Case Report
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Giant Soft Tissue Hemangioma of the Neck with Laryngeal Extension

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Abstract

Introduction:

Soft tissue hemangiomas are among the most prevalent soft tissue tumors and can pose diagnostic challenges due to their propensity to extend into various regions. In our case, imaging studies facilitated effective characterization of the mass, and timely intervention with sclerotherapy enabled adequate initial control of the lesion. Subsequently, medical management with propranolol and a second scheduled surgical intervention with sclerotherapy contributed to a reduction in the lesion size, alleviation of symptoms, and improvement in prognosis.

Case Report:

We present a rare case of a giant cervical soft tissue hemangioma with laryngeal extension in an adult female, initially misdiagnosed as a primary laryngeal hemangioma. This case underscores the critical role of diagnostic imaging in assessing the extent of these vascular tumors.

Conclusions:

A thorough assessment of the suspected site, as well as the entire head, neck, and chest, should be conducted for all patients with suspected hemangiomas.

Keywords: Hemangioma, Hemoptysis, Magnetic Resonance Imaging, Propranolol, Sclerotherapy.

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Introduction

Hemangiomas are benign vascular tumors characterized by endothelial cell hyperplasia, frequently found in the head and neck region. Approximately 65% of all hemangiomas occur in the head and neck, with the most common sites being the parotid gland, tongue, and larynx in children (1). Laryngeal hemangiomas are generally classified into two types: infantile and adult. The infantile type is more common in females and typically localized to the subglottic area, where it primarily presents with respiratory distress, although these lesions often undergo spontaneous resolution. In contrast, adult hemangiomas are rare, more prevalent in males, and frequently located in the supraglottic area, with hoarseness as the main symptom; unlike the infantile type, they do not regress spontaneously. Other symptoms may include dysphonia, dysphagia, shortness of breath, and occasionally hemoptysis (2,3). These tumors are classified based on age of onset, origin, location, and histology (4). Histologically, hemangiomas are categorized by the types of blood vessels present. Capillary hemangiomas consist of small, mature blood vessels, while massively dilated vessels are referred to as cavernous hemangiomas. Hemangiomas with arterial structures are termed arteriovenous hemangiomas, and venous and mixed hemangiomas can also be identified (5). Most laryngeal hemangiomas are of the cavernous type, typically located in the subglottic area, and primarily cause ventilatory failure and hemoptysis (6,7).

Current literature lacks reports of laryngeal hemangiomas with extensive involvement of the base of the skull, trachea, and skin, with a radiological epicenter in the soft tissues of the neck. This article aims to report and discuss the clinical presentation and radiological findings of a patient initially diagnosed with a complicated laryngeal hemangioma, ultimately identified as a giant soft tissue hemangioma extending to the trachea and skull base. Intraoperative and imaging findings enabled precise diagnosis and appropriate interventions, emphasizing the importance of accurate identification in these rare cases.

Case Report

A 23-year-old female patient presented to the emergency department at Hospital Universitario Clínica San Rafael in Bogotá, Colombia, with massive hemoptysis, tachycardia, diaphoresis, and hypotension. Physical examination revealed non-elevated serpiginous lesions on the skin of the posterior-inferior cervical region (Figure 1).

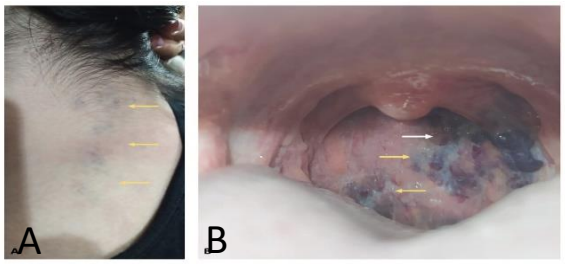

Fig 1: Findings from the initial physical examination. A) Non-elevated, non-specific serpiginous lesions in the nuchal region (yellow arrows). B) Diffuse, non-pulsatile lesions with a vascular appearance in the posterolateral and superior oropharynx. A larger lesion is observed at the left superolateral aspect of the oropharynx (white arrow), with vascular lesions extending downwards (black arrows).

