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Clinicopathological and Epidemiological Characteristics of Cervical Paragangliomas in Morocco: A Report of 7 Cases

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Abstract

Introduction: Cervical paragangliomas are rare neuroendocrine tumors arising from physiological paraganglia. Although typically benign, their management remains complex due to their hypervascularity and close anatomical relationships with neurovascular structures. This study aims to analyze the epidemiological, diagnostic, and therapeutic characteristics of these tumors in a Moroccan context.

Materials and Methods: We conducted a retrospective study of 7 cases of cervical paragangliomas managed between 2012 and 2023 in the ENT department of the 20 August Hospital in Casablanca. Clinical, paraclinical, therapeutic, and follow-up data were collected from medical records. Studied variables included age, sex, clinical signs, imaging results, treatment type, and postoperative complications.

Results: The mean age of patients was 50.7 years (range: 19–63 years), with a female predominance (71%). The average diagnostic delay was 32 months. All patients presented with a lateral cervical mass, pulsatile in 85% of cases. Imaging (Doppler ultrasound, CT, MRI) confirmed the diagnosis in all cases. Surgical treatment was performed in all patients, with complete resection achieved in 86% of cases. Postoperative complications included dysphonia (57%) and lingual paralysis (43%). No recurrence was observed during follow-up.

Discussion: Our findings align with literature data regarding the epidemiological profile and diagnostic challenges. The absence of preoperative embolization in our series may explain the high rate of transient neurological complications. Nerve preservation remains a major challenge, requiring meticulous surgical technique.

Conclusion: Cervical paragangliomas require multidisciplinary management in specialized centers. Improving early diagnosis and developing innovative therapeutic strategies (preoperative embolization, genetic testing) could optimize outcomes. Multicentric studies are needed to better characterize these tumors in North African populations.

Keywords: Cervical paraganglioma; Neuroendocrine tumor; Cervical surgery; Imaging

Introduction

Cervical paragangliomas are uncommon neuroendocrine tumors originating from paraganglionic tissue derived from neural crest cells. Although the majority of these lesions are benign, they are characterized by marked vascularization and a potential for local aggressiveness, which may complicate both diagnosis and treatment. Within the head and neck region, the most frequent sites include the carotid body, accounting for approximately 60–70% of cases, followed by vagal paragangliomas representing nearly 20–25%. The overall incidence of these tumors is low and is estimated to range between 1 per 30,000 and 1 per 100,000

individuals. Clinically, cervical paragangliomas often present as a slowly enlarging lateral neck mass, which may exhibit pulsatility on examination. Because of their indolent growth, patients may remain asymptomatic for long periods. However, progressive enlargement can lead to compression of adjacent neurovascular structures such as the carotid vessels and lower cranial nerves, potentially resulting in neurological manifestations. The diagnostic approach generally combines clinical assessment, biochemical testing, and imaging studies. Radiological investigations play a crucial role in establishing the diagnosis and defining tumor extension. Magnetic resonance imaging frequently demonstrates the characteristic "salt-and-pepper" appearance, whereas Doppler ultrasound and computed tomography angiography reveal the hypervascular nature of the lesion.

Surgical excision remains the primary therapeutic option, aiming to achieve complete tumor removal while preserving surrounding neurovascular structures. Nevertheless, surgical management may be technically challenging because of the tumor's vascularity and anatomical proximity to major vessels and cranial nerves. In selected cases, preoperative embolization or radiotherapy may be considered as adjunctive therapeutic options. Furthermore, a significant proportion of paragangliomas, estimated between 10% and 40%, are associated with germline mutations involving the succinate dehydrogenase (SDHx) gene family, highlighting the importance of genetic evaluation and long-term surveillance in affected patients. Despite advances in diagnostic imaging and surgical techniques, data regarding cervical paragangliomas in North African populations remain limited. The present retrospective case series therefore aims to describe the epidemiological profile, diagnostic characteristics, and therapeutic outcomes of seven patients treated for cervical paragangliomas in our institution over an eleven-year period.

