Unusual Presentation of Laryngeal Cavernous Hemangioma

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ABSTRACT
Cavernous hemangioma is a rare tumor of the adult larynx. These hemangiomas are confined to the larynx and generally asymptomatic. We present a rare case of a huge cavernous hemangioma in a 22-year-old patient who presented with stridor and a huge swelling in the neck, of acute onset. Detailed evaluation including 70° Hopkins laryngoscopy, contrast-enhanced computed tomography (CT) scan and magnetic resonance imaging (MRI) revealed a vascular malformation with both intra- and extralaryngeal components. The typical findings of hemangioma with its management are highlighted in this article. Postoperatively, patient’s voice improved and the stridor was relieved.

CASE REPORT
A 22-year-old woman presented to the emergency room with a long-standing swelling in the anterior neck region (around 5 years). The swelling had been painless and nonprogressive until 3 days back when she developed a sudden increase in the size of swelling associated with severe pain. She also developed hoarseness and difficulty in breathing, especially in the supine position. There was no history of fever or an inciting trauma to the neck. The patient did not give any history of sudden phonotrauma. On physical examination, the patient was tachypneic and had stridor. There was a 7 × 5 cm swelling over the anterior neck extending between the anterior margins of bilateral sternocleidomastoid muscles and superiorly extending up to the level of hyoid bone (Fig. 1). The swelling moved with deglutition but not on protrusion of tongue. It was soft, tender, nonpulsatile and rubbery to touch. Hopkin’s 70° laryngoscopy revealed a submucosal bluish bulge involving the right aryepiglottic fold and right false vocal fold (Fig. 2).

With the possibility of a vascular mass in mind, contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) scans of the neck was ordered. Computed tomography scan revealed heterogeneous lobulated swelling with serpiginous enhancing vessels in the neck and in the larynx, at the

Fig. 1: Huge anterior neck swelling extending between the two sternocleidomastoid heads

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level of false cord obscuring the airway (Fig. 3) and MRI scan revealed T2 hyperintense lesion and T1 hypointense multilobulated lesion 7.5 × 3.4 × 2.4 cm displacing the epiglottis, involving the thyroid cartilage and causing airway occlusion at the level of C4 (Fig. 4). The patient was posted for a combined external and transoral excision of the mass. Patient was intubated with a small sized endotracheal tube. A horizontal neck incision was taken and strap muscles were divided in midline and retracted to expose the vascular mass (Fig. 5A). Thyroid gland was found to be displaced toward left. Right recurrent laryngeal nerve was identified and preserved. The mass was separated from the thyroid and cricoid cartilage and excised (Fig. 5B) and a part of the vascular mass extending into the larynx was tackled via exposing the larynx using a Kleinsasser laryngoscope. With the bulge still present, a small incision was made on the right aryepiglottic fold using CO2 laser and vaporization of the mass was done achieving adequate hemostasis.

Patient could be successfully extubated. Postoperatively, she did not have any breathing difficulty and her voice improved considerably. Postoperative histopathology report revealed cavernous hemangioma. A follow-up Hopkin’ s 70° laryngoscopy revealed normal endolarynx at 3 months follow-up period.
DISCUSSION

Laryngeal hemangiomas are broadly classified into infantile and adult types. This classification was first proposed by Sweeter in 1921. Infantile hemangiomas are associated with multiple skin and gastrointestinal manifestations. These hemangiomas in the neonatal period can be associated with dyspnea and stridor, and they generally involute by 5 years of age.

Adult hemangiomas in contrast do not involute spontaneously. They are more commonly of the cavernous type and are mostly located at or above the level of vocal cords.

In our case, the location of the huge cavernous hemangioma was supraglottis and in addition it was also spreading extralaryngeally. The important point clinically in this case was to differentiate it from a thyroid swelling. Since the patient had a long-standing history of anterior neck selling before the acute symptoms developed, the first diagnosis would have been of a thyroid swelling. But the rubbery feel on palpation along with a bluish bulge on endoscopy, clinched the diagnosis of hemangioma of larynx, which was further confirmed by imaging. On MRI, cavernous hemangiomas appear as well-demarcated, homogeneously hyperintense lesions on T2-weighted images, and hypointense on T1-weighted images and on contrast-enhanced T1-weighted images, all of these lesions have homogenous enhancement.

Sudden onset hemorrhage in a long-standing hemangioma caused the acute symptoms in our case and tracheostomy could be avoided in this case because of the rapid and timely diagnosis and intervention.

The surgical approach was combined extra- and intralaryngeal because of the size of the hemangioma. Bridger and associates recommended that adult laryngeal hemangiomas should be left alone if there are no symptoms.

If there are obstructive symptoms (like in our case), like sudden stridor and breathing difficulty caused by rapid growth (because of the internal hemorrhage), active intervention is warranted. The treatment options of laryngeal and upper airway hemangiomas include steroid injection, systemic steroid therapy, beta blockers therapy, laser ablation and surgical removal. Radiotherapy use for hemangiomas is now obsolete, since radiation therapy has a host of side effects. Use of beta blocker like propranolol is the modality of choice in infantile hemangiomas and has been extensively studied in the recent past. CO2 laser is the treatment of choice for adult laryngeal hemangiomas. In cases of extended hemangiomas, staged procedures may be required. Lucioni et al in their 3-year study of use of CO2 Laser for hemangioma cases, found it successful in five out of six cases.

CONCLUSION

To summarize, one needs to be aware of the following facts:

- Adult laryngeal hemangiomas, although silent for many years, can rapidly grow and cause obstructive symptoms
- Bluish submucosal bulge on endoscopy clinches the diagnosis
- Imaging confirms the diagnosis
- Timely diagnosis and intervention can avoid tracheostomy
- Surgery with CO2 laser is feasible option.

REFERENCES