Extradural paraganglioma of the cervical spine is rarely seen. Few reports of this functioning disorder appear in the literature. A 29-year-old woman presented with a 1-year history of left shoulder pain and occasional transitional dizziness. This is the first case of a functioning cervical spinal paraganglioma with symptoms of catecholamine excess throughout the operation. A mass in the neck region was discovered by the patient 2 months prior to hospital admission. Physical examination revealed an egg-shaped soft and unflexible mass with no clear boundary in the anterior left part of the neck. Neither sensory disturbance nor motor weakness was evident in the upper and lower extremities. Laboratory studies were normal. Both computed tomography and magnetic resonance imaging implied neurilemmoma. Embolization of the branches supplying the mass was taken to reduce perioperative bleeding complications 1 day preoperatively. The patient demonstrated frequent hypertensive spikes with tumor handling. The blood pressure changed between 80/40 mm Hg and 200/105 mm Hg throughout the surgery. The tumor was dissected successfully from the paraspinal tissues, followed by spinal cord decompression of C4 to C6, C5 corpectomy, anterior column reconstruction, and anterior internal fixation with a plate. The histopathological examination yielded a postoperative diagnosis of paraganglioma. Diagnosis and treatment of this rare case require comprehensive perioperative management and meticulous surgical excision.

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Figure: CT images showing an epidural soft tissue mass located between the trachea and cervical spine including C2 to C7. The big paravertebral tumor was connected with the little intracranial tumor on the left side. The left transverse process and foramen of C4, C5, and C6 were destroyed by the tumor. The tumor extended into the spinal canal and compressed the spinal cord (A, B).
Parangliomas are benign tumors arising from the heterotopic sympathetic ganglion. They occur more often in the carotid body, glomus jugulare, mediastinum, and para-aortic region, unlike adrenal pheochromocytomas, and are rarely functional. Spinal parangliomas are less common and usually located in the intradural, extramedullary compartment at the level of the conus medullaris and the filum terminale. Extradural parangliomas of the cervical spine are extremely rare. This article describes the case of a functioning paranglioma of the cervical spine in a woman who presented with symptoms of catecholamine excess throughout the operation. To the best of our knowledge, there have been no previous reports of this condition.

Case Report
A 29-year-old woman presented with a 1-year history of left shoulder pain and occasional transitional dizziness. A mass in the neck region was discovered by the patient 2 months prior to hospital admission. She had no history of headache or palpitations.

The patient was afebrile and normotensive with a regular pulse and was in no obvious respiratory distress. Physical examination revealed an egg-shaped soft and unflexible mass with no clear boundary in the anterior left part of the neck. Neither sensory disturbance nor motor weakness was evident in the upper and lower extremities. Both Hoffman and Babinski signs were negative.

Laboratory studies included total blood count, hormone levels, sedimentation rate, C-reactive protein, electrolyte values, and the levels of catecholamines and their metabolites in plasma and urine, all of which were within normal ranges. Computed tomography (CT) scans revealed a 4×5×6-cm³ epidural soft tissue mass located between the trachea and cervical spine, including C2 to C7. The big paravertebral tumor connected with the little intracanal tumor on the left side. The left transverse process and foramen of C4, C5, and C6 were destroyed by the tumor. The tumor extended into the spinal canal and compressed the spinal cord (Figure 1). Magnetic resonance imaging (MRI) confirmed the lesions. On MRI, the lesions were found to be low-signal intensity on T1-weighted and high-signal intensity on T2-weighted images. After intravenous gadopentetate-dimeglumine administration, T1-weighted MRI demonstrated enhanced lesions located between the trachea and cervical spine. While the signal was not changed on the spinal cord, it was significantly atrophic at the medullary–cervical junction (Figure 2). Both CT and MRI implied neurilemmoma.

Embolization of the branches supplying the mass was taken 1 day preoperatively to reduce perioperative bleeding complications. A reverse L-shaped skin incision was made at the C4 to C6 level. The hypervascular tumor was dissected from the paraspinal tissues, followed by spinal cord decompression of C4 to C6, C5 corpectomy, anterior column reconstruction, and posterior internal fixation with a plate. Intraoperative findings revealed the epidural tumor invading the part of C5 vertebral body and causing spinal cord compression. The patient demonstrated frequent hypertensive spikes with tumor handling. The tumor compression made the blood pressure increase to 200/105 mm Hg and lasted approximately 1 minute each time. The blood pressure changed between 100/60 mm Hg and 80/40 mm Hg throughout the operation. The whole resection of accessible tumor lasted 5 hours. Hemostasis was not problematic, and blood loss was estimated at only 400 mL. However, the blood pressure decreased to 80/40 mm Hg immediately after the tumor removal. The blood pressure became stable and was maintained at 85/60 mm Hg by the anesthetist with the use of norepinephrine and dopamine at the end of the operation. Surgical specimens were submitted for pathology.

