

CASE REPORT

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Bilateral carotid paraganglioma: A case report

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ABSTRACT

Introduction: Paragangliomas are rare tumors derived from the neural crest. The carotid location is the most common in the neck.

Case Report: We report the case of a 17-year-old Burundian girl who presented with a bilateral laterocervical mass syndrome evolving for several years. The interrogation reveals a case of laterocervical swellings of very slow evolution without any other clinical manifestation. The diagnosis was made by nuclear magnetic resonance imaging and angiography.

Conclusion: The young age of the patient, the bilateral location of the tumors in the carotid artery, and the family history suggest a hereditary etiology.

Keywords: Age, Bilaterality, Carotid location, Paraganglioma, SDH gene

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INTRODUCTION

Paragangliomas are tumors of the autonomic nervous system that develop from the paraganglia. Paraganglia are cells derived from the neural crest. They are rare tumors. The incidence is estimated at 1/300,000 per year, paragangliomas and pheochromocytomas combined and whatever their location (Lefebvre 2014) [1]. Although the evolution of paragangliomas is very slow and uncertain, signs of severity can be present very early on. It is important to know how to look for them and to recognize them because their presence leads to specific diagnostic and therapeutic measures. Our aim is to look for these signs in this case and then compare them with those of the most recent publications.

CASE REPORT

This case concerns a 17-year-old girl, student, without any particular history, referred to the medical imaging department for exploration of two laterocervical masses.

These masses became clinically apparent about three years before the radiological exploration. The family history revealed a cervical mass syndrome in a 27-year-old brother and hypertension in a 26-year-old sister. Examination revealed a hyperpigmented spot of several centimeters on the right forearm. Cervical magnetic resonance imaging showed two well-demarcated tissue masses located at the level of the two carotid bifurcations. The two masses measured respectively: 3.5 (L) × 2.7 (W) × 2.6 (AP) cm on the right and 1.5 (L) × 1.3 (W) × 1.2 (AP) cm on the left. In T1 sequence they appeared isosignal and in T2 sequence they appeared hypersignal with intense enhancement. Punchy areas in hyposignal appear on T2 giving the "salt and pepper" appearance, suggestive of paraganglioma. Carotid angiography showed tissue masses with intense enhancement at the level of the carotid bifurcations and spreading the two carotids. The

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right tissue mass surrounded the internal carotid artery by more than 1800. On the left, the internal carotid artery was not surrounded.

DISCUSSION

This is the first case reported in Burundi. The first serious element is the rarity of the disease. We found very few published cases in Africa. The only publications we could find were from Morocco (Soumia 2021) [2], South Africa (Cassandra Bruce-Brand 2021) [3], and Egypt (Zainine Rim Besbes 2015) [4]. The early age of onset of the disease was the second most important factor in our patient's case. In a review published in 2017, Lenders states that young age and tumor size, the third sign of severity, predispose to a high risk of metastasis and recurrence [5]. The dimensions of the patient's two cervical masses are very large compared to the size of a normal carotid corpuscle, which varies between 3 and 5 mm (Figures 1 and 2). Karren (2020) estimates the potential for carotid paragangliomas to progress to malignancy to be around 10% [6]. Bilaterality is the fourth sign of severity. The fifth element we encounter in the girl is the family history. One of the brothers has a cervical mass of the same topography, painless, asymptomatic, and evolving for several years. This mass would have appeared even before that of his sister. An older sister has episodes of hypertension although she is currently only 26 years old.

The family history, the young age of the patient, the bilaterality, and the location are elements that plead in favor of a hereditary etiology (Garcia-Carbonero 2021) [7, 8]. It is currently recognized that paragangliomas/phaeochromocytomas of genetic origin account for 30–33% of cases of carotid paragangliomas, which may be bilateral in 5% of cases and multiple in 80% of cases. Familial cases are due to mutations in the subunits of the succinate dehydrogenases gene: SDHB, SDHC, and SDHD (Amr Gad 2014) [9]. The transmission is autosomal dominant, i.e., there is a 50% chance that a child will inherit the mutation from a parent. People carrying a maternal gene mutation are in the vast majority of cases healthy, whereas those who inherit the paternal gene mutation are usually ill. The penetrance of the gene transmission depends on the gene, age, and tumor location. Genetic diagnosis is very important for the screening and follow-up of patients and their relatives. Indeed, it is now recognized that the presence of a constitutional mutation in the SDHB gene of a paraganglioma carrier is an important risk factor for malignancy (38%) and poor prognosis (Amr Gad 2014) [9]. These mutated SDHB tumors are mainly located in the thoraco-abdomino-pelvic region. They can take the familial or multiple forms, but they are less frequent (22% for familial forms vs. 21% for multiple forms), in contrast to mutated SDHD paragangliomas, where familial expression of the disease and multiple forms are more frequent, respectively 59% and 67% (Guo 2015) [10].

