary (EM) proliferation of myeloblasts with resultant tissue effacement. It can either herald or be concurrent with or follow the onset of acute myeloid leukemia (AML) in 2.5%–9.1% of AML patients.[1] Maladjusted homing signals due to the overexpression and abnormal interaction of matrix metalloproteinases and leukocyte surface integrins play a vital role in myeloblast propagation to extramedullary location.[2] The sites of predilection of myeloid sarcoma (MS) include lymph nodes, bone, central nervous system, soft tissue, mediastinum, lung, uterus, ovaries, and gastrointestinal tract. Nasal and sinus involvement are seldom noticed. Lack of large prospective clinical trials and availability of merely a handful of case reports of sinonasal MS adds to its recondite nature. Clinical, pathological, and imaging similarity to lymphoma, Langerhans cell histiocytosis, and poorly differentiated carcinoma contributes to the reporting dilemma.

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CASE REPORT

A 33-year-old female with complaints of nasal obstruction, head ache, right-sided proptosis, and hearing loss for the past 6 months was referred to our department for ^{18}F-fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) scan. Nasal endoscopy revealed a grayish white nasal mass extending into the posterior nasal cavity. Endoscopy-guided biopsy of the nasal

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mass was done. Initial histopathology suggested poorly differentiated malignant neoplasm. PET/CT from the vertex to the mid-thigh was performed after IV administration of 250 MBq of ^{18}F-FDG.

Maximum intensity projection images of ^{18}F-FDG PET/CT showed abnormal increased tracer uptake in the sinuses, breasts, retrosternal, and sacral region [Figure 1]. An FDG-avid ill-defined soft-tissue mass, measuring 4.5 (AP) × 5.2 (T) × 4.0 (CC) cm, involving the bilateral nasal cavity, ethmoid, sphenoid, and maxillary sinuses with right intraorbital extension, displacing right medial rectus, and posteriorly extending into the nasopharynx with maximum standardized uptake value (SUV) of 9.9, was seen [Figure 2]. Hypermetabolic focal pleural thickening in the bilateral hemithorax and multiple well-defined lobulated homogenously enhancing deposits in the bilateral breasts with SUV_{max} of 4.5 were noted [Figure 3]. There were associated FDG-avid soft-tissue thickening in the retrosternal region, dural deposit in the high parietal region, and ill-defined soft-tissue thickening in the spinal canal at S1 vertebral level extending into the 1st left sacral neural foramen with SUV_{max} of 8 [Figure 3]. Non-FDG-avid soft-tissue deposits involving the bilateral lacrimal glands were also found. Overall imaging features were more in favor of lymphoma.

Immunohistochemistry (IHC) results conversely detected MS. IHC showed Ki index of 80%–90% and diffuse positivity for vimentin, myeloperoxidase, leukocyte common antigen, CD43, CD34, and C-KIT [Figure 4]. Retrospective analysis of her preliminary blood reports revealed 27% blasts and her peripheral blood smear was indicative of acute leukemia.

However, the patient did not consent for a bone marrow biopsy. Although the peripheral smear suggested acute leukemia, the association with AML in this case could not be confirmed.

DISCUSSION

In our patient, as the major disease burden concentrated in the sinonasal region, with an infiltrative pattern of involvement, the possibility of sarcoma was considered initially. However, after further scrutiny of the overall

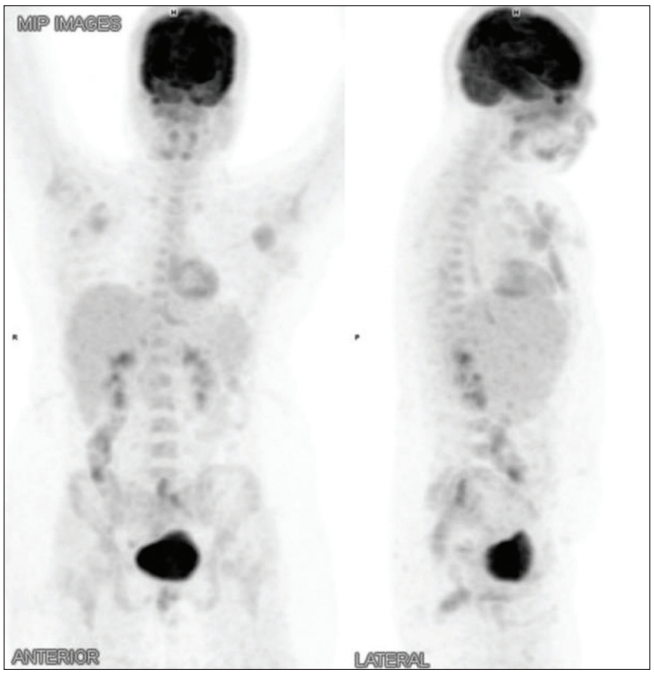

Figure 1: Maximum intensity projection of 18-fluorodeoxyglucose positron emission tomography/computed tomography scan showing abnormal increased tracer uptake in the sinuses, breasts, retrosternal, and sacral region

