Clinical Case Report

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Esophageal neuroendocrine tumor with initial presentation as painless forehead and neck masses

A case report

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Abstract

Rationale: Esophageal neuroendocrine tumors (NETs) are a rare type of esophageal tumor which are usually positive for chromogranin A, synaptophysin, and CD56 in tumor immunohistochemical staining. The most common symptoms of esophageal NETs are gastrointestinal symptoms such as dysphagia and/or abdominal discomfort. While esophageal NETs have the potential for distant metastasis, there have only been a few reports of brain metastasis originating from esophageal NET.

Patient concerns: We report the case of a 60-year-old Taiwanese female who initially presented with a 1 month history of painless forehead and bilateral neck masses. She did not complain of any other symptoms, which complicated the diagnosis.

Diagnoses: Chest and abdominal computed tomography were arranged for a thorough evaluation, and a paraesophageal lesion as well as multiple metastases in the liver, bilateral adrenal glands, and bone were found. Panendoscopy and pathology confirmed the diagnosis of an esophageal NET.

Interventions: Best supportive care.

Outcomes: The clinical course of this case deteriorated drastically, and she died of tumor progression 10 days after the diagnosis had been made.

Lessons: To the best of our knowledge, this is the first article in the literature to report a case of esophageal NET whose initial presentation was painless forehead and bilateral neck masses. Clinicians should be aware of the early signs and symptoms of esophageal NET to allow for a prompt diagnosis.

Abbreviations: CT = computed tomography, NET = neuroendocrine tumor.

Keywords: esophageal neuroendocrine tumor, esophageal tumor, forehead mass, neuroendocrine tumor

1. Introduction

Esophageal neuroendocrine tumors (NETs) are a rare type of esophageal tumor which are usually positive for chromogranin A, synaptophysin, and CD56 in tumor immunohistochemical staining. The reported incidence and prevalence of NETs have increased since the 1970s due to improved diagnostic techniques.[1] However, esophageal NETs are still rare, comprising only 1.4% of all gastroenteropancreatic NETs[2] and 0.15% to 2.80% of esophageal carcinomas.[3] The most commonly reported symptoms of esophageal NET are gastrointestinal symptoms such as dysphagia and/or abdominal discomfort.[2] While esophageal NETs have the potential for distant metastasis, there have only been a few reports of brain metastasis originating from an esophageal NET.[4–6] In this article, we report the first case of an esophageal NET whose initial presentation was painless forehead and bilateral neck masses, without any gastrointestinal symptoms.

2. Case report

A 60-year-old Taiwanese female who lived in a nursing home presented to the outpatient clinic of the otorhinolaryngology department of our hospital due to a 1 month history of painless swelling over her forehead and bilateral neck. She had a past medical history of a cardiovascular accident. She had lost a substantial amount of body weight, and a physical examination revealed forehead and bilateral neck masses. The results of nasopharyngolaryngoscopy were unremarkable; however, computed tomography (CT) showed a huge osteolytic mass over the frontal bone that also protruded into the skin and brain with multiple brain lesions (Fig. 1A), and bilateral neck lymphadenopathy (Fig. 1B). She was then scheduled for admission 2 weeks later for further survey and management.

However, 1 week later she was admitted to our emergency department due to fever (38.7°C), hypotension (96/60 mm Hg), tachycardia (pulse rate: 150 beats/min), tachypnea (36 breaths/min), and drowsiness for the past 2 days. A blood test showed

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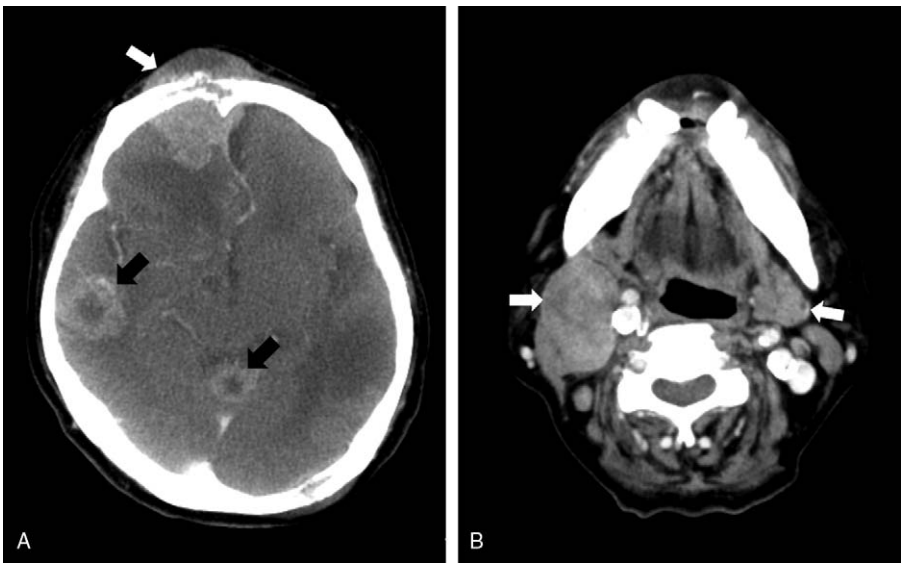