Due to hemodynamic instability, fluid resuscitation was initiated. Once stability was achieved, a flexible nasolaryngoscopy performed by the otorhinolaryngology department revealed a non-pulsatile vascular lesion in the glottic and supraglottic regions.The lesion involved the piriform sinuses and partially obstructed the airway, leading to an initial diagnosis of primary laryngeal hemangioma. During her hospital stay, the patient experienced another episode of massive hemoptysis, resulting in hemodynamic instability. Consequently, she underwent an urgent micro-endoscopic laryngeal examination followed by sclerotherapy of the lesion (Figure 2).

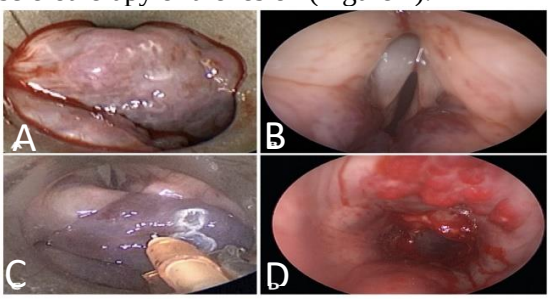

Fig 2: Intraoperative findings from laryngeal microendoscopy. A) A large supraglottic mass completely obstructing the supraglottis. B) Visualization of the glottis by laterally displacing the lesion and advancing the lens. C) View through the glottis and subglottis of multiple lesions with a vascular appearance at the tracheal level, with downward extension identified during tracheostomy. D) Intraoperative volume reduction achieved through sclerotherapy.

However, profuse bleeding occurred during sclerotherapy, posing a significant ventilatory risk; therefore, an intraoperative tracheostomy was performed to secure the airway. The surgery continued until further reduction in lesion volume was achieved, and hemostasis was established. Post-procedure, computed tomography (CT) and magnetic resonance imaging (MRI) of the neck were performed, revealing a giant soft tissue lesion extending towards the larynx. This indicated that the initially diagnosed laryngeal hemangioma was, in fact, a giant soft tissue hemangioma extending to the larynx (Figures 3 and 4).

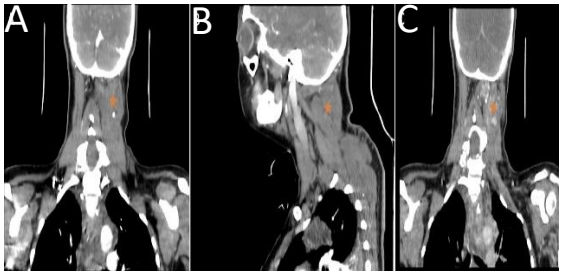

Fig 3: Contrast-enhanced CT of the neck. A and B) A poorly defined hypodense mass (orange star) displacing the mucopharyngeal and parapharyngeal spaces as well as the carotid space, extending from the mandibular angle to the suprathyroid region. C) Enhancement with contrast medium during the venous phase.

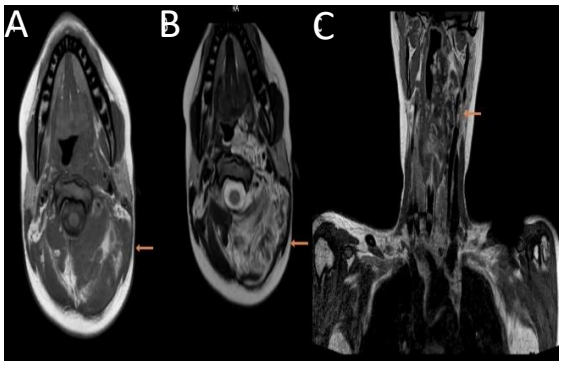

Fig 4: Contrast-enhanced cervical MRI. A) Hypointense mass (orange arrow) on T1-weighted images. B) Hyperintense mass on T2-weighted images. C) STIR T2 lesion showing intense enhancement following contrast medium administration, with evidence of involvement in the left hypopharyngeal region, extending through the scalene, parathyroid, and left paravertebral muscle planes towards the nape, surrounding large vessels, and extending towards the larynx and thyroid gland.

