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CASE REPORT

Unusual presentation of an adult pedunculated hemangioma of the oropharynx

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Key Clinical Message

Hemangioma is a benign tumor rarely found in adult, especially in oropharynx. This study describes the first case of mixed hemangioma occurring as an oropharyngeal asymptomatic pedunculated mass. Biopsy is excluded given the risk of hemorrhage. Diagnosis and treatment are based on the surgical resection and the histopathologic examination.

Keywords

Hemangioma, peduncle, pharyngeal, tongue, vascular.

Introduction

Hemangiomas are the most common congenital vascular abnormality. Previous studies have reported that they are congenital with 70% present at birth as 85% are discovered during the first year of life [1] with an incidence in up to 2.5% of the neonate population [2]. The male:female ratio in children is 1:2 and mostly regresses spontaneously and disappears by 2–3 years of life [3]. They can occur at any age in adults and seem more commonly found in men [4]. It seems that 60% appear in the head and neck (i.e., skin, subcutaneous tissues, tongue, nasal mucosa, oral cavity, larynx, and salivary glands) [5, 6] even if hemangiomas occurring in the pharynx are uncommon [7]. Close observation can be sufficient, in the case of silent hemangioma, while functional compromise requires aggressive treatment [8]. In this study, an unusual case of a mixed oropharyngeal pedunculated hemangioma inserted into the base of tongue in a young female is described. The relevant literature on the clinical features, histological subgroups, diagnosis, and treatment of these lesions will be reviewed. To the best of our knowledge, pedunculated oropharyngeal hemangioma has never been documented, especially with a base of tongue origin.

Case Report

A 33-year-old woman was referred to the department of otolaryngology for progressive dysphagia, hemoptysis, intermittent dyspnea, and a globus sensation over a period of 2 months. The patient did not smoke and did not

drink alcohol, and her personal and medical history were unremarkable. The videolaryngoscopic examination of the aerodigestive tract showed a very large pedunculated red-blue tumor occupying the oropharyngeal cavity with an uncertain anatomical origin. The larynx was uninvolved with normal mobility. Computed tomography (CT) revealed a pharyngeal mass measuring 30 mm along the axis with numerous phleboliths (Fig. 1). An MRI was not performed because the appearance of the mass on the CT clearly confirmed that a resection en bloc was possible. Both clinical and radiological explorations of the head and neck area did not reveal another lesion. Based on the clinical features, with additional imaging investigations, the decision to surgically resect the tumor was made and accepted by the patient. Given the macroscopic highly vascularized appearance, no biopsy was performed owing to the risk of uncontrollable hemorrhage. Direct laryngoscopy reported a pedunculated, lobulated, and hypervascularized lesion of the pharyngeal cavity (Fig. 2). Tumor resection was performed by endoscope surgery under controlled hypotensive anesthesia. This reported a mass found to have originated in the base of the tongue with a long peduncle attached above the vallecular. No active bleeding episodes or other intraoperative complications were reported during surgery. The macroscopical examination showed that the mass measured $29 \times 16 \times 14$ mm and had a nodular, pedunculated, and lobulated appearance. Histopathologic analysis identified a mixed vascular lesion with both capillary and venous components (Fig. 3). CD31 and CD34 were strongly positive in immunohistochemistry, confirming the vascular nature of the tumor. Kaposi sarcoma was excluded (HHV-8: immunohistochemistry LANA-1). The proliferation index Ki67 was low, and at the six-month follow-up, no recurrence was observed.

Discussion

Pharyngeal hemangiomas are extremely rare, particularly in the oropharynx [9]. Including this case, the team also reviewed 12 cases of pharynx hemangioma in the literature published in English [7–17] (Table 1). Of these 12 cases, 10 were located in hypopharynx with various extensions, only one was in oropharynx, and one was unspecified. The 12 tumors were found primarily in subjects with an average age of 45 years old (33–77); 55% of them were women. The pharyngeal cases reported were rarely exophytic and occurred as a spot or flat mass on the posterior pharyngeal wall or in the pyriform sinus. This patient seems to be the first case described in literature of an adult pedunculated hemangioma of the oropharyngeal cavity. Indeed, besides some cases described in the olfactory cleft or nasal cavity [18], there was only one pedunculated case reported in the hypopharyngeal cavity [14]. Moreover, in this case, it is possible that the hemangioma was present and asymptomatic since birth, as reported by Brown et al. [19].