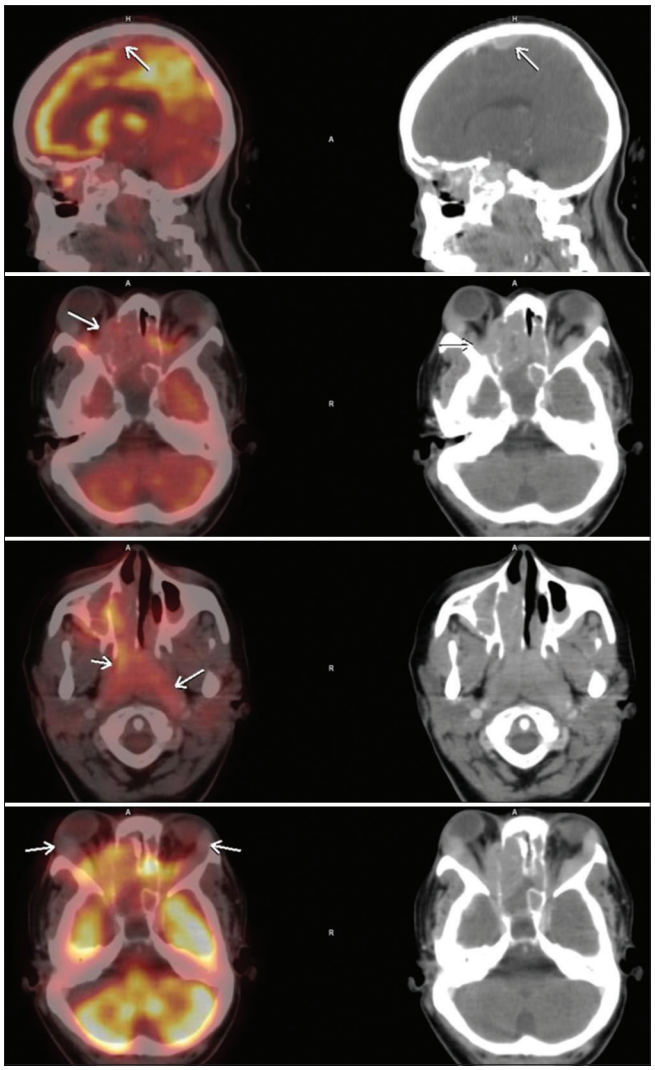

Figure 2: Positron emission tomography/computed tomography images of head and neck showing fluorodeoxyglucose-avid dural deposit in the high parietal region and ill-defined soft-tissue mass involving bilateral nasal cavity, ethmoid sphenoid, and maxillary sinuses with non-fluorodeoxyglucose-avid soft-tissue deposits in bilateral lacrimal glands

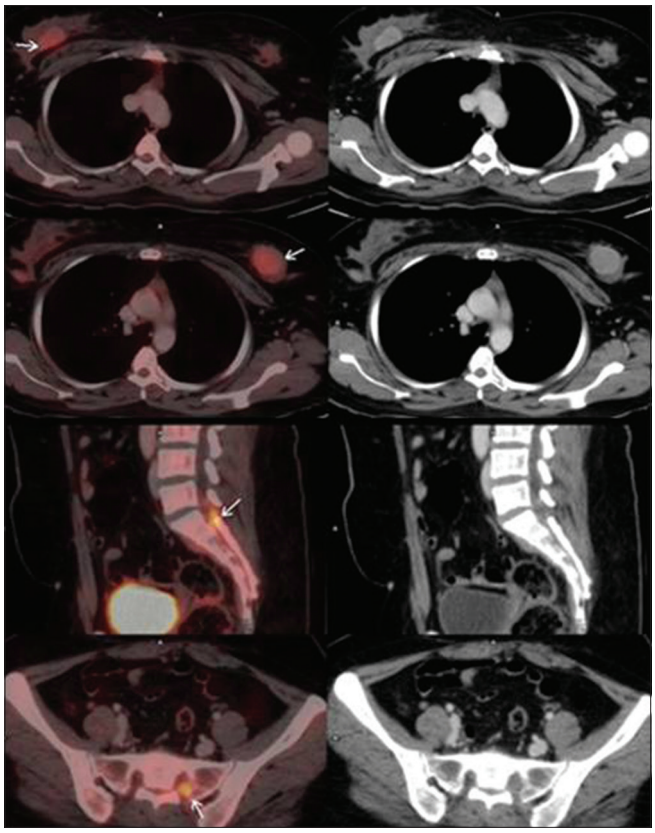

Figure 3: Positron emission tomography/computed tomography images showing fluorodeoxyglucose-avid deposits in bilateral breasts and ill-defined soft-tissue thickening in spinal canal at S1 vertebral level extending into 1st left sacral neural foramen

