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# JUGULAR FORAMEN EXTRAMEDULLARY PLASMACYTOMA PRESENTING AS AN EXTERNAL AUDITORY CANAL MASS WITH LOWER CRANIAL NERVE PALSIES: A CASE REPORT

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**Abstract**

**Background**: Solitary extramedullary plasmacytoma is a rare plasma cell neoplasm characterized by localized proliferation of monoclonal plasma cells outside the bone marrow. Jugular foramen involvement is particularly uncommon and often presents with multiple cranial neuropathies.

**Case Presentation:** We report a woman who presented with 3 months of progressive left occipital stabbing headache and 1 week of left otalgia, tinnitus, and hearing loss, followed by deviation of angle of mouth to the right, dysphonia, and dysphagia. Otoscopy showed a fleshy mass arising from the posterior wall of the left external auditory canal. Neurological examination revealed left sided LMN type of facial nerve palsy, reduced palatal movement with left vocal cord palsy, and left hypoglossal nerve palsy. MRI demonstrated a large avidly enhancing extra-axial lobulated lesion centered at the left jugular bulb/foramen, extending into the middle ear cavity with surrounding osseous erosion. PET-CT showed no other lesions. Serum protein electrophoresis with immunofixation and urine Bence–Jones proteins were negative. A subtotal excision was performed, achieving decompression of the lower cranial nerves. Histopathology was suggestive of an extra medullary plasmacytoma. Postoperative MRI confirmed residual disease around the jugular foramen. Adjuvant radiotherapy was initiated.

**Conclusion**: Skull base EMP should be considered in the differential diagnosis of jugular foramen masses with EAC involvement and lower cranial neuropathies. Tissue diagnosis is essential, and combined surgical decompression with adjuvant radiotherapy can provide durable local control in unresectable or residual disease.

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**Keywords**: extramedullary plasmacytoma; jugular foramen; skull base tumor; cranial nerve palsy; external auditory canal mass; radiotherapy.

## INTRODUCTION

Extramedullary plasmacytomas (EMPs) are clonal plasma-cell neoplasms arising outside the bone marrow and account for a minority of plasma-cell tumors, most commonly in the upper aerodigestive tract [1]. Skull base EMP is uncommon, and involvement of the jugular foramen is particularly rare, with only scattered reports in the modern literature [2,3]. Given the intimate relationship of the jugular foramen with cranial nerves IX–XI and the hypoglossal canal (XII), patients typically present with combinations of jugular foramen (Vernet) and Collet–Sicard syndromes, and occasionally with facial nerve (VII) dysfunction due to contiguous spread [4]. Radiologically, these lesions may mimic more prevalent entities such as jugulotympanic paraganglioma or schwannoma, especially when avidly enhancing and associated with bone erosion, underscoring the need for histopathological confirmation [2]. Current consensus statements and guidelines recommend thorough systemic evaluation to exclude multiple myeloma and favor radiotherapy (RT) as a cornerstone of local control, with surgery utilized for diagnosis, decompression, or when anatomically feasible [1].

### Case Presentation

A previously healthy woman was apparently normal until 3 months prior to presentation, when she developed insidious-onset, gradually progressive stabbing pain in the left occipital region. The pain was intermittent and not associated with nausea, vomiting, photophobia, or phonophobia. One week before presentation, she developed left ear pain of gradual onset without discharge, associated with hardness of hearing, on-and-off tinnitus, and intermittent giddiness. There was no neck rigidity. Over the same week, she noticed deviation of the angle of the mouth to the right, change in voice, and difficulty swallowing; she also reported loss of weight.

On otological examination, the right ear was normal. The left external auditory canal (EAC) showed a fleshy mass arising from the posterior wall; probing around the mass was not possible and there was no bleeding on touch. The tympanic membrane on the left was not visualized. Rinne's test was negative on the left and positive on the right; Weber test lateralized to the left; absolute bone conduction was not recordable bilaterally. There was no tragal tenderness; three-finger and fistula tests were negative. Cranial nerves I–VI were normal. There was Grade IV left facial nerve palsy with absence of forehead wrinkling, inability to close the eye, deviation of the angle of the mouth to the right, and loss of the left nasolabial fold. Soft-palate movement was reduced on the left. Tongue deviated to the left, consistent with hypoglossal nerve involvement. Videolaryngoscopy demonstrated immobility of the left false and true vocal cords with compensatory movement of the right cords and pooling of saliva in the pyriform sinus; the subglottis and trachea were normal.

MRI of the brain with contrast showed a 4.9 × 3.6 × 3.8 cm enhancing extra-axial lesion centered at the left jugular foramen involving left facial, vestibulo-cochlear, glossopharyngeal, vagus and hypoglossal nerve, extending into the middle ear with skull base erosion, vascular abutment, and non-visualization of the left internal jugular vein; CT confirmed extensive bony erosions with widening of the jugular foramen and hypoglossal canal. These features favored a jugulotympanic paraganglioma or jugular schwannoma radiologically.