From a clinical point of view, these tumors are known to have unpredictable clinical behavior [20]. Patients usually complain of hoarseness, dysphagia, dyspnea, and there is a risk of bleeding and consequential complications, that is, false swallowing and hematemesis. Globus (7), dysphagia (5), bleeding (4), and voice complaints (3) were the most usual symptoms reviewed in the literature. The patient had clinical complaints of dysphagia, dyspnea, and bleeding for only 2 weeks, which was surprising given the size of the mass. However, this tumor with its thin pedicle could have been more dangerous in the case of airway obstruction. It is important to clarify that, in some cases, there may be superinfection phases leading to the development of symptomatology and the discovery of a hemangioma [17].

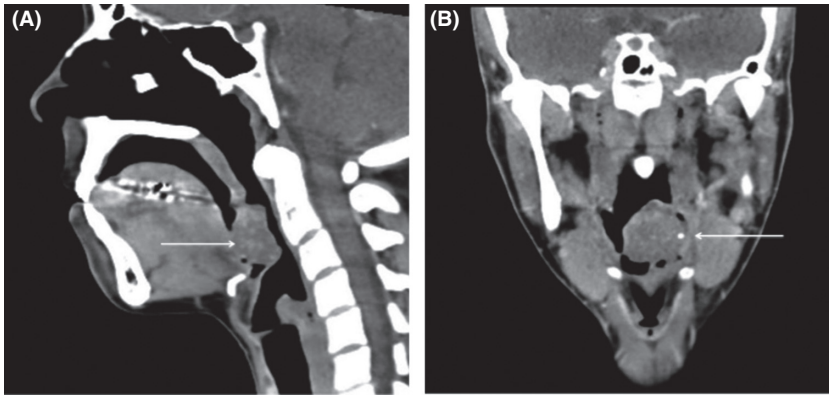

Figure 1. Radiological findings. Pre-operative computed tomography (CT) without contrast administration reported an oropharyngeal tumor (A) occupying 80% of the oropharyngeal space with numerous phleboliths (B). The peripharyngeal tissues, that is, tongue, seems free from any secondary invasion of the lesion. The pedicle is not shown in these images.

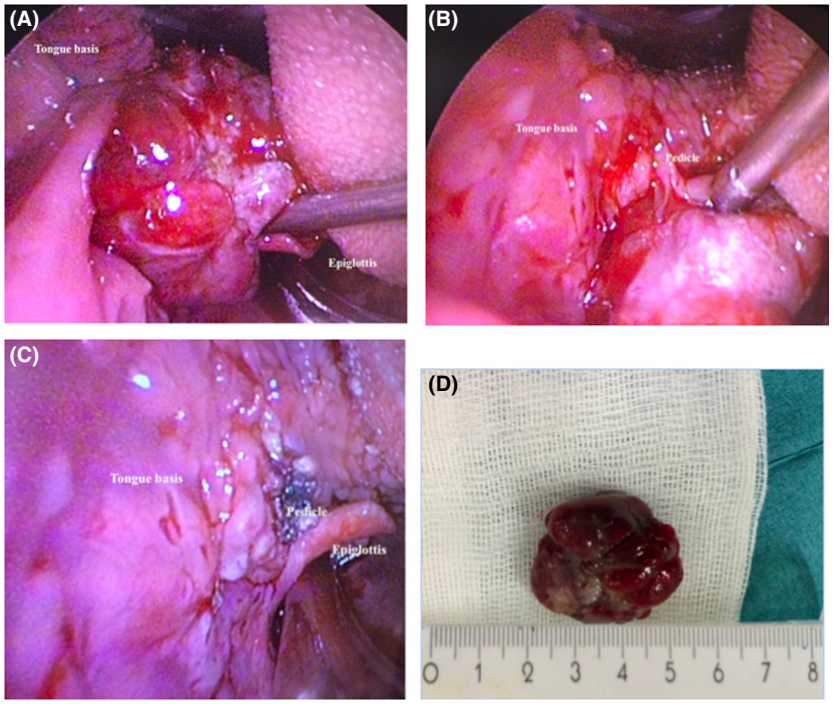

Figure 2. Clinical findings. Endoscopic examination of the upper aerodigestive tract shows a red-blue oropharyngeal lobulated tumor (A). During endoscopic surgery, it was observed that the tumor had originated at the base of tongue and was attached by a 1-cm pedicle containing blood vessels (B), which was better visualized after endoscopic resection (C, D). The pedicle was cut with bipolar coagulation forceps without significant bleeding, and the hemangioma was resected without complications.