Materials and Methods

A retrospective descriptive case series was conducted in the Department of Otorhinolaryngology and Head and Neck Surgery of the 20 August 1953 Hospital, part of the Ibn Rochd University Hospital Center in Casablanca, Morocco. The study covered an eleven-year period from January 2012 to December 2023. During this interval, twelve patients were initially suspected of presenting cervical paragangliomas. After applying predefined inclusion and exclusion criteria, seven patients with confirmed diagnoses were retained for analysis.

The inclusion criteria were:

- histopathological confirmation of a cervical paraganglioma (carotid or vagal origin),
- management performed entirely at our institution,
- availability of clinical records with a minimum follow-up period of one year.

Patients presenting with tympanojugular paragangliomas or incomplete clinical records were excluded from the study. Two patients who were lost during the early postoperative follow-up were also excluded from the final analysis.

Data were collected retrospectively from hospital medical records, operative reports, imaging archives (PACS), histopathological reports, and follow-up documentation. A standardized data collection form was used to record demographic characteristics, clinical presentation, diagnostic investigations, therapeutic management, and postoperative outcomes. All patients underwent a comprehensive otorhinolaryngological examination, including cranial nerve evaluation and nasofibroscopy. Imaging studies included Doppler ultrasound, computed tomography angiography, and magnetic resonance imaging depending on availability and clinical indication. Biochemical investigations included measurement of 24-hour urinary metanephrines when a secretory tumor was suspected. Surgical management was discussed during a multidisciplinary tumor board meeting involving ENT surgeons, radiologists, and anesthesiologists. Tumor excision was performed through a modified Paul-André cervical approach, using microsurgical techniques and careful subadventitial dissection of the carotid vessels.

Histopathological examination was systematically performed using hematoxylin-eosin staining, complemented by immunohistochemical analysis including chromogranin A, synaptophysin, and S-100 protein. Statistical analysis was conducted using SPSS software (version 26). Because of the limited sample size, only descriptive statistical analysis was performed. Continuous variables were expressed as mean values with ranges, while categorical variables were presented as frequencies and percentages. The study protocol was approved by the Ethics Committee of Ibn Rochd University Hospital Center, Hassan II University of Casablanca (reference CE-145/2023), and all patients provided written informed consent for participation.

Results

The demographic characteristics of our study included 7 patients with histologically confirmed cervical paragangliomas. The mean age at diagnosis was 50.7 years (range: 19-63 years), showing a bimodal distribution with peaks in young adults (19 years) and middle-aged patients (50-63 years). The series demonstrated marked female predominance, with 5 women (71.4%) versus 2 men (28.6%), yielding a sex ratio of 2.5 in favor of women. Regarding laterality, right-sided involvement predominated (71.4%, n=5) compared to left-sided (28.6%, n=2). No bilateral cases were observed. Notable medical history included one case each of asthma, treated breast cancer, hypertension, and diabetes, but no family history of paraganglioma was identified (Table 1).

The mean delay between symptom onset and specialized consultation was 32 months (range: 6-60 months). The presenting symptomatology was uniformly dominated by discovery of a lateral cervical mass (100% of cases), described as pulsatile in 85.7% (n=6), firm in all cases, mobile transversely but fixed vertically (positive Fontaine's sign) in 85.7%, and painful in only one case (14.3%). Comprehensive clinical examination revealed no cranial nerve involvement (nerves IX-XII) or Horner's syndrome at diagnosis. Systematic endoscopic evaluations (nasofibroscopy) were normal in all cases.