Given the extensive involvement and history of intraoperative bleeding, treatment with oral propranolol for six months was initiated to reduce the size of the lesion. A second intralesional sclerotherapy was then performed, resulting in significant volumetric reduction and symptom control (Figure 5). The patient continues to present violaceous lesions in the neck and oropharynx; however, there is no airway compromise.

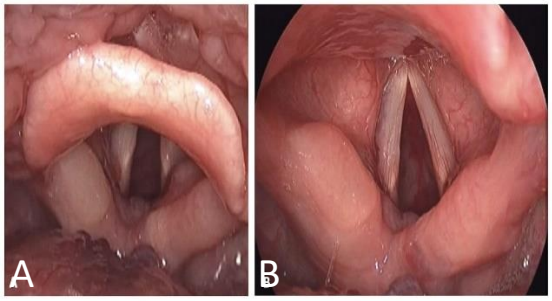

Fig 5: Posterior airway after two surgical interventions and medical therapy. Images A and B demonstrate complete regression of the lesion, with no obstruction or remnants of vascular lesions present at the supraglottic and glottic levels.

Discussion

Soft tissue hemangiomas, accounting for approximately 7% of all benign soft tissue tumors, can occur in cutaneous, subcutaneous, muscular, or synovial tissues. While superficial lesions are typically easy to diagnose without imaging, atypical and deep-seated lesions require appropriate imaging studies to differentiate them from malignant tumors and assess adjacent structure involvement, with MRI being the modality of choice. Some authors argue that hemangiomas can be diagnosed and distinguished from malignant tumors based solely on imaging features, thus negating the need for biopsy. Ultrasonography can also accurately diagnose soft tissue hemangiomas without biopsy, though ultrasound-guided biopsy is a safe option if tissue sampling is necessary (4,8).

Management of adult laryngeal hemangiomas is not well-established and depends on lesion localization, size, and symptomatic status. For larger symptomatic lesions, treatment options may include intralesional or systemic steroids, laser surgery, or a combination of both. For asymptomatic lesions, conservative management is typically recommended. Other treatment modalities for nasopharyngolaryngeal hemangiomas include embolization, radiation, sclerotherapy, and surgical excision. Some authors advocate for endoscopic surgery through

natural cavities (oral and/or nasal) for clear and wide exposure to ensure safe and successful resection (2,3). Surgical treatment of these hemangiomas carries risks, including intraoperative hemorrhage that may be difficult to control, compounded by a limited operative field. Sclerotherapy offers advantages such as simplicity and minimal invasiveness, leading to an increase in its use in clinical practice (9).

Propranolol has been shown to significantly decrease the size of cutaneous and airway hemangiomas, with few adverse effects due to the lower dosing compared to the dose used for cardiovascular treatments. However, clinicians should monitor for bradycardia, hypotension, hypoglycemia, and exacerbation of reactive airway diseases (10).