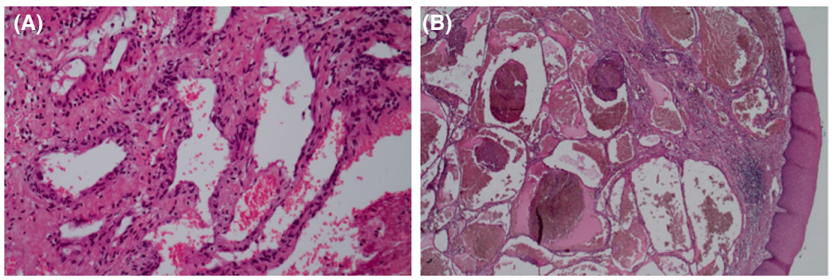

Figure 3. Histological findings. Histology showed vascular proliferation with numerous dilated vessels. Some of them were partially filled by thrombi (A). On the right, some vessels with variably muscular thick walls were identified as the venous component (B) (original magnification x65 (A) and x200(B)).

Most pharyngeal tumors, or those at the base of the tongue, can be recognized by a cross-sectional CT scan or MRI [21]. CT usually provides information about the size, the composition, and the expansion of the lesion. Although the variable vascularization characterizing hemangiomas may influence the nature of the enhancement, they are often characterized by a weak-demarcated enhancing pattern as a result of intratumoral nonenhanced thrombosis or phleboliths. When an MRI is done, hemangioma lesions appear as solid masses with isointense or slightly high signal intensity compared with muscle on T1 and with a heterogeneous signal intensity on the T2 sequence [21]. In the present case, an MRI was not performed for the reason explained in the case report description. Not surprisingly, the CT performed showed a very large pharyngeal mass with phleboliths and punctuate enhancement [21]. The pedicle was not seen with tomodensitometry.

Concerning histopathology, some authors mistakenly use the term "vascular malformations" to describe hemangiomas, whereas these two clinical entities seem to be two different disease entities with different histopathologic and clinical features, and natural courses [22]. There are several hemangioma types including capillary,

Table 1. Cases of pharyngeal hemangiomas available in the literature published in English.

References	Year	Gender	Age	Tumor extent	Tumor aspect	Histological	Clinical findings	Treatment	Complications	Follow-up	Outcome
Edwin [10]	1961	N.A.	N.A.	Hypopharynx Oral cavity	N.A.	Cavernous	N.A.	N.A.	N.A.	N.A.	N.A.
Li [11]	1990	F	32	Hypopharynx *Pyriform sinus* *Lateral wall*	Smooth swelling	Cavernous	Hoarseness Globus Dyspnea & stertor & chocking	Surgery	Slight Hoarseness	180 m	Recurrence
Kornfehl [12]	1995	M	32	Hypopharynx	Exophytic	Cavernous	Dysphagia Hoarseness	CO2/Nd laser	None	24 m	Recurrence
Yellin [13]	1996	F	39	Hypopharynx *Lat. & post. Walls* Oral cavity Thyroid lobe Mediastinum	Soft tissue mass	Cavernous	Odynophagia Dysphagia Globus	Nd-YAG LP	None	6 m	Residual hemangioma
Guo [14]	2001	M	77	Hypopharynx *Pyriform sinus*	Polypoid mass Pedunculate	Mixed Hemangioma	Dysphagia Globus	CO2 Laser	None	24 m	Recurrence Voice quality
Qureshi [9]	2004	M	65	Hypopharynx Base of tongue	Fleshy mass	Capillary	Dysphagia Bleeding	Surgery	None	36 m	Speech Swallowing Recurrence
Hazarika [15]	2006	F	36	Hypopharynx *Pyriform sinus*	Cystic mass	Cavernous	Globus	KTP Laser	None	6 m	Reducing symptoms
Hussain [3]	2011	F	44	Hypopharynx *Pyriform sinus* Right ary-epiglottic fold	Soft tissue mass	Cavernous	Bleeding Dysphonia Globus Epigastric pain Weight loss	CO2 Laser	None	3 m	Reducing pain Reducing hemoptysis Reducing dysphonia
Kozakiewicz [16]	2012	M	59	Pharynx	Soft tissue mass	N.A.	Worsening hearing in the left ear	Embolization	N.A.	N.A.	Reducing the tumor size Decreasing symptoms
Won [4]	2013	M	42	Hypopharynx Esophagus	Flat lesion	N.A.	Asymptomatic	None	None	30 m	Symptoms Signs
Reder [16]	2014	F	38	Hypopharynx	Soft tissue mass	Cavernous	Dysphagia Weight loss Globus	CO2 Laser	Postcricoid edema	2 m	Decreasing symptoms Reducing postoperative Decreasing edema
Lechien	2015	F	33	Oropharynx	Pedunculate	Mixed Hemangioma	Dysphagia Bleeding Globus Dyspnea	Surgery	None	6 m	Decreasing symptoms Recurrence