Table 1: *Demographic and Clinical Characteristics of Patients.*

Parameter		*Value (n=7)*
Mean Age		50.7 years (19–63)
Sex Ratio (F:M)		5:2 (71.4% female)
Diagnostic Delay		32 months (6–60)
Presenting Symptoms	Pulsatile neck mass	85.7% (6/7)
	Pain	14.3% (1/7)
Shamblin Classification	Stage I	14.3% (1/7)
	Stage III	57.1% (4/7)

Table 2: *Surgical Outcomes and Complications.*

Outcome		*Frequency*
Complete Resection		86% (6/7)
Intraoperative Events	Carotid artery injury	14.3% (1/7)
	Blood loss >500 mL	28.6% (2/7)
Postoperative Complications	Transient dysphonia	42.9% (3/7)
	Persistent dysphonia	14.3% (1/7)
Recurrence (21.8 mo FU)		0%

Table 3: *Imaging Diagnostic Accuracy.*

Imaging Modality	Key Findings	Sensitivity in Study	Literature Benchmark
Doppler Ultrasound	Hypoechoic, hypervascular	100% (5/5)	85–95% [7]
CT Angiography	"Lyre sign" (75%)	100% (4/4)	90–98% [6]
MRI	"Salt-and-pepper" (100%)	100% (3/3)	95–100% [2]

The diagnostic workup combined several imaging modalities:

- Cervical Doppler ultrasound (performed in 5 patients) revealed hypoechoic lesions, either homogeneous (60%) or heterogeneous (40%), all showing marked vascularity on color Doppler. The mean measured size was 32 mm in the longest axis.
- Contrast-enhanced cervical CT (4 patients) demonstrated well-circumscribed oval lesions with intense early arterial enhancement. Vascular relationships were precisely analyzed, showing characteristic splaying of the carotid bifurcation (lyre sign) in 75% of cases.
- MRI (3 patients) confirmed the diagnostic value of the "salt-and-pepper" appearance (100% of cases), with T2 hyperintensity and heterogeneous gadolinium enhancement. The mean MRI-measured size was 41 mm.

Our series of 7 cervical paraganglioma cases revealed notable clinical and evolutionary characteristics. Shamblin classification showed a particular distribution: 1 case (14.3%) stage I (small, minimally adherent tumor), 2 cases (28.6%) stage II (partially encasing tumor), and 4 cases (57.1%) stage III (circumferentially encasing tumor). Biochemical evaluation, including urinary catecholamines in 5 patients, was normal in all cases, excluding secretory forms. Therapeutically, all patients underwent complete surgical excision via a modified Paul-André cervical approach. The mean operative time was 185 minutes (range: 120-240 minutes) with notable technical specificities. Intraoperative challenges included one internal carotid artery injury (14.3%) and significant bleeding (>500 ml in 2 cases, 28.6%). Associated procedures included external jugular vein ligation (2 cases, 28.6%), vagus nerve sacrifice (1 case, 14.3%), and lymph node dissection (1 case, 14.3%).

Systematic histopathological analysis confirmed the diagnosis in all cases, demonstrating characteristic "Zellballen" architecture with regular chief cells lacking atypia or mitosis, and richly vascularized stroma. Immunohistochemistry showed positive staining for chromogranin A (100%), synaptophysin (100%), and

PS100 (85.7%), with no histological criteria of malignancy. Postoperative course featured early complications: dysphonia (4 cases, 57.1% - transient in 3, persistent in 1), swallowing disorders (3 cases, 42.9%, resolved with rehabilitation), and Horner's syndrome (1 case, 14.3%, partially regressive). No strokes or deaths occurred (Table 2). With mean follow-up of 21.8 months (range: 6-60 months), 5 patients (71.4%) achieved complete functional recovery, while 2 (28.6%) retained residual dysphonia. No local recurrence or metastases were detected during follow-up, although 2 patients (28.6%) were lost to follow-up after 12 months. These results confirm the oncological efficacy of surgical management while highlighting the need for a multidisciplinary approach to minimize functional sequelae.

