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Collet-Sicard Syndrome as the Presentation of Malignant Pheochromocytoma

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A 51-year-old man developed progressive dysarthria. Hours after presenting, he experienced paroxysmal diaphoresis and blood pressure fluctuations (Figure 1A). Examination showed weak cough, hoarseness, flaccid dysarthria, weak left shoulder shrug, and left tongue deviation. Our findings of lower cranial neuropathies prompted brainstem and neck imaging, revealing contrast-enhancing mass in the jugular foramen (Figure 1B). His dysautonomia compelled neuroendocrine studies, which showed elevated chromagranin A (7047 ng/mL; normal <93), epinephrine (3334 μg; normal <21), and norepinephrine (1117 μg; normal 15-80). Abdominal imaging indicated a large retroperitoneal mass (Figure 1C), confirmed as a pheochromocytoma (pheo) on biopsy.

Malignant pheo is rare, accounting for 8% to 13% of catecholamine-secreting tumors; this is classically described as the "10% rule."[1] There are sparse reports of neuraxis involvement.[2,3] Diagnostic challenges arise, given that hyperadrenergic spells (ie, diaphoresis, tachycardia, hypertension) are nonspecific, and abdominal tumor burden can remain asymptomatic for years—as seen in our patient.[4,5] Local tissue invasion is often the only clue.[4,6] In our case, cranial nerve 9 to 12 involvement offered localizing value to the jugular foramen (Figure 1D), eponymously known as the Collet-Sicard syndrome (CSS).[7,8] The differential diagnosis for CSS is broad, including tumor (eg, glomus jugulare, meningioma, schwannoma), trauma (vertebral fracture), inflammatory disorders (sarcoid, systemic lupus), infection (varicella zoster), and vascular phenomena (carotid aneurysm, venous thrombosis).[8,9] Here, malignant pheo represented a possible unifying diagnosis between CSS and hyperadrenergic spells. This concern prompted neuroendocrine studies. Given that ~95% of malignant pheo's are intra-abdominal in origin, elevated catecholamines should further compel abdominal imaging for both prognostication and treatment guidance.[10]

Declaration of Conflicting Interests

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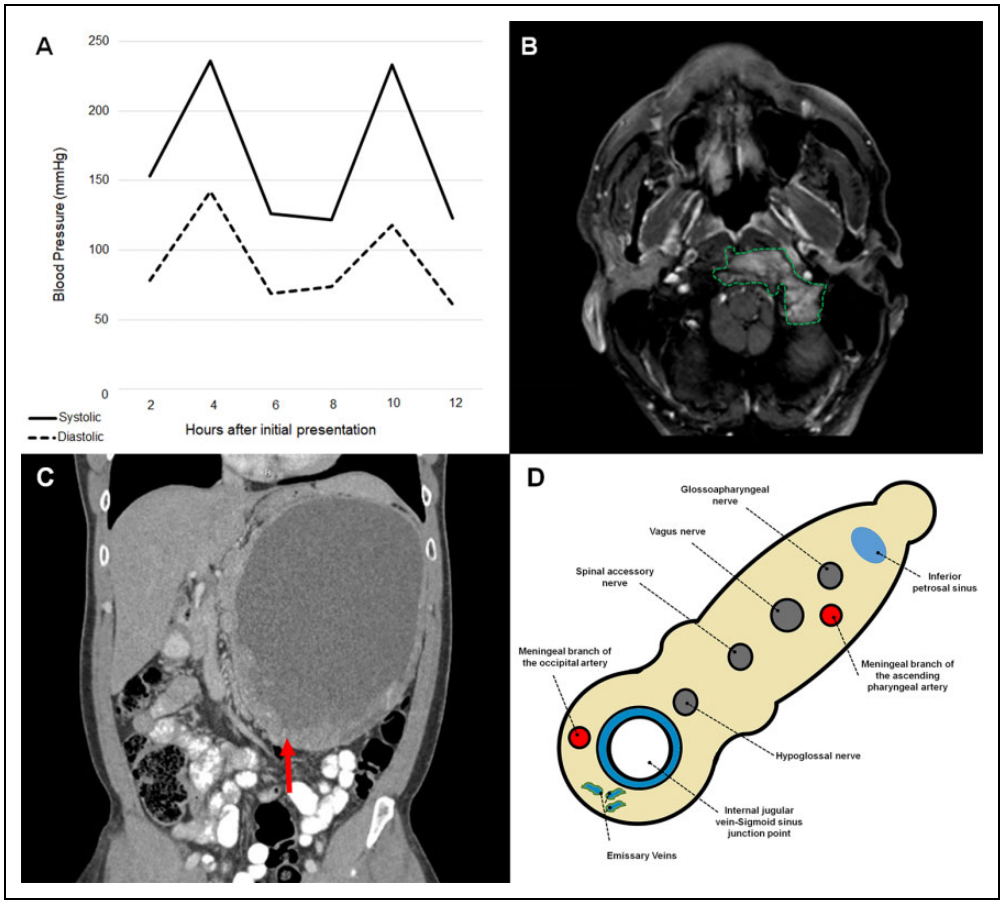

Figure 1. A, Line plots of blood pressure fluctuations during the first 12 hours after initial presentation. B, Axial T1-weighted magnetic resonance image of the neck with contrast demonstrating avidly enhancing mass (outlined) within the left jugular foramen, extending medially and eroding the left aspect of the clivus. C, Coronal computed tomography of the abdomen showing large retroperitoneal soft tissue mass (arrow), likely the primary tumor, displacing loops of bowel inferiorly and medially. D, Cross-sectional schematic representation of structures passing through the jugular foramen.

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