Figure 1. (A) Head computed tomography revealed an osteolytic mass lesion over the frontal bone protruding into the skin and brain (white arrow), with multiple brain metastases (black arrows). (B) Neck computed tomography revealed bilateral neck lymphadenopathy at the right IB-II level and left III level (white arrows).

leukocytosis (10390 $mm^3/\mu L$) and anemia (hemoglobin: 7.6 mg/dL). We arranged chest and abdominal CT for a thorough evaluation, which revealed a paraesophageal lesion and multiple metastases (liver, bilateral adrenal glands, bone, and neck lymphadenopathy) (Fig. 2A–C). She had high levels of tumor markers including beta-HCG, CEA, CA-125, CA-199, and CA-153. Panendoscopy revealed a 7-cm ulcerated tumor in the lower third of her esophagus (Fig. 2D). The pathologist diagnosed an esophageal NET based on positive staining for CD56, synaptophysin, and chromogranin A (Fig. 3). We then referred the patient to the hospice unit of our hospital to receive palliative care due to multiple metastases and poor clinical condition, and she died of tumor progression 10 days after the diagnosis.

3. Discussion

Esophageal tumors have the potential for distant metastases, including the brain. Song et al[7] reported an incidence of brain

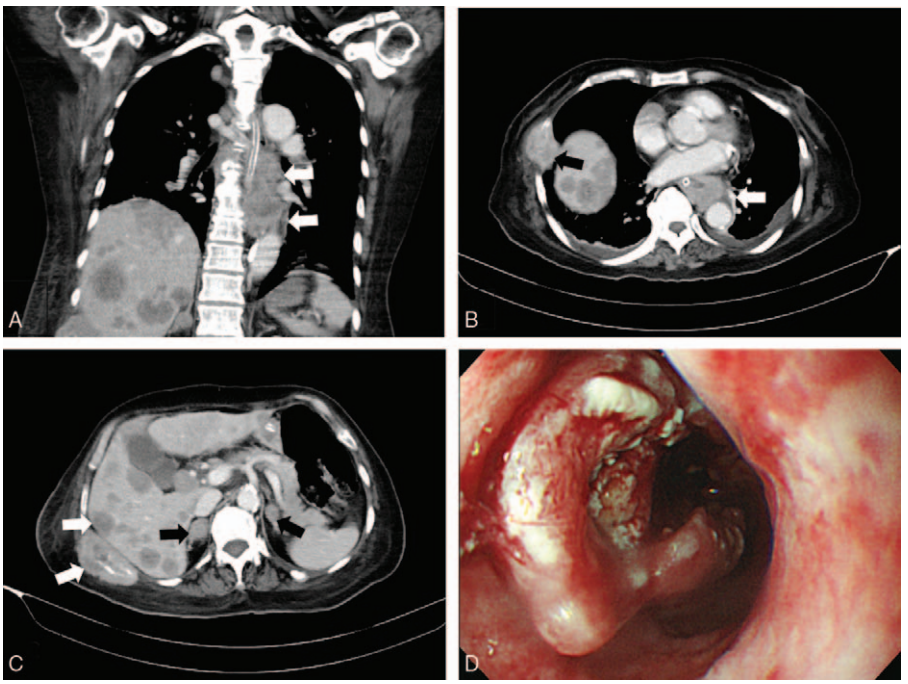

Figure 2. (A) Chest computed tomography in the coronal view showed that the length of the esophagus mass was about 7.9 cm (white arrows). (B) Chest computed tomography in the axial view revealed that the esophagus tumor had invaded the descending aorta (white arrow), and metastasis over the right 4th rib (black arrow). (C) Abdominal computed tomography revealed metastases over bilateral adrenal glands (black arrows), right 9th rib and multiple metastases of the liver (white arrows). (D) Panendoscopy showed a 7-cm ulcerated lesion in the lower third of the esophagus.