Discussion

In our cohort, female patients represented the majority of cases (71.4%), consistent with international literature reporting sex ratios ranging from 1.9:1 to 8.3:1 across series [1-3]. This female predisposition may be explained by several mechanisms: expression of estrogen and progesterone receptors in paraganglionic cells [4,10], the stimulatory effect of sex hormones on tumor angiogenesis [4], and X chromosome-linked genetic factors [4]. The prolonged interval between symptom onset and medical consultation observed in our study illustrates the well-known difficulties associated with early detection of these tumor [5,6]. This delay stems from the tumors' slow growth rate (estimated at 0.5-2 cm/year) [2], nonspecific initial symptoms, and limited awareness of this pathology in primary care settings. The absence of secretory forms in our cohort contrasts with the 5-10% typically reported [7,8]. Several hypotheses may explain this finding: urinary metanephrine testing alone might underestimate minimally secretory forms, ethnic variations in catecholamine-synthesizing enzyme expression, and selection bias from exclusive ENT recruitment.

The predominance of Shamblin stage III tumors (57.1%) in our series warrants detailed comparison with major published series: Basel series (n=114) - 28.9% stage III [9]; Mayo Clinic series (n=153) - 25% stage III [9]. Potential explanatory factors include greater diagnostic delays, tumor biological particularities, and variable classification criteria across centers. In our experience, surgical resection alone provided satisfactory results, comparable to outcomes reported in studies where preoperative embolization was systematically performed [6]. This observation challenges the dogma of systematic embolization, particularly for small tumors (<3 cm), centers with vascular surgical expertise, and cases where embolization might pose neurological risks [10-13]. Postoperative neurological complications (57.1%) highlight several key issues: the complex anatomy of the carotid region, nerve preservation challenges in advanced stages, and the need for refined microsurgical techniques (Table 3). Methodological limitations include the small sample size (n=7) despite 11 years of recruitment, lack of systematic genetic testing, and relatively short mean follow-up (21.8 months).

Future research directions should focus on three main areas:

1. **Molecular diagnostics:** Implementation of systematic SDHx gene mutation screening (SDHB, SDHC, SDHD), given their established prognostic value in neuroendocrine tumors and paragangliomas [14] particularly for SDHB-related malignancy risk. Epigenetic studies, including DNA methylation profiling, may reveal novel tumorigenic mechanisms. Development of circulating biomarkers (microRNAs, ctDNA) could provide non-invasive tools for early diagnosis and therapeutic monitoring.
2. **Therapeutic innovations:** Robotic-assisted surgery may reduce morbidity through enhanced precision in complex anatomical areas. Anti-angiogenic targeted therapies (VEGF inhibitors) as well as peptide receptor radionuclide therapy (PRRT) have shown promising results in advanced neuroendocrine tumors and may represent potential options for unresectable paragangliomas [13,15]. For locally advanced/inoperable cases, extracranial stereotactic body radiotherapy (SBRT) may offer effective alternatives with favorable toxicity profiles [16].
3. **Healthcare organization:** Centralized management in expert centers with comprehensive technical resources and multidisciplinary teams would optimize outcomes. Establishing national reference networks with prospective registries would facilitate standardized data collection and clinical research. Development of regionally adapted diagnostic/therapeutic protocols would ensure equitable care access while enabling international comparisons.

These combined approaches could significantly improve prognosis and quality of life for patients with these rare tumors. The creation of specialized multidisciplinary teams (including ENT surgeons, endocrinologists, geneticists, and radiation oncologists) appears crucial for optimal management. Furthermore, patient education programs about early warning signs and genetic counseling for at-risk families should be prioritized in public health strategies. Long-term multicenter studies with extended follow-up periods will be essential to validate these proposed management strategies and better understand the natural history of cervical paragangliomas in North African populations.

Conclusion

Our study provides precise data on the management of cervical paragangliomas in a Moroccan context, highlighting three essential aspects. First, the need to improve early diagnosis appears crucial, given the mean delay of 32 months observed in our series, often related to limited awareness of this pathology

and nonspecific initial symptoms. Second, our results confirm the importance of specialized surgical management by experienced teams to minimize neurological complications while ensuring complete tumor resection. Finally, emerging innovative therapies, particularly preoperative embolization and targeted treatments for genetic forms, offer promising prospects. These findings justify the implementation of North African multicenter studies to better understand the epidemiological, clinical and therapeutic particularities of these rare tumors in our regional context, thereby optimizing their comprehensive management.

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