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What is This?
Case Report

A Rare Case of a Second Primary Nasopharyngeal Angiofibroma

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Keywords
angiofibroma, nasal mass, endoscopic sinus surgery, nasopharyngeal mass, nasopharynx, vascular mass, nasal cavity

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Juvenile nasopharyngeal angiofibromas (JNAs) are benign tumors that mainly develop in male adolescents between the ages of 14 and 25 years. They grow in an unpredictable pattern and may involve the nasal cavity, nasopharynx, paranasal sinuses, pterygopalatine fossa, infratemporal fossa, orbit, and cranial cavity. Patients with JNA typically present with unilateral tumor masses. We present a case of a 24-year-old man with a second primary angiofibroma 5 years after removal of the primary tumor through a completely endoscopic approach. To the best of our knowledge, this is the first case of a second primary angiofibroma reported in the English literature.

Case

We obtained exemption from the Institutional Review Board at the University of Puerto Rico, School of Medicine. The patient is a 24-year-old man who was first evaluated in our clinics in September 2005 because of progressive right nasal obstruction and epistaxis for several years. Initial endoscopic evaluation revealed a large, encapsulated mass that completely occluded the right posterior nasal cavity and nasopharynx. A preoperative computed tomography (CT) scan revealed a heterogeneously enhancing mass (3.4 × 3.6 × 3.5 cm) involving the posterior right nasal cavity and extending to the nasopharynx, the right ethmoid, and the right sphenoid sinuses (Figure 1). Preoperative embolization was deferred after a 6-vessel angiogram revealed a stenosis at the takeoff of the right external carotid artery and a feeder into the tumor from the right cavernous carotid. Normal vascular anatomy was noted on the left internal and external carotid systems.

The patient was taken to the operating room in November 2005 with a working diagnosis of JNA. Resection of the angiofibroma was accomplished through a completely endoscopic approach. Postoperative pathology results confirmed the diagnosis of angiofibroma.

The patient was seen in our clinics for follow-up. Nasal endoscopy evaluations were unremarkable until 2009, when a small purplish nasal mass was found attached to the posterior edge of the left middle turbinate. A CT scan revealed a heterogeneously enhancing left nasal mass, limited to the posterior left nasal cavity, with extension to the left choana (Figure 2).

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Figure 1. Computed tomography scan of the primary angiofibroma showing a heterogeneously enhancing right nasal mass involving the posterior right nasal cavity. From here the mass extended to the nasopharynx, right ethmoid, and right sphenoid sinuses.
The decision was made to surgically resect the left nasal mass in November 2010 through a completely endoscopic resection. It was noted that the tumor mass was highly vascular, with blood supply from the left posterior nasal and sphenopalatine arteries. Pathology results again came back positive for angiofibroma. On follow-up examinations since, the patient has remained asymptomatic and without evidence of recurrence for 15 months.

Discussion

Juvenile nasopharyngeal angiofibromas are highly vascular tumors that commonly receive their blood supply from the external carotid artery system. The most common feeding vessel of these tumors is the ascending pharyngeal artery, but other vessels such as the internal maxillary artery and the descending palatine artery are also commonly involved. Feeding vessels may also come from the internal carotid artery system, as was the case with the first tumor diagnosed in our patient.

A case of bilateral angiofibromas was previously reported in the literature. In 2011, Wu et al presented a case series of 4 patients with JNA whose preoperative angiograms revealed vascular supply from bilateral external carotid systems. It has also been described that angiofibromas may recur after initial surgery. Sun et al reported that the total recurrence rate of JNA is 39.2%, with age at diagnosis <18 years old and tumor size >4 cm being the most important predictive factors for recurrence. Carrillo et al added extension pattern as a significant risk factor for recurrence, which occurred in 67% of their cases that had intracranial extension.

The case we present here differs from the ones previously discussed in that it deals with a contralateral second primary angiofibroma that appeared years after resection of the primary tumor. Before endoscopic removal of the first angiofibroma, a 6-vessel angiography confirmed that its main feeder came from the ipsilateral internal carotid artery system, with the contralateral vascular anatomy normal. With all these factors considered, we excluded the possibility of this being a case of recurrent angiofibroma, even though this was considered part of the differential diagnosis.

Conclusion

We present a case of a contralateral second primary angiofibroma diagnosed 5 years after endoscopic removal of the original primary tumor. To the best of our knowledge, this is the first case of such nature reported in the English literature.

Author Contributions

Carlos Torre-León, collected data, wrote article; José M. Busquets, revised article.

Disclosures

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