Collet-Sicard Syndrome Attributable to Extramedullary Plasmacytoma of the Jugular Foramen

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Key words
- Collet-Sicard syndrome
- Jugular foramen
- Multiple myeloma
- Plasmacytoma
- Skull base

Abbreviations and Acronyms
CT: Computed tomography
ENT: Ear, nose, and throat
EP-MM: Extramedullary plasmacytoma—multiple myeloma
JFS: Jugular foramen syndrome
MM: Multiple myeloma
MRI: Magnetic resonance imaging
PET-CT: Positron emission tomography—computed tomography
SEP: Solitary extramedullary plasmacytoma
SPEP: Serum protein electrophoresis
UPEP: Urine protein electrophoresis

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BACKGROUND: **Collet-Sicard syndrome is a rare manifestation of skull base disease involving the jugular and hypoglossal foramina. We report the first case of Collet-Sicard attributable to extramedullary plasmacytoma—multiple myeloma (EP-MM) and the second case of EP-MM precipitating a jugular foramen syndrome (JFS)-spectrum disorder.**

CASE DESCRIPTION: **A 59-year-old woman presented with 4 months of left aural fullness and pulsatile tenderness, positional vertigo, hoarseness, and dysphagia. Examination identified left tongue weakness and nonspecific sensory abnormalities of the ear, pharynx, and throat localizing to cranial nerves IX–XII. Imaging revealed a 3.4 × 1.4 × 2.8 cm lytic lesion extending from the left jugular foramen into the posterior fossa, for which she was referred to neurosurgery and otolaryngology for consideration of resection. A second, much smaller (1.1-cm) lytic lesion in the left posterior occipital bone was incidentally discovered by the surgeon during preoperative consultation. A stereotactic biopsy of the occipital lesion was subsequently recommended, which identified plasma cell neoplasm. Serum studies and skeletal survey were consistent with MM, and a definitive pathologic diagnosis of MM with cranial EP was confirmed by bone marrow biopsy.**

CONCLUSION: **Tumors of the jugular foramen present with a diverse array of lower cranial nerve deficits, including Collet-Sicard syndrome, a rare subset of JFS-spectrum diseases. Paragangliomas are the most common jugular foramen neoplasms, followed by schwannomas and meningiomas; however, many other rare entities have been reported as masqueraders, and diligent work-up with consideration for preliminary biopsy is recommended, particularly in the presence of additional lesions, equivocal imaging findings, or cases arousing high suspicion.**

INTRODUCTION

Collet-Sicard syndrome is an uncommon clinical manifestation of skull base disease involving the jugular and hypoglossal foramina, with consequent palsies of cranial nerves IX, X, XI, and XII, along with potentially raised intracranial pressure and other sequelae of jugular venous compression.[1,2] One specific variant in a spectrum of clinical syndromes under the umbrella of jugular foramen syndromes (JFS), Collet-Sicard, is potentially attributable to a wide range of infectious, traumatic, and other rare causes, although neoplastic processes are by far the predominant causes, most frequently a jugular paraganglioma, or glomus jugulare, or less commonly a lower cranial nerve schwannoma.[3-6] By contrast, extramedullary plasmacytoma-multiple myeloma (EP-MM) is a very uncommon intracranial neoplasm that very rarely develops within the jugular foramen and, where present, rarely presents with neurologic deficits attributable to mass effect.[7] We report the first case of Collet-Sicard attributable to EP-MM, which is also only the second such case of EP-MM neoplasm precipitating a JFS-spectrum disorder, accompanied by a review of the pertinent literature on plasma cell neoplasms involving the jugular foramen.

CASE HISTORY

A 59-year-old woman presented with a 4-month history of aural fullness, followed by positional vertigo, pulsatile tenderness of the left ear, hoarseness, and dysphagia to both solids and liquids. Before evaluation, she had experienced a single episode of isolated facial weakness, which was presumed to be Bell palsy and treated with steroids, after which baseline function was restored. The result of motor examination

