Clinical usefulness of ^{68}Ga -DOTA-NOC PET/CT in staging a vagal paraganglioma associated with a novel *SDHB* mutation

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DESCRIPTION

Paragangliomas are rare tumours arising from the autonomic nervous system. Most are benign, however, a few are malignant. The diagnosis of malignancy is dependent on the evidence of metastases. Approximately, 40% of all paragangliomas/pheochromocytomas harbour a germ-line mutation in one of the susceptibility genes identified so far.¹

We present a case of a 29-year-old Caucasian man, referred to us on suspicion of a neck paraganglioma based on CT findings. MRI confirmed a lesion (80×40×40 mm) within the right parapharyngeal space, slightly intense on T2 and posterior to the carotid bifurcation, likely to be a vagal paraganglioma; 24 h urine metanephrines were normal. A subtotal resection was performed due to incarceration of the internal carotid artery in the carotid canal of the skull base. Histopathological examination established the diagnosis of malignant paraganglioma based on the

documentation of a metastatic lymph node (figure 1). Following surgery, the patient was submitted to Intensity Modulated Radiation Therapy (total dose 54 Gy).

Genetic testing disclosed a novel germ-line mutation in the *SDHB* gene (c.721T>G; p.Tyr241Asp), predicted to be deleterious by in silico analysis using SIFT and PolyPhen software. The same mutation was identified in the patient's father, who is known to have Parkinson's disease and who is awaiting imaging examinations to exclude paragangliomas.

To screen for synchronous paragangliomas and/or distant metastases,^{2 3} a ^{68}Ga -DOTA-NOC positron emission tomography/CT was performed (figure 2). Our patient's examination documented, as expected, right parapharyngeal uptake, excluded synchronous paragangliomas and revealed left acetabular uptake consistent with bone metastasis, making the patient a candidate for lutetium therapy.

Figure 1 (A) Vagal paraganglioma (H&E staining, ×200); (B) Metastatic lymph node (chromogranin A positive staining, ×100); (C) Metastatic lymph node (synaptophysin positive staining, ×100).

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Figure 2 ^{68}Ga -DOTA-NOC positron emission tomography/CT: uptake in the vagal paraganglioma (arrow 1) and the left acetabular metastatic lesion (arrow 2).

Learning points

- ▶ A novel mutation identified in the *SDHB* gene (c.721T>G; p. Tyr241Asp) confirms the high malignant potential of tumours related to *SDHB* germ-line mutations.
- ▶ The development of somatostatin (SST) analogues radiolabelled with ^{68}Ga for positron emission tomography/X-ray CT (positron emission tomography/CT) imaging, such as ^{68}Ga -DOTA-NOC, improved the diagnosis and follow-up of patients with SST receptor (SSTR)-positive tumours.

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