A 30-year-old man with suspected neuroendocrine tumor (NET), due to persistently increased chromogranin-A serum levels (>300 U/L), underwent somatostatin receptor positron emission tomography (PET)/computed tomography (CT) using Gallium-68-DOTANOC (68Ga-DOTANOC-PET/CT) to look for a possible NET.

68Ga-DOTANOC PET/CT showed a focal area of increased radiopharmaceutical uptake (maximum standardized uptake value, 4.5) corresponding to a 1-cm nodule in the right middle ear (Figure 1). 68Ga-DOTANOC-PET/CT did not detect other areas of pathological increased radiopharmaceutical uptake.

Based on this PET/CT finding, the patient underwent surgery. Histology showed the presence of a well-differentiated NET (Figure 2). After surgery, a normalization of chromogranin-A serum levels was observed, and the patient was judged as free from disease.

The most frequent sites of NETs are the gastrointestinal tract and the lungs; less often, patients develop NETs in the head and neck region. One particularly uncommon site of origin of NETs in the head and neck area is the middle ear. NETs of the middle ear are usually benign entities; nevertheless, in a small number of cases they may metastasize to regional lymph nodes. The most frequent symptom of the NETs of the middle ear is hearing loss (1–3).

A recent meta-analysis demonstrated that somatostatin receptor PET/CT is an accurate diagnostic method in patients with thoracic and gastroenteropancreatic NETs (4). This functional imaging method may also be applied for the evaluation of NETs of uncommon origin (5). Our case demonstrated that 68Ga-DOTANOC-PET/CT has been very useful in the correct staging of a rare case of NET of the middle ear.

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References


* G.T. and R.B. equally contributed to this paper, sharing the first authorship. Abbreviations: CT, computed tomography; NET, neuroendocrine tumor; PET, positron emission tomography.
Figure 1. Maximum-intensity projection $^{68}$Ga-DOTANOC-PET image (A), PET (B), CT (C), and PET/CT (D) images in different projections and 3-dimensional PET/CT reconstruction of head and neck region (E) showing a focal area of uptake corresponding to a 1-cm nodule in the right middle ear (yellow arrows). Physiological uptake in the pituitary was evident (white arrows).

Figure 2. Histological examination of the nodule in the right ear revealed tumor cells forming gland-like and cribriform structures without relevant atypia or mitotic activity at hematoxylin and eosin staining (A). On immunohistochemical staining, tumor cells were positive for cytokeratins (B), chromogranin (C), and synaptophysin (D). The final diagnosis was well-differentiated NET.