A neurosurgical opinion was obtained. Whole-body PET-CT revealed no other lesions. Under endotracheal general anesthesia, the patient was positioned laterally with head fixation in a Sugita head clamp. Neuromonitoring electrodes were placed for lower cranial nerves. Through a hockey-stick incision, a far-lateral approach exposed the left suboccipu t and C1. Intraoperatively, the tumor was very vascular, firm, and extradural, eroding the lateral suboccipital region with extension anterior to the left vertebral artery. Craniotomy was extended; borders were defined; and subtotal excision was performed, leaving residual tumor within the jugular foramen. The lower cranial nerves were decompressed.

Histopathology of the excised specimen revealed dense fibrous stroma infiltrated cells of, plasmablastic morphology with open chromatin and prominent nucleoli with strong CD138 positivity, suggestive of extramedullary plasmacytoma. Bone marrow aspiration with biopsy demonstrated normal hematopoiesis without evidence of multiple myeloma. Serum protein electrophoresis with immunofixation and urine Bence–Jones protein were negative. Post-operative MRI showed a residual 2.4 × 4.1 × 2.8 cm extra-axial

lesion in the left petrous–mastoid region with heterogeneous enhancement, partial encasement of the petrous ICA, and compression of the sigmoid sinus; CT confirmed postoperative bony defects and erosions. After multidisciplinary discussion, adjuvant external-beam radiotherapy was initiated, and the patient remains under follow-up.

## DISCUSSION

This case illustrates a rare presentation of jugular foramen extramedullary plasmacytoma (EMP) masquerading radiologically as a jugulotympanic paraganglioma or schwannoma, with clinical manifestations driven by mass effect on multiple lower cranial nerves and extension into the external auditory canal (EAC). EMPs account for a minority of plasma-cell neoplasms and are classically seen in mucosal sites of the upper aerodigestive tract; skull base disease is distinctly uncommon [1]. Involvement of the jugular foramen is particularly rare but clinically important because of the tight neurovascular relationships: cranial nerves IX–XI traverse the foramen, XII lies anteromedially within its canal, and the facial–vestibulocochlear complex (VII/VIII) may be affected through contiguous extension, explaining our patient's mixed pattern of VII, IX **,** X, and XII deficits with otologic symptoms. Such patterns overlap with Vernet and Collet–Sicard syndromes and are well-documented in skull base case series and reports [4].

Differential diagnosis hinges on radiological patterns but remains imperfect without histology. Hypervascular, avidly enhancing lesions at the jugular foramen with bone erosion and middle ear extension classically suggest paraganglioma; schwannoma and meningioma are additional considerations. EMP typically appears iso- to hypointense on T1 and variable on T2 with homogeneous or heterogeneous enhancement; however, flow voids from tumor vascularity and the presence of bone erosion can blur distinctions with paraganglioma, as occurred here. Contemporary reports of jugular foramen EMP highlight this radiological ambiguity and emphasize biopsy or resection for diagnosis [2,3].

The current diagnostic framework for solitary plasmacytoma requires exclusion of systemic multiple myeloma by marrow assessment and advanced imaging (preferably whole-body low-dose CT, MRI, or PET-CT), along with demonstration of a clonal plasma-cell tumor at a single site [1,5]. Our patient's PET-CT was negative for other lesions and serum studies (including immunofixation and urine Bence–Jones proteins) were non-contributory, bone marrow aspiration and biopsy showed no features of multiple myeloma Immunohistochemistry demonstrated CD138 positivity, a reliable plasma cell marker consistent with EMP. Histologically, dense sheets of plasma cells with variable maturation and plasmablastic morphology confirm the diagnosis; immunohistochemistry for CD138 is frequently supportive **as seen in our case**, and modern series underscore the need for expert hematopathology review [1,6].

Management is individualized, balancing anatomical constraints, neurological compromise, and the tumor's marked radiosensitivity. Evidence synthesized by expert panels and guideline bodies consistently supports definitive radiotherapy as the primary local treatment for solitary EMP, achieving high local control rates—typically 80–95%—with conventional fractionation [1,7]. The enduring question is optimal dose. Multiple retrospective series and pooled analyses suggest superior control with doses ≥45 Gy, particularly for head-and-neck EMP or tumors >3–5 cm, whereas lower doses are associated with higher local failure [8,9,10]. A recent multi-institutional analysis reported overall response rates of 87% with high-dose RT (>45 Gy) compared with 67% at ≤45 Gy, translating into longer local control [8]. Similarly, a contemporary series (median 45 Gy) demonstrated 76% local control overall, with primary EMP showing significantly better control than secondary EMP [9]. National guideline compendia (e.g., NCCN) and recent narrative syntheses continue to endorse diameter-adjusted dose selection in the ~40–50 Gy range, with higher doses considered for larger lesions or residual disease after surgery [7].