Summary of cases of adult isolated pharyngeal hemangioma reported in the literature published in English. All of these hemangiomas have a starting point in the hypopharynx. N.A., Not available; Nd-YAG LP, neodymium:yttrium-aluminum-garnet laser photocoagulation; KTP laser, potassium-titanyl-phosphate laser; F, Female; M, Male.

cavernous, and mixed tumor [23]. Capillary and cavernous hemangiomas are identified by thin-walled vessels with a flattened endothelium. The caliber of the vessels is more dilated in cavernous hemangiomas in comparison with capillary ones, where they are very small [24]. Mixed hemangiomas are characterized by small and dilated vessels, as in this case. In the literature, seven cavernous, one capillary, and two mixed hemangiomas were found (Table 1). Macroscopic examination shows that hemangiomas in adults are often characterized by a red-blue appearance [25], cystic or soft tissue mass. Large lesions can lead to ulceration, necrosis, or chronic infections of the concerned area. In this subject, the tumor had a nodular, lobulated, and atypical pedunculated appearance originating from the oropharynx, especially from base of tongue. The most common pedunculated tumors in oro- and hypopharynx remain fibrolipomas and leiomyomas, but not hemangiomas. The exact pathogenesis of hemangioma is still not understood although it would appear that gene mutations or deletions, microtrauma, and hormonal factors could be the cause of the disease.

Currently, there is some controversy regarding the optimal treatment for hemangiomas of the aerodigestive tract [25]. Indeed, they represent a therapeutic challenge depending on their location, functional impairments, size, and bleeding risk, as seen in the literature (Table 1). Endoscopic surgery with or without sclerosing drugs, cryotherapy, radiotherapy, and laser CO2 have varying degrees of effectiveness [25]. Unlike in children where hemangiomas could involute spontaneously, hemangiomas in adults persist over time and do not respond to medical treatment. In fact, the treatment choice for hemangiomas between the various options depends on the primary site, various stages of growth, if it is detected early, the depth of the lesion, and any previous treatments [26]. Main indications to aggressive treatment include accommodating symptoms and/or functional disorders, that is, bleeding, chronic troublesome complaints, airway obstruction, dysphagia, and infectious episodes [25]. The use of laser therapy or surgery excision is often recommended in accessible exophytic hemangiomas [12, 28]. Laser CO2 is a safe approach with a high success rate, greater than 77%, to treat superficial and smaller proliferating lesions, and to reduce the size of the lesion, creating a favorable situation for subsequent therapeutic approaches [26, 27]. Laser procedures are also recommended if the adult hemangioma is pedunculated and if the tumor can be excised across its base [14]. In this case, given the pedunculated aspect, an endoscopic surgical approach was the first choice to resect the tumor. Cauterization of the peduncle vessels was performed with bipolar coagulation forceps, to reduce bleeding during the operation, which led to the exeresis of the hemangioma without complications. Embolization makes sense especially when hemangiomas can lead to serious complications, especially laryngeal and pharyngeal hemorrhages, which sometimes require intensive treatment [20, 25, 29]. Nevertheless, it should be borne in mind that radiotherapy, radioactive gold grain implantation, cryosurgery, and sclerosing drugs increased the possibility of delayed complications, such as scarring and stenosis [14]. Regardless of the treatment used, there is still the risk of local recurrence which is why careful follow-up should be offered to every patient. Local recurrence and the decrease in symptoms are the two most common outcomes referred to in the literature, wherein the mean follow-up time is 35 months.

The case described here is the first reported case of a very large mixed pedunculated hemangioma of the oropharyngeal cavity in an adult subject, who had been asymptomatic for a long time. Given the pedunculated aspect, endoscopic surgery was carried out to extract the mass without massive intra-operative bleeding. The rationale to publish this case is the unusual histopathologic presentation of this hemangioma, the uncommon site of the lesion, and the atypical clinical history of the symptoms.

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Authorship

JRL: wrote the paper, analyzed the case, conducted literature review, and performed the research. LGDM: analyzed the case and wrote the part: "case report." IT: is the pathologist who provided the images relating to the case, and he also corrected a large part of the paper (clinical arguments and English language). MK and JRL: conducted the patient's surgery. MK and SS: coordinated, read, and corrected the paper, and SS: is the head of the department.

Conflict of Interest

None declared.

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