The histopathological examination yielded a diagnosis of paranglioma. Typical Zellballen nests of neoplastic cells were present. The cells possessed granular eosinophilic cytoplasm and demonstrated nuclear pleomorphism. Immunohistochemically, the tumor showed positive staining for chromogranin, S-100, and synaptophysin (Figure 3). Postoperatively, the patient recovered with no neurological symptoms. Her previous presentation disappeared. At 15-month follow-up, the patient was asymptomatic. Urinary noradrenaline levels were within the normal range. Magnetic resonance imaging scans demonstrated no obvious residual tumor.

Discussion
Parangliomas are rare neoplasms of extra-adrenal chromaffin tissue associated with the autonomic nervous system. The overall incidence of extra-adrenal parangliomas is still unknown. It is the only neuroendocrine tumor that is more common in women (M:F, 1:3) and has been seen in
almost all ages. Paragangliomas, unlike adren al pheochromocytomas, are rarely functional, and 80% to 90% arise in the glomus jugulare or carotid bodies. Paragangliomas locate less frequently in the spine and are found most commonly in the intradural extramedullary compartment at the level of the cauda equina. Extradural-spinal paragangliomas are unusual. Cases of functional spinal paragangliomas are rare, with only 4 cases reported in the literature.

Paragangliomas of the spine, due to their rarity and nonspecific clinical and imaging features, are seldom considered in the differential diagnosis preoperatively. Magnetic resonance imaging typically shows a low-signal intensity on T1-weighted images and a high-signal intensity on T2-weighted images. Gadolinium administration produced intense heterogeneous enhancement, and high-velocity flow through the hypervascular tumors produced multiple serpiginous areas of signal void with all sequences. However, the specialty of the features for paragangliomas was controversial. Others argued that the features were impossible to differentiate from the myxopapillary ependymoma, neur oroma, and meningioma in this region. Paragangliomas have distinctive histological features. Clusters of large polyhedral chief cells are arranged in nests (Zellballen) surrounded by a delicate fibrovascular stroma. However, one must not rely on the histologic finding alone. This well-known cellular arrangement can be found in the carcinoid tumors, melanomas, and medullary carcinomas of the thyroid. Microscopically, characteristic findings include granular eosinophilic cytoplasms and round or ovoid nuclei. Most commonly used for differential diagnosis, immunohistochemical staining typically confirms the neuroendocrine nature of the paragangliomas with being strongly positive for synaptophysin, neurofilament, chromogranin, and neuron-specific enolase stains. The sustentacular cells are positive for S100 protein. Malignancy cannot be determined with histological assessments but is demonstrated by metastases and invasion. Complete surgical excision is believed to be the suitable treatment of choice for both neurilemmoma and paraganglioma. To reduce perioperative bleeding complications, preoperative embolization in either neurilemmoma or paraganglioma should be considered. In addition, the relationship of the mass to the carotid artery, jugular vein, trachea, thyroid gland, and spinal cord makes surgical resection technically demanding. In previous reports, preoperative embolization has been described in cases of neurilemmoma or paraganglioma occurring in the neck. Embolization was usually carried out 1 to 7 days preoperatively with no serious complications. Perioperative bleeding and postoperative complications in these patients were significantly decreased. In the present case, preoperative embolization significantly reduced the vascularity of the cervical tumor, thereby facilitating surgical resection.

More attention should be paid to preoperative treatment of patients with functioning paragangliomas. Jeffs et al suggested at least 2 weeks of α-adrenergic receptor blockade with phenoxybenzamine to allow the chronically contracted extravascular space to expand. In this case, because of misleading preoperative imaging diagnosis and lack of presentation with symptoms of catecholamine excess, the patient did not receive the above preoperative treatment. Handling of the tumor caused a rapid increase in the pressure and heart rate intraoperatively, which implied that the tumor could be a functioning paraganglioma. The blood pressure decreased to 80/40 mm Hg after all of the accessible tumor tissues were excised. The possible reason was that expansion of the chronically contracted extravascular space resulted in the decrease in the blood pressure with not enough catecholamine secreted in plasma after excision of the tumor. Therefore, a large-volume infusion to increase blood volume should be considered pre- and intraoperatively.

Surgical removal is the gold standard treatment and must be total, possibly en bloc. Radiotherapy and chemotherapy have minor efficacy and are often used palliatively in cases of aggressive or multicentric paragangliomas. Due to the potentially aggressive nature of this disease, long-term follow-up is necessary.

REFERENCES