The role of surgery remains critical in selected skull base cases despite the radiosensitive nature of EMP. Surgical objectives include establishing diagnosis when imaging is equivocal, relieving mass effect on neurovascular structures, and reducing tumor volume in anatomically constrained corridors to optimize subsequent RT planning. In jugular foramen disease—where intimate relationships with the lower cranial nerves, ICA, jugular bulb, and sigmoid sinus complicate complete resection—subtotal excision for decompression followed by adjuvant RT is a pragmatic and commonly reported strategy that balances neurological recovery with durable local control [2,3]. Our patient underwent a far-lateral approach with subtotal resection and decompression of the lower cranial nerves; adjuvant RT was then initiated for the residual lesion, consistent with guidelines**.**

Oncologic outcomes for solitary EMP are favorable relative to systemic myeloma. Local control after RT exceeds 80–90% in many series; cranial neuropathies may improve partially with decompression and RT, although deficits from chronic nerve compression may persist [8–10]. A minority (roughly 10–30%) will ultimately progress to multiple myeloma, most commonly within the first 2–3 years, though risk estimates vary with site, size, persistence of monoclonal protein, and imaging findings. Contemporary institutional experiences continue to explore prognostic factors and report median delivered doses around 45 Gy, with combined-modality therapy used selectively for skull base or anatomically complex sites [6].

Follow-up and surveillance should adhere to consensus guidance: periodic clinical evaluation; serum protein electrophoresis with immunofixation and serum free light chains; and imaging of the primary site as clinically indicated. Advanced whole-body imaging (PET-CT, whole-body MRI, or low-dose CT) is recommended at baseline and considered during surveillance if symptoms or laboratory markers evolve [1,5,7].

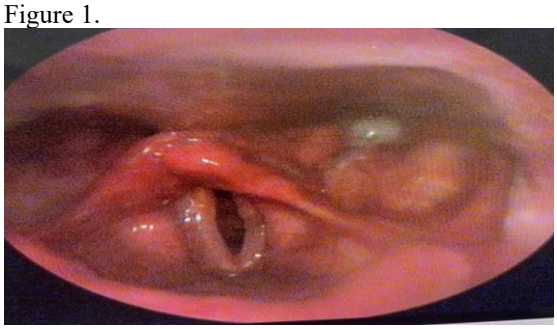
This case contributes several practical points. First, EAC masses with ipsilateral otalgia, conductive hearing loss, and tinnitus—especially when accompanied by lower cranial neuropathies—should prompt evaluation for skull base pathology, not only canal-confined disease. Second, avidly enhancing jugular foramen masses with bone erosion are not synonymous with paraganglioma; persistent diagnostic uncertainty warrants biopsy or resection. Third, in anatomically complex skull base disease, subtotal surgical decompression followed by adjuvant RT aligns with guideline-endorsed management and contemporary series demonstrating high local control for EMP. Finally, long-term hematologic surveillance is essential, given the risk of progression to myeloma even after excellent local control [1,11].

Recent literature specific to jugular foramen EMP reinforces these themes. A 2023 report described a solitary plasmacytoma of the jugular foramen closely mimicking a jugulotympanic paraganglioma radiologically, successfully managed with RT after histologic confirmation [2]. A 2024 skull base series in younger patients emphasized neurological presentations and favored RT-based local control strategies, highlighting the disease's radiosensitivity and the value of multidisciplinary care [3]. In parallel, case-based descriptions of Collet–Sicard syndromes due to skull base plasmacytoma continue to underscore the proximity of these lesions to the jugular and hypoglossal foramina and the need for tailored treatment plans [4].

