

Case Report

Giant haemangioma of the naso-oro-hypopharynx treated by midline mandible split

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Abstract:

Hemangioma is a benign tumour characterised by an increased number of normal or abnormal blood vessels. A 22-year-old male presented with occasional hemoptysis of 3 weeks duration. Clinical examination revealed an angry looking red mass in the oropharynx suggestive of hemangioma. MRI and MRAngiography revealed the mass to be a hemangioma extending from the skull base to the vocal cord level. The hemangioma was excised surgically using a midline labio mandibulo glossotomy approach. Follow up till date has been uneventful.

Key words: Haemangioma, Pharynx

Introduction:

50% of the haemangiomas present in the head and neck.¹ Hemangiomas are usually not present at birth but are antedated by a pale, well-circumscribed flat area that may contain some central telangiectasia.² Hemangiomas of the head and neck region can pose a therapeutic challenge depending on their size, location and symptoms. Haemangiomas are benign proliferative nests of vasoformative tissue. We report a case of a 22-year-old male patient who presented with a hemangioma involving the nasopharynx, oropharynx and hypopharynx. Extensive workup included CT, MRI and MRAngiography. Surgical excision using mandible split was done.

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Case report:

A 22-year-old male patient presented with complaints of blood stained sputum on and off for 3 weeks. There was no preceding history of epistaxis or hematemesis. Past history was not significant.

On examination of the oral cavity; an angry looking red mass was seen in the oropharynx completely occupying the posterior pharyngeal wall. The upper and lower extent of the mass could not be established on oral examination. The mass was seen attached to the posterior pharyngeal wall and did not bleed on touch. Nasal endoscopy revealed the mass occupying the nasopharynx and its upper limit could not be visualized. Indirect laryngoscopy was not possible due to the mass occupying the oropharyngeal space.

CT and MRI neck and skull showed a large lobulated well defined mass extending from the skull base to the C₅ vertebral level, occupying the nasopharynx, oropharynx and

hypopharynx upto C₅ level with T₁ Hypointense and T₂ Hyperintense signal suggestive of haemangioma. No invasion of the surrounding soft tissue or musculature of the pharynx was noted. No bony invasion or intracranial extension was noted.

MR Angiography revealed a mildly vascular lesion along the posterior pharyngeal wall. Multiple thin hair like branches were seen directly arising from the left external carotid artery and supplying the lesion. These arteries were not amenable for embolization due to their small size and hence embolization was not performed. Right carotid artery was not involved.

MRI and MR Angiography features were suggestive of haemangioma.

Management:

Due to inadequate glottic space and hence visualization, elective tracheostomy was performed under local anaesthesia. Patient was intubated through the tracheostomy and subsequently general anaesthesia was administered. The excision of the haemangioma was done as follows:

The lower lip, mandible and the tongue were divided in the midline

Soft palate and posterior half of the hard palate were divided in the midline.

Left external carotid artery was exposed using a neck incision and was clamped temporarily.

The tumour was resected completely from its origin from the posterior pharyngeal wall extending from the skull base to the hypopharynx. No involvement of the soft tissue or musculature of the posterior pharyngeal wall or bony involvement was

seen. Mucosa over the posterior pharyngeal wall was closed by mobilizing the mucosa medially on either side. Divided ends of the mandible were fixed using steel plate. Palate and tongue were sutured with 3-0 vicryl, lips and skin were sutured with 4-0 ethilon.

Nasogastric tube no.16 was inserted. The recovery from anaesthesia was uneventful.

As there was not a significant amount of blood loss, no blood transfusion was required.

Patient was kept nil by mouth for 5 days post operatively. Intravenous antibiotics, analgesics, regular antiseptic mouth washes (2nd day onwards) and appropriate tracheostomy care were given. Nasogastric feeding was continued till 5th post operative day after which patient was started on liquids and then slowly graduated to a semisolid diet. On day 7th the tracheostomy tube was removed. The patient was discharged on the 14th day.

On follow up after 15 days (1 month post operative) the suture line and the pharyngeal mucosa were healthy.



Figure-1: Haemangioma seen in the oropharynx, posterior pharyngeal wall not visualized.



Figure-2: MRI of the Neck showing the extent of the haemangioma: from the base of the skull to the Level C₅. No involvement of the surrounding structures or intracranial extension noted.



Figure-4. Haemangioma measuring 12 x 5 x 2 cms. Lobulated surface. Histopathological report consistent with hemangioma.

