A RARE CASE OF ARTERIO-VENOUS MALFORMATION OF LARYNX.
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Abstract: Arterio-Venous Malformation of the Larynx is a rare vascular lesion, and a very few reports of such lesions are reported in the medical literature. We report a rare case of A-V malformation located over the left pyriform fossa of larynx in a 22-year-old female, who presented with a swelling in the left side of the neck. On videolaryngoscopy, smooth bluish swelling was seen in the left pyriform fossa. Surgical excision was performed without complications. The 2-month postoperative follow-up showed good results with no recurrence.

Keywords: Arterio-Venous Malformation, Larynx, Pyriform fossa, Vascular lesion

INTRODUCTION
Vascular anomalies are among the most common congenital and neonatal dysmorphogenesis which are separated into hemangiomas and vascular malformations. They can occur in various areas throughout the body with 60% being located in head and neck. True mechanism of the pathogenesis of vascular anomalies is still unclear.

CASE SUMMARY
22 years old female patient came with complaints of gradually progressive swelling in left side of neck since childhood. This was not associated with any difficulty in swallowing or change in voice, hematemesis/hemoptysis or respiratory distress. Clinically, swelling of about 4x4 cm present at level of thyroid cartilage on the left side extending to anterior border of sternocleidomastoid. It was soft, compressible, non-transilluminant, fluctuant, non-tender.

On General Examination
- Moderately built and nourished, alert, conscious and oriented
- Not Anemic/Jaundiced
- No cyanosis/clubbing
- Temp: 98F
- PR: 78/min
- BP: 110/70 mmHg
- Respiratory Rate: 16/min
- Cardiovascular system - S1 S2 heard, No Murmur
- Respiratory System - Normal Vesicular breath sounds, No Added sounds
- Per Abdomen - soft, No Organomegaly
- Central Nervous system - Higher Mental Function - Normal, No Focal Neurological deficit
- Skin - Normal

Videolaryngoscopy examination smooth swelling present in left Pyriform fossa which was bluish in colour.

USG finding revealed highly vascular lesion, less likely to be a nerve sheath tumour.

CT neck with contrast shows multi-compartmental soft tissue lesion with contrast enhancement in left pyriform fossa, paraglottic space. ?Hemangioma ?AV malformation.
The key to embolization is to use sufficient embolization to control the growth of AVMs and frequent bleeding. Angiography and embolization should be considered as the purpose of treatment. If the embolization agent causes more harm than benefit, it should not be used. Once the diagnosis of AVMs is confirmed, an angiography and embolization should be considered as the purpose of embolization to control the growth of AVMs and frequent bleeding. The key to embolization is to use sufficient embolizing agents to eradicate the nidus. The currently used liquid agents are ethanol and N-butyl-2-cyanoacrylate. Successful embolization is completed when active bleeding stops, localized pulsation has disappeared, the lesion is lighter in color, the expanded veins in neck return to normal and new bones are formed in cystic zones. For females with AVMs, who are planning to become pregnant, it is best to do embolization before pregnancy, because the hormonal changes during pregnancy may accelerate the progress of AVMs. Ethanol is probably the most effective sclerosing agent that can denature the blood proteins and denude the vascular wall of endothelial cells. However, the risks with use of ethanol and the procedure requires skill and experience. If the procedure is not adequately performed, tissue necrosis and more serious complications such as pulmonary arterial hypertension may occur. Soft tissue AVM can be divided into 3 types: infiltrative, nidus and fistula. For infiltrating AVMs, it is suggested to use a mixture of ethanol and contrast at a ratio of 1:1 for embolization where the nidus and AV fistula require absolute alcohol for embolization. If AVMs of soft tissues affect important anatomic structures with severe disfigurements, the most effective treatment isoperative embolization and radical resection. Surgery is indicated when embolization fails or endovascular access of the nidus is not possible. Surgery is very difficult because of vascularity and lack of distinct margins. Surgery should be performed by an experienced surgeon with AVMs and ability to reconstruct immediately. It is common for AVMs to recur after surgery, and the surgeon must be ready to reoperate. The goal of surgery must be to remove the entire nidus and AV fistula will recur. The nidus is very difficult to define because of diffuse feeder vessels and draining veins which do not necessarily have to be resected. In our case, preoperative embolization was not done because the feeding vessels could not be identified by CT angiography. LASER is less effective with AVMs and should rarely be used. The only curative treatment of AVMs is the radical resection which is possible only after a preoperative embolization.

Conclusion

Laryngeal AV malformation is a rare vascular lesion which is usually diagnosed by CT angiography. Preoperative embolization followed by radical surgical resection is the treatment of choice. In our case, since CT angiography could not trace the feeding vessel of the lesion, only surgical resection was done. Regular follow up is necessary to identify recurrence early.


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