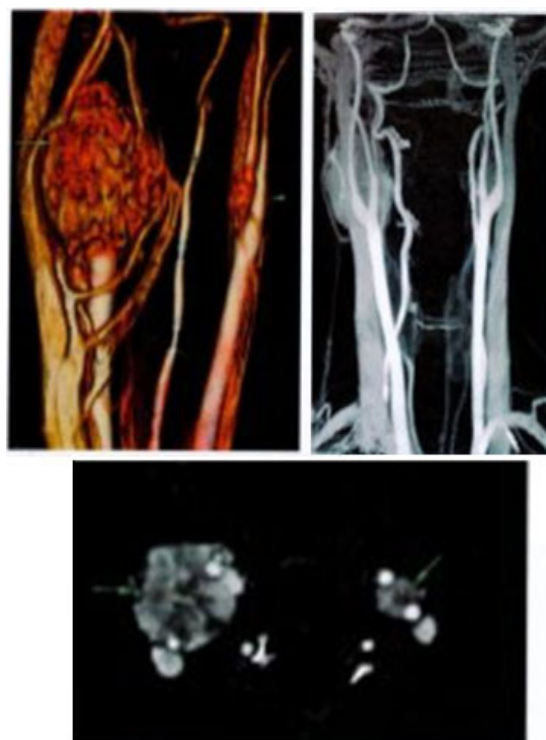


Figure 1: Carotid angiography: two tissue masses with intense enhancement at the carotid bifurcations and spreading the two carotids.

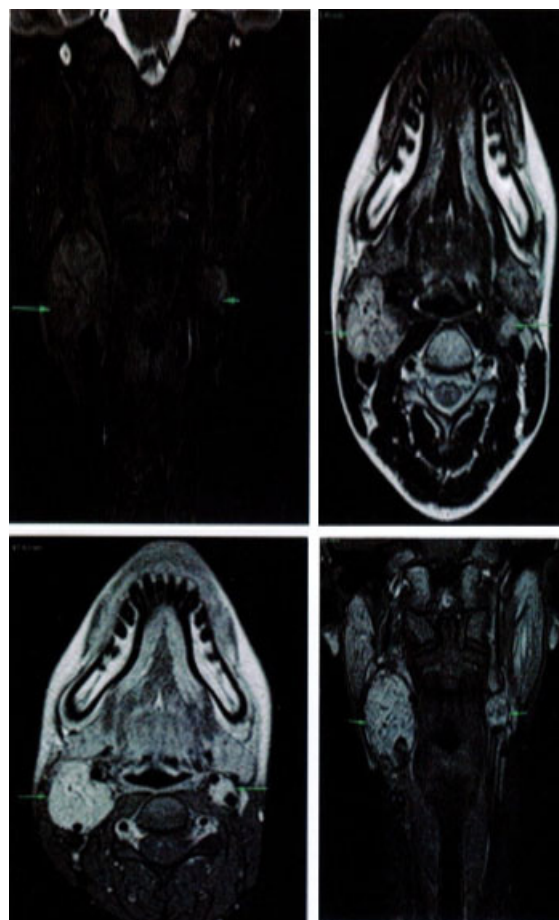


Figure 2: MRI, two well-demarcated tissue masses with a “salt and pepper” appearance.

CONCLUSION

The patient needs to complete her work-up as far as possible with genetic, biochemical, immunohistochemical, and imaging tests to ensure optimal management.

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Author Contributions

Jean Michel Nzisabira – Conception of the work, Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Eugène Ndirahisha – Analysis of data, Interpretation

of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Sébastien Manirakiza – Design of the work, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Linda Samantha Nzisabira – Design of the work, Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Guarantor of Submission

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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