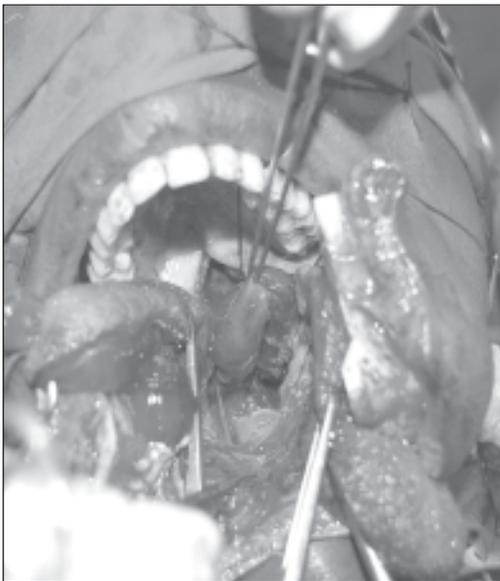


Figure-3. Midline labio glosso mandibulotomy done. Soft palate divided in midline. Haemangioma being dissected off the posterior pharyngeal wall.

Discussion:

Hemangiomas are benign vascular anomalies of blood vessels which may occur in various areas throughout the body, 60% being located in the head and neck.

As to the epidemiology³ of hemangiomas, they occur in 10 to 12 percent of all white children. They are almost twice as common in pre-term infants weighing less than 1000 grams. Interestingly, they occur one-tenth as frequently in Asian and Black children. They are not familial, but 10 percent of patients have a positive family history. Hemangiomas have a 6 to 1 female predominance versus vascular malformations, which have an equal incidence in both sexes.

Classification:

Although the classification systems vary for vascular tumors, Mulliken's scheme divides

hemangiomas into two categories, capillary and cavernous.⁴ Capillary hemangiomas are the most common with an incidence of 1-1.5% in infants. They are characterized by raised, circumscribed, red lobulated lesions. Histologically, they are composed of small thin-walled capillary sized vessels which are lined by single layer of flattened or plump endothelial cells and surrounded by discontinuous layer of pericytes and reticular fibers, generally they are low flow lesions. Cavernous hemangiomas can be high flow lesions, and consist of deep, irregular, dermal blood-filled channels which impart a purple-blue hue to the overlying skin. They are comprised of tangles of thin-walled cavernous vessels or sinusoids that are separated by a scanty connective tissue stroma. Further classification includes a compound hemangioma containing both components also called capillary cavernous hemangiomas.

Clinical course:

Hemangiomas may not be congenital but may be preceded by a pale, well-circumscribed flat area that may contain some central telangiectasia. The actual hemangioma will appear within the first month and will continue to increase in size for the next 3-8 months. A stable phase of relatively no growth then occurs over the next 6-12 months followed by slow involution of the tumor by ages 5-7 years. They can occur just about anywhere in the head and neck, but are more common in the parotid, lip, oral cavity, perinasal region, and larynx or subglottis. The complications of these lesions are ulceration, infection, bleeding, compression syndromes including airway compromise, thrombocytopenia, and even high output cardiac failure. Psychiatric symptoms may be an occasional presentation due to the severe cosmetic deformities that are associated with facial tumours.

Investigations:

Available modalities include CT, MRI, MR Angiography, coloured flow ultrasound.

The best imaging is a MRI, which is the test of choice. Hemangiomas will give you an intermediate signal density on T1. They will be hyperintense on T2 and T1 with contrast they are very hyperintense. They do show high signal flow-voids and you can also see this with arterial venous malformations, but arterial venous malformations are not bright on T2's because they do not have any parenchymal component. The MRI can actually help differentiate between the different types of vascular malformations based on their T1 and T2 contrast enhancement.

There is no uniformly accepted treatment of head and neck hemangiomas. The various modalities of therapy are dependent upon the age of the patient, the site and size of the lesion, and the hemodynamic flow of the hemangioma. It is important to note that congenital lesions typically regress while adult onset lesions do not. The treatment options include: observation, steroids, embolization, cryotherapy, sclerotherapy, antifibrinolytic agents, radiation therapy, laser photocoagulation, surgery with or without preoperative embolization, or any combination of the above.

Observation is encouraged in uncomplicated cases given the natural regression with onset in infancy, but intervention should be considered with lesions that threaten function.

Steroids⁵ may act to increase the sensitivity of the hemangioma to circulating vasoconstrictors, but the overall mechanism is unclear. Dosing can range from 20 to 40 mg per day and have been continued from 2 weeks to 17 months. The response to steroid ranges from 30% to 93% depending on the dose, the duration and the age of onset of treatment.