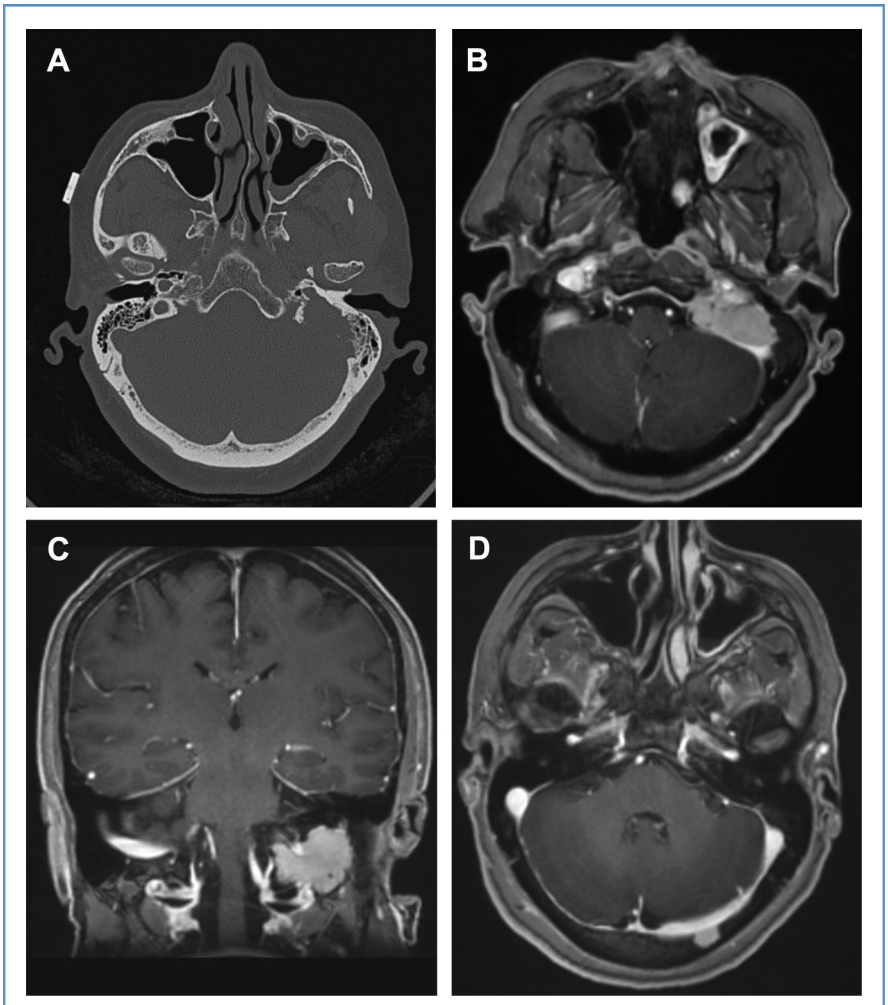

Figure 1. (**A**) Axial computed tomographic scan demonstrating destructive lytic changes extending into the left jugular tubercle and posterior left temporal bone, suggesting an underlying neoplastic process. (**B**) Axial and (**C**) coronal gadolinium-enhanced T1-weighted magnetic resonance images identifying a destructive 3.4 × 1.4 × 2.8 cm lesion of the left skull base centered within the left jugular foramen. (**D**) Additional, discrete, 1.1-cm lytic lesion separately identified in the left posterior occipital bone.

by an outside physician was remarkable for left tongue weakness, with nonspecific left-sided sensory abnormalities of the ear, pharynx, and throat localizing to cranial nerves IX–XII; upper aerodigestive examination with nasopharyngoscopy was not completed. Interestingly, the extent and severity of reported cranial nerve IX–XII symptoms improved markedly after steroid initiation, and therefore before her first examination at our institution. A computed tomographic (CT) scan showed a destructive 3.4 × 1.4 × 2.8 cm lesion centered within the left jugular foramen, from which it expanded eccentrically with associated lytic changes (Figure 1A).

Both findings were subsequently characterized more fully by magnetic resonance imaging (MRI), which was read by radiology as most consistent with jugular paraganglioma; however, on our review, the homogenous appearance and absence of prominent flow-voids raised suspicion for other causes, including plasmacytoma and lymphoma (Figure 1B–D). Consideration was also given to endolymphatic sac tumor and meningioma, but each was thought to be highly unlikely, given the radiologic appearance on MRI and CT. Perhaps most interestingly, during our clinical consultation, we noted that a second, much smaller (1.1-cm) lytic lesion in the left posterior occipital bone was also identified on preoperative consultation. Considering the abnormal radiographic features of the jugular foramen mass, taken together with the newly discovered secondary lesion, the diagnosis of jugular paraganglioma was thought to be significantly less likely. Correspondingly, a stereotactic biopsy of the posterior occipital lesion was recommended, and the patient was taken to surgery.

Intraoperatively, left occipital skin incision was made overlying the lesion, and the tumor was exposed using careful blunt dissection to preserve an adequate specimen. The lesion grossly appeared to be a nonspecific soft tissue neoplasm with mottled discoloration, suggestive of hematologic origin, and intraoperative pathologic examination suggested a plasma cell disorder. The final pathologic examination confirmed a dense, homogenous, κ light chain-restricted plasma cells neoplasm with positive CD138 expression, rare scattered CD3 positive reactive T cells, and no CD20-positive cells (Figure 2). Serum protein electrophoresis (SPEP) demonstrated a monoclonal (M) protein spike in the γ fraction—confirmed by serum immunofixation—and skeletal survey PET-CT identified multiple lytic skeletal lesions demonstrated avid uptake of ^{18}F-fluorodeoxyglucose, consistent with myeloma. Definitive pathologic confirmation was carried out by bone marrow biopsy, which confirmed cranial EP-MM. Combination therapy of bortezomib, lenalidomide, and dexamethasone was recommended, to be followed by autologous stem cell transplantation. The patient elected to pursue chemotherapy near her permanent residence.

DISCUSSION

Extramedullary plasmacytoma—multiple myeloma neoplasms of the jugular foramen are extremely rare lesions, with our patient marking the second case to present with a clinical JFS-spectrum disorder, and the first instance of EP-MM—associated Collet-Sicard syndrome.[5,8-11]

Multiple myeloma is a neoplastic proliferation of plasma cells productive of monoclonal antibodies and primarily seated within the bone marrow, where the expansile intramedullary process manifests the characteristic bony