Synchronous Detection of Extra-Adrenal Paraganglioma in a Follow-Up Case of High-Grade Gastrointestinal Stromal Tumor

Abstract

Extraadrenal paragalionoma at multiple sites and its association with GIST is a rare finding. Although ^{18}F-FDG PET-CT is not a modality of choice for primary diagnosis of paraganglioma, and its use is restricted for detection of metastases/multiple sites and disease staging. However, in this case that we describe here, its role in an already-proven/recurrent case of paraganglioma is emphasized by its simultaneous assessment of disease at several different sites.

Keywords: *Aortic pulmonary ganglion, fluorodeoxyglucose positron emission tomography-computed tomography, paraganglioma*

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We present an intresting image of a follow up and post operated case of Glomus vagale(left) and Glomus jugulare (right) GIST stomach. The case is a 44yr-old young man, a k/c/o Glomus vagale Lt (excision 2009, thyroplasty done), Glomus jugulare (excision 2009, GKS done) and high grade GIST (infra pyloric region, resection of lesion in Aug 2013, followed by Tab Imatinib since then).

The patient is on follow up since 2013, and multiple fluorodeoxyglucose positron emission tomography computed tomography (PET CT) scans were done during this period. On a recent follow-up, there was persistence of metabolic active lesions at multiple Paraganglioma sites, with no change in size. These sites were at the left jugular fossa (jugulotympanic ganglion/glomus jugulare), at the left cervical level II (glomus vagale) and close to the right atrium (aortic pulmonary ganglion/aortico pulmonary ganglion) [Figure 1a-d].

The last scan dated July 8, 2020, reveals that in addition to the above lesions (paraganglion), there is recurrence at the primary site (GIST – FDG-avid endophytic lesion in the pylorus) [Figure 1e-j]. A recent Contrast Enhanced Magnetic Resonance Imaging (CEMRI) brain showed postoperative status, with no significant change in the extent of lesion in the left jugular fossa.

Cardio Thoracic Vascular Surgery (CTVS) surgeon suggested no active intervention for the lesion near the right atrium. The patient is asymptomatic presently.

Extra adrenal paraganglioma is a relatively uncommon entity (incidence-1/300000) and especially at multiple sites is a rare phenomenon.[1] While majority of them are asymptomatic or nonclinical, hardly 1-3% are secretary in nature. Paraganglias can be divided into parasympathetic or sympathetic types, are collections of neuroendocrine cells at various locations in the body (involving neck, thorax, and trunk).[2] Although mostly benign and slow growing, paraganglionomas have a high recurrence rate. There is a established role of Mutations in SDH complex genes which is predisposing to gastrointestinal stromal tumors (GIST) and familial paraganglioma-pheochromocytoma syndrome (FPG).[3] Carney triad, (described in 1977) is a triad of 3 tumors namely gastrointestinal stromal tumor (GIST), extra-adrenal paraganglioma, and pulmonary chondroma.[4] In this case FDG PET CT did simultaneous assessment of disease at several different sites (glomus vagale, glomus jugulare, and aorticopulmonary ganglion) with a description of stability of metabolic activity and size during an extended course of time

How to cite this article: Husain A, Jain A, Singh H, Hegde A. Synchronous detection of extra-adrenal paraganglioma in a follow-up case of high-grade gastrointestinal stromal tumor. Indian J Nucl Med 2021;36:205-6.

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Received: 19-08-2020
Revised: 24-08-2020
Accepted: 15-09-2020
Published: 21-06-2021

Access this article online

Website: www.ijnm.in

DOI: 10.4103/ijnm.IJNM_193_20

Quick Response Code:

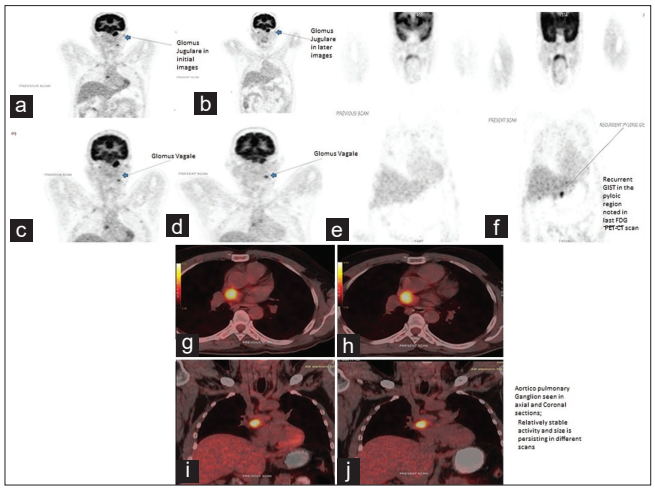

Figure 1: Maximum Intensity Projection (MIP) image showing metabolically active glomus jugulare and vagale in the initial images (a and c) and during the follow-up images (b and d) MIP image showing metabolic lesion of gastrointestinal stromal tumor in the stomach region in follow-up scan (f) that was not appreciated in the initial images (e). Axial and sagittal section positron emission tomography-computed tomography scan showing fluorodeoxyglucose-avid lesion in the mediastinum, closely abutting the right pulmonary artery, right atrium inferiorly, superior vena cava with preserved fat planes (aorticopulmonary ganglioma) in the initial (g and i) and follow-up (h and j) images

(no clinical progression is noted as well). Furthermore, its association with a special malignancy (GIST) is likely due to common genetic alterations.[5-7]

In our case of multiple head-and-neck paragangliomas and GIST, FDG PET scan showed the true extent of disease at follow-up. Overall, FDG PET-CT scan findings indicated no significant changes in multiple paraganglions, thus giving no definite indication for any active intervention. However, there is recurrence of GIST in pylorus of the stomach despite being on tyrosine kinase inhibitors, hence active intervention is contemplated here. Thus, the role of FDG PET-CT scan can also be seen in giving directions for treatment.

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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