Interferon⁷ was found useful in hemangiomas while treating HIV patients who also had Kaposi's sarcoma. The Kaposi's sarcoma lesions would get better on interferon therapy. There have been prospective trials, which showed that interferon does work.

The use of the laser to treat hemangiomas by photocoagulation or excision has become popular with the advent of new laser technology. The copper vapor laser⁸ is limited to use with superficial lesions. The argon laser⁹ is more useful for superficial lesions, and may cause skin pigmentation changes as a result of its effects on melanin containing cells. The CO₂ laser¹⁰ is a valuable tool for subglottic hemangiomas where limited penetration will protect the nearby esophagus and great vessels. The Nd-YAG laser¹¹ is especially useful for the deeper cavernous or mixed hemangiomas. The YAG laser, by utilizing the scalpel tip adaptor can also be transformed into a hemostatic excisional tool.

Surgical excision appears the most effective treatment and often results in complete cure. It is reserved for small lesions that fail to regress or the larger lesions that compromise function or cause severe cosmetic deformity. Superselective embolization¹² is recommended pre-operatively in order to reduce the complications of bleeding that can occur. The use of the laser as a pre-operative coagulator can also be utilized in conjunction with surgical excision.

Other modalities such as antifibrinolytic agents¹³, cryotherapy, and sclerotherapy¹⁴ have shown variable efficacy in reducing the size of the hemangioma and are not generally considered as first line therapies. Radiation therapy¹⁵ once considered useful for large haemangiomas has fallen out of favor because of its potential to disrupt growth and induce malignancies.¹⁵

References:

1. Frederick J. Schoen & Ramzi S, Cotran. Ch.12: Blood vessels, Robbins pathologic basis of disease, 6th ed. Pg. 532.
2. Bert WO Malley. Haemangioma of the Head and Neck, Baylor College of Medicine Journal 1992; 25: 29-36.
3. James Ludwick. Hemangiomas and Vascular Malformations of the Head and Neck, Baylor College of Medicine Journal 2002; 35: 46-49.
4. Mulliken JB. Classification of vascular birthmarks. In: Vascular birthmarks: hemangiomas and malformations. Philadelphia: WB Saunders, 1988: 24 - 7.
5. Hawkins DB, Crockett DM, MacLaughlin EF. Corticosteroid management of airway hemangiomas: long-term follow-up. Laryngoscope 1984; 94 : 633 - 7.
6. Sadan N, Wolach B. Treatment of hemangiomas of infants with high doses of prednisone. J Pediatr 1996; 128 :141 - 146
7. Ohlms L, McGill T, Jones D, Healy G. Interferon alfa-2A therapy for airway hemangioma. Ann Otol Rhinol Laryngol 1994; 103 : 1 - 8.
8. Key JM, Waner M. Selective destruction of facial telangiectasia using a copper vapor laser. Arch Otolaryngol Head Neck Surg 1992; 118: 509 - 13.
9. DiBartolomeo JR. Response of vascular lesions of the head and neck to argon laser radiation. Otolaryngol Head Neck Surg 1983; 91: 203 - 6.
10. Muzono G, Dedo HH. Subglottic hemangiomas in infants: treatment with

- CO₂ laser. Laryngoscope 1984; 94: 638-41.
11. McCaffrey TV, Cortese DA. Neodymium:Yag laser treatment of subglottic hemangioma. Otolaryngol Head Neck Surg 1986; 94 : 382 - 4.
 12. Azzolini A, Bertani A, Riverti C. Superselective embolization in immediate surgical treatment: our present approach to treatment of large vascular hemangiomas of the face. Ann Plast Surg 1982; 9: 42 - 60.
 13. Fradis M, Podoshin L, Simon J, Lazarov N, Shagraw I, Boss JH. Combined treatment of large head and neck capillaro-venous malformations by a fibrosing agent. J Laryngol Otol 1089; 103: 390 - 8.
 14. Govrin-Yuhudain J, Moscona AR, Calderon N, Hirschowitz B. Treatment of hemangiomas by sclerosing agents. Ann Plast Surg 1987; 18 : 465 - 9.
 15. Schild SE, Buskirk SJ, Frick LM, Cupps RE. Radiotherapy for large symptomatic hemangiomas. Int J Radiat Oncol Biol Phys 1991; 21: 729 - 35.