## CONCLUSION

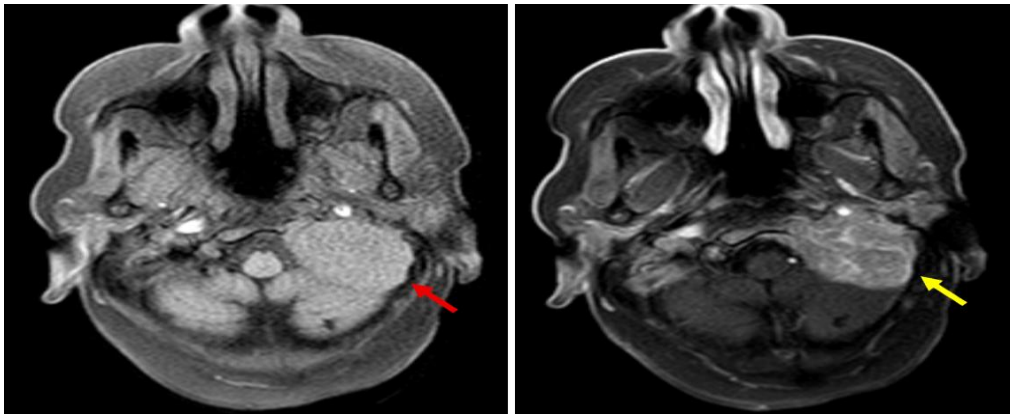
Jugular foramen extramedullary plasmacytoma is a rare skull base tumor that can masquerade as paraganglioma or schwannoma on imaging. In patients with otologic complaints and multiple lower cranial nerve palsies, clinicians should maintain a high index of suspicion and pursue tissue diagnosis. For anatomically complex disease, subtotal surgical decompression followed by adjuvant radiotherapy offers an evidence-supported path to durable local control while minimizing neurological risk. Long-term hematologic surveillance is essential to detect progression to multiple myeloma. This case underscores the importance of integrating radiological, histopathological, and systemic evaluations in establishing a definitive diagnosis. Early multidisciplinary involvement—neurosurgery, otolaryngology, hematology, and radiation oncology—optimizes outcomes. Reporting such rare presentations adds to the growing body of literature and enhances awareness of EMP as a skull base mimic, ensuring timely recognition and management in future cases.

Figure 1.

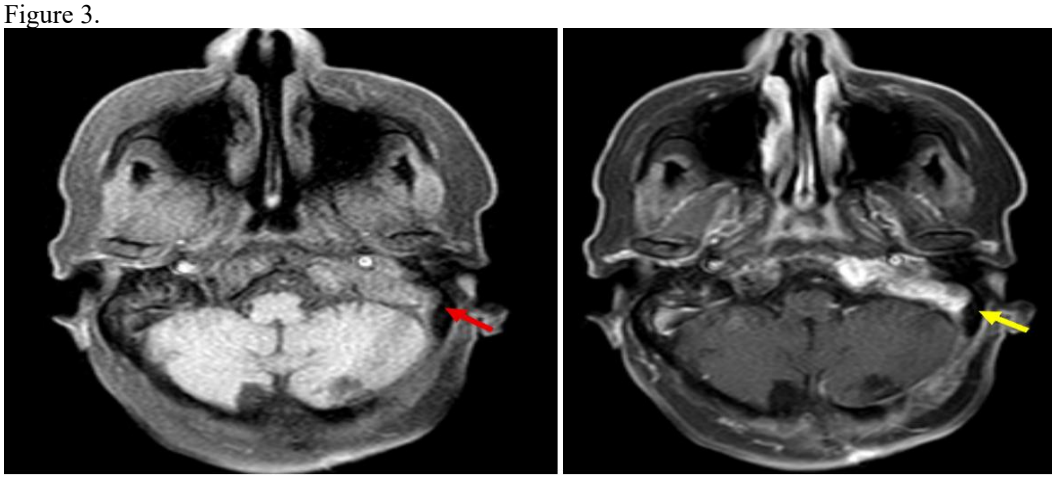


Videolaryngoscopy image showing left true and false vocal cord immobility with compensatory movement of the right cords and pooling of saliva in the pyriform sinus..

Figure 2.



MRI brain with contrast revealed a fairly defined infratentorial extra-axial lobulated lesion (Red arrow) along the left cerebellomedullary angle with epicenter at the jugular bulb/foramen, measuring approximately 4.9 × 3.6 × 3.8 cm (AP × TR × CC). The lesion was T1/T2/FLAIR isointense with foci suggestive of flow voids on T2, contained SWI hypo intensities, and showed avid homogeneous enhancement (yellow arrow) without diffusion restriction. The mass extended into the left jugular foramen and middle ear cavity; medially it abutted the left lateral cerebellomedullary cistern, flocculonodular lobe, and vertebral artery; anteriorly it reached toward the middle cranial fossa and was limited by the carotid canal; posteriorly it abutted the left cerebellar hemisphere; superiorly it involved the jugular foramen and abutted the IAC and sigmoid sinus with non-visualization of the left internal jugular vein.
Figure 3.



Postoperative MRI with contrast showed a postoperative bony defect in the left occipitotemporal region and an ill-defined infra-tentorial extra-axial lesion involving the petrous and mastoid temporal bone (residual disease – Red arrow), measuring ~2.4 × 4.1 × 2.8 cm. It was T1-isointense (Red arrow) and T2/FLAIR heterointense (predominantly hyperintense) without diffusion restriction or SWI blooming and enhanced heterogeneously (Yellow arrow). The lesion extended medially to the left lateral cerebellomedullary cistern and anteromedially to Meckel's cave; anteriorly it abutted and partially encased the petrous internal carotid artery. The left sigmoid sinus was severely compressed with suspicious invasion and non-enhancing filling defect; there was preserved but reduced flow in the left transverse sinus and internal jugular vein.

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