Neuroendocrine tumor in the mandible: a case report with imaging and histopathologic findings

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Neuroendocrine tumors (NETs) arise from neuroendocrine cells and are mostly observed in the gastrointestinal tract, pancreas, and lungs. NETs in the oral and maxillofacial region are extremely rare. We report a case of a 59-year-old woman with an NET in the mandible. The patient did not show any symptoms except for remarkable swelling and bleeding. The lesion appeared as a radiolucent honeycomb abnormality with bone destruction on panoramic radiography. The histopathologic diagnosis following a biopsy was NET. Contrast-enhanced computed tomography (CT), ^{18}F-fluorodeoxyglucose positron emission computed tomography (^{18}F-FDG PET/CT), and adrenal scintigraphy-labeled meta-iodobenylguanidine were the modalities added to identify the primary site. Multiple lesions were confirmed in the gastrointestinal tract. Endoscopy was performed to identify the lesions, and several lesions were observed protruding from the mucous membranes. However, the endoscopy specimens did not yield an accurate diagnosis because adequate samples were not acquired. Blood and urine tests revealed no functional activity caused by the tumors. Although the origin was not histopathologically confirmed with endoscopy, this patient was situationally diagnosed with nonfunctional NET originating from the duodenum, as demonstrated by the metastases in the mandible. (Oral Surg Oral Med Oral Pathol Oral Radiol 2015;119:e41-e48)

Neuroendocrine tumors (NETs) originate from hormone-producing cells. Neuroendocrine cells are found throughout the body in such organs as the gastrointestinal tract, pancreas, and lungs. Well-differentiated NET was previously described as "carcinoid" until this labeling was clarified by the World Health Organization (WHO) classification. In 2010, the WHO indicated a new classification of NETs based on both the mitotic count and Ki67 index and introduced a grading system (Grade 1 to Grade 3). The European Neuroendocrine Tumor Society (ENETS) also proposed a grading system (G1, G2, and G3).[1-3] According to their proliferative activity, G1 and G2 neuroendocrine tumors are well differentiated, and G3 tumors are poorly differentiated and are called carcinomas (NECs). The diagnosis of a NET is based on the histopathology of tumor specimens, circulating biomarkers, and imaging.[2]

Most NETs are located in the gastrointestinal tract and the pancreas. The incidence has been estimated to range from 1 to 2 per 100,000 people in Western countries.[4,5] In Japan, the latest report on the status of gastroenteropancreatic NETs (GEP-NETs) in 2005 estimated their prevalence to be 3.45 in 100,000 persons, with an annual onset incidence of 2.10 in

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Statement of Clinical Relevance

Neuroendocrine tumors (NETs) are extremely rare and arise from the secretory cells of diffuse neuroendocrine cells. These tumors are particularly rare in the oral and maxillofacial region, and only a few cases have been reported. NETs are classified as malignant tumors because of their metastatic character. However, the malignant features are difficult to diagnose from the imaging findings in well-differentiated NETs because the tumors grow very slowly and the patients lack severe clinical symptoms. In this article, we report a case of NET (Grade 2 in the World Health Organization 2010 classification) in the mandible.

Fig. 1. Intraoral photography revealing an easily bleeding mass in the left buccal mucosa.

Fig. 2. Panoramic radiograph showing a large, ill-defined, soap bubble—like radiolucency of the left mandible extending from the retromolar area to the ramus.

100,000.[6] The increase in the incidence of GEP-NETs during the last 30 years can be attributed to the increase in the detection rate due to advances in endoscopic and imaging methods.[7-9]

This report describes an NET in the mandible and includes imaging and pathologic findings, symptoms, and the process leading up to diagnosis. In this case, despite having extensive bone swelling and resorption, the patient did not have numb chin syndrome or trismus. Hence, this lesion was difficult to diagnose as a malignancy. NETs in the head and neck region, particularly the well-differentiated types, are often underdiagnosed.[10]

CASE REPORT

A 58-year-old woman visited a dental practitioner for a new denture and was taken for a dental impression. After 10 days, she visited the dentist again because a bleeding mass had appeared in the left buccal mucosa. She was referred to a hospital for further examination.

Her left cheek was swollen, and a 20 × 15 × 15 mm reddish brown bleeding mass (Figure 1) was observed on the left buccal mucosa. Trismus, paresthesia, and spontaneous pain were absent, and her lymph nodes were not palpable. She

Fig. 3. Contrast-enhanced CT showing a very large osteolytic lesion in the left ramus. **A**, axial image. **B**, coronal image.

had a history of hypertension and hyperthyroidism. Her father had died at 83 years of age from pancreatic cancer, and her sister had died of colorectal cancer.

Panoramic radiography revealed an ill-circumscribed, multilocular, radiolucent area in a honeycomb pattern around the left ramus of the mandible, which was bulging and displaying a thin cortical bone (Figure 2). Contrast-enhanced CT demonstrated a weakly enhanced lesion, which had a relatively clear boundary, around the left ramus. Moreover, the lesion was accompanied by sporadic bone destruction enlarged in the lingual and buccal directions, along with compressive bone resorption at the posterior part of the maxilla (Figure 3).

The patient had undergone magnetic resonance imaging (MRI) for torticollis 3 years ago, and a mass had been confirmed in the left mandibular ramus at that time. The lesion was observed as a round, homogeneous, well-demarcated mass on the T1-weighted image. Furthermore, the lesion showed iso-signal intensity to the parotid gland on a T2-weighted image (Figure 4, A and B).