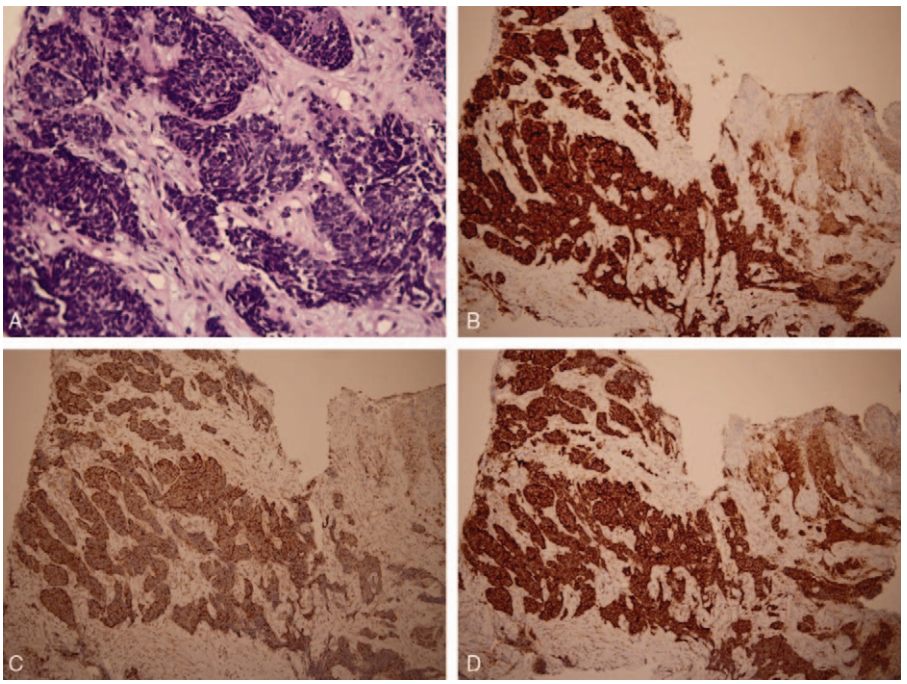

Figure 3. (A) Microscopic findings of the resected specimen of the tumor showed scant cytoplasm, hyperchromatic nuclei, nested, and molding arrangement under hematoxylin and eosin staining (400×). (B–D) Immunohistochemical findings revealed positive results for CD56 (neural cell adhesion molecule), chromogranin A, and synaptophysin (all 100×).

metastasis from esophageal tumors of 1.61%, however, all of the esophageal tumors in their study were either squamous cell carcinomas or adenocarcinomas, and there have only been a few reports of brain metastases originating from an esophageal NET.[4–6] To the best of our knowledge, this is the first case in the literature of an esophageal NET that initially presented with painless forehead and bilateral neck masses.

Although the most commonly reported symptoms of esophageal NETs are gastrointestinal symptoms, such as dysphagia and/or abdominal discomfort,[2] our patient had no gastrointestinal symptoms, which complicated diagnosis. As esophageal NETs are rare, there is currently no consensus as to the optimal treatment. Lee et al[2] proposed a treatment algorithm for esophageal NETs, in which patients with widespread metastases receive palliative chemotherapy mainly with etoposide and cisplatin. A tumor size >2 cm and American Joint Committee on Cancer TNM (Tumor extent; extent of spread to lymph Nodes; and Metastasis) advanced stage have been reported to be prognostic factors for overall survival of patients with esophageal NETs.[2,8] Our patient had a tumor size of 6.4 cm × 2.4 cm × 7.9 cm, with TNM stage IV, so the prognosis was poor. However, compared to reported medium survival of 27.04 months by Lee et al[2] and 22.4 months by Deng et al,[8] she expired exceptionally soon after the diagnosis (10 days), which may have been due to rapid tumor growth and extensive metastases.

4. Conclusion

We report the case of an esophageal NET with an uncommon initial presentation of painless neck and forehead masses without any gastrointestinal symptoms. The clinical course of this case deteriorated drastically. Clinicians should be aware of the early signs and symptoms of esophageal NETs to allow for a prompt diagnosis.

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