In this case, timely intervention with sclerotherapy enabled adequate initial control. Imaging studies, including CT and MRI, provided effective characterization of the lesion, demonstrating their diagnostic accuracy and utility in assessing lesion extent, which was not achievable intraoperatively. Medical management with propranolol and a subsequent surgical intervention with sclerotherapy led to significant reduction in lesion size, symptom relief, and improved prognosis. Understanding this extensive and variable condition is crucial for clinicians, as precise diagnosis affects morbidity, mortality, and clinical outcomes. This case is significant because the extent of the hemangioma caused the patient to present with massive bleeding and airway compromise, complicating the initial diagnosis and necessitating unplanned emergency management. Despite its extensive nature and associated hemorrhage, the hemangioma was managed effectively through an initial approach that included securing the airway, controlling bleeding, and reducing the volume of the lesion, followed by medical management and a second surgical procedure before tracheostomy removal. It is vital to emphasize the importance of an accurate initial diagnostic assessment, which provides insight into the lesion's extent based on observed signs and symptoms. The presence of superficial skin lesions associated with hemoptysis, respiratory distress, and stridor, as seen in this case, may indicate extension beyond the larynx, involving the surrounding soft tissues. Notably, it is essential to conduct not only routine endoscopic studies but also sequential MRI studies as an initial step to identify the lesion type and its extent. Diagnostic imaging criteria exist to recognize these lesions. On T1-weighted sequences, hemangiomas appear as masses with soft tissue density and intermediate signal, with hyperintense areas corresponding to fatty proliferation, resulting in a heterogeneous appearance. On T2-weighted sequences, the mass exhibits hyperintensity interspersed with areas of low/intermediate signal (11). Occasionally, small low-signal masses corresponding to phleboliths may be found, which are typical findings in hemangiomas. The use of contrast medium enhances the identification of highly vascularized lesions, making its administration critical for accurate diagnosis (11).

Conclusion

A comprehensive assessment of not only the suspected site but also the entire head, neck, and chest should be performed for all patients with suspected hemangiomas. If available and the patient's condition permits, complementary angiography and venography studies should be conducted. In patients with adequate hemodynamic and ventilatory stability, a stepwise approach that combines endovascular embolization with medical management should be considered, aiming to avoid surgery whenever possible.

References

1. Gauna P F, Moreno S P, Abarca S A, Cancino C M, Arregui V R. Hemangioma cavernoso laríngeo en adultos: Reporte de un caso y revisión de la literatura. Rev Otorrinolaringol Cir Cabeza Cuello. March 2018; 78(1):84-8.

2. Laohakittikul C, Srirompotong S. Adult Vocal Fold Hemangioma: A Case-Series Study and Review of Literature. J Voice Off J Voice Found. may 2023; 37(3): 471.e1-471.e5.

3. Shen Z, Zhang D, Li G, Huang D, Qiu Y, Xie C, et al. Clinical Characteristics, Classification, and Management of Adult Nasopharyngolaryngeal Hemangioma. The Laryngoscope. December 2021; 131(12):2724-8.

4. Pourbagher A, Pourbagher MA, Karan B, Ozkoc G. MRI manifestations of soft-tissue haemangiomas and accompanying reactive bone changes. Br J Radiol. December 2011; 84(1008): 1100-8.

5. Tang P, Hornicek FJ, Gebhardt MC, Cates J, Mankin HJ. Surgical treatment of hemangiomas of

soft tissue. Clin Orthop. June 1st 2002; (399): 205-10.
6. Sims HS, Cundiff J, Chejfec G. Adult supraglottic capillary vascular malformation in an African-American female patient. Respir Med Cme. December 31st de 2008;1:131-2.
7. Bridger GP, Nassar VH, Skinner HG. Hemangioma in the adult larynx. Arch Otolaryngol Chic Ill 1960. November 1970; 92(5): 493-8.
8. Rimondi E, Mavrogenis AF, Errani C, Calabrò T, Bazzocchi A, Facchini G, et al. Biopsy is not necessary for the diagnosis of soft tissue hemangiomas. Radiol Med (Torino). July 1st 2018; 123(7): 538-44.
9. Qu H, Lei X, Hu L, Yin X, Du X, Zhang L, et al. Successful Endoscopic Sclerotherapy Using Lauromacrogol Injection for Laryngopharyngeal Hemangioma. Ear Nose Throat J. November 2021; 100(9):662-6.
10. Shukla TS. Recurrence of Laryngeal Hemangioma in an Adult: A Case Report. Cureus. 14(11):e31090.
11. Marler JJ, Mulliken JB. Current management of hemangiomas and vascular malformations. Clin Plast Surg. January 2005;32(1):99-116, ix.