disease distribution, sinonasal mass with dural, pleural, well-circumscribed lobulated breast deposits and soft-tissue thickening involving spinal canal, primary differential was narrowed down to lymphoma. Sinonasal involvement in MS being extremely rare an entity, differential diagnosis of myeloid sarcoma was overlooked. The clinching step toward the diagnosis of MS here was the IHC findings. *Ex post facto* review of the hemogram and peripheral blood smear results of the patient surprisingly unfolded preimaging possibility of MS, underscoring the need of detailed assessment of patient history and simple laboratory parameters before concluding imaging differentials.

Even though the diagnosis of MS involves a multidisciplinary approach, it is to be emphasized that PET/CT as a baseline imaging technique gives a lucid picture of the overall disease burden. In our patient, there was no symptomatic or clinical indication toward pleural, dural, paraspinal, or neural foramen soft-tissue involvement, which was brought to light by PET/CT. Prior studies conducted by Aschoff *et al*., Stölzel *et al*., and Cribe *et al*., substantiate our claim of significantly higher detection of clinically silent EM sites of involvement in MS.[3-5] The recently concluded prospective PET AML study

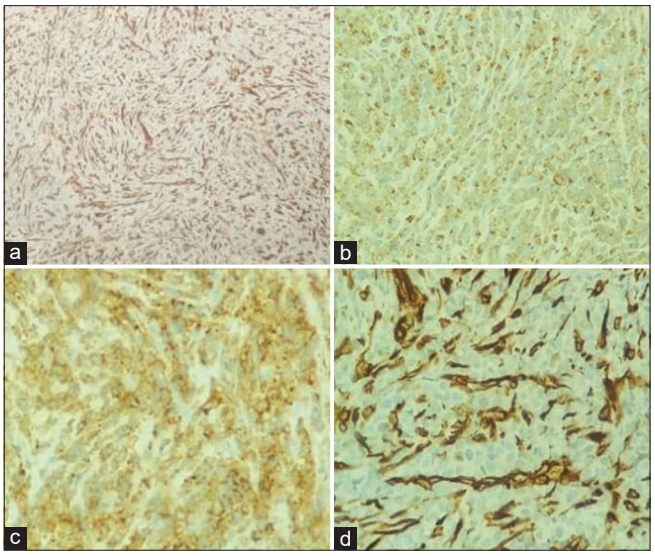

Figure 4: Immunohistochemistry images showing (a) vimentin positive, (b) myeloperoxidase positive, (c) leukocyte common antigen positive, (d) CD43 positive

surmised that ^{18}F-FDG PET/CT had a sensitivity of 90% in EM AML and discerned additional EM sites in 60% of patients.[6]

The association of increased expression of matrix metalloproteinase in MS in itself suggests poor prognosis, though adequate documentation to this effect is lacking. There have been prior retrospective studies eliciting poor median survival of 5.4 months in AML patients with MS in comparison to 59.5 months in AML patients without MS.[4] This implies the early utilization of superior imaging modality like PET/CT in expediting the detection, targeting biopsy, and in thorough assessment of the whole-body disease extent. Knowledge of all the sites involved can also help us in planning targeted radiotherapy in addition to the conventional chemotherapy and thereby arrest the progression of the disease as well as development of grave symptoms and hence improve the overall quality of life (QOL).

PET AML trial also revealed that ^{18}F-FDG PET/CT discerned relapses in patients who were in remission as per bone marrow findings, adding to the profitability of PET/CT in MS.[6] Prognostic value of glucose metabolism in AML patients is established in prior studies.[7] Extrapolation of the same in MS can prove to be of benefit in using FDG PET/CT in response assessment and relapse identification in this subset of patients.

CONCLUSION

Sinonasal involvement in MS is a rarity on its own. Holistic evaluation of patient history and basic laboratory parameters is mandated preceding imaging diagnosis in such cases. Imaging

suspicions should always be confirmed with IHC in similar nonspecific occurrences. Although clinical, pathological, and imaging deceptions command for a multimodality approach in the diagnosis of sinonasal MS, the contribution of PET/CT in early detection, mapping, and prognostication is becoming crucial in panning the treatment choices and improving the overall survival and QOL in these patients.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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