INTRODUCTION

The term “hemangioma” represents the most common growth of infancy and childhood and can vary from small innocent birthmarks to large disfiguring tumors. However, for a number of reasons the lesion is commonly used as a misnomer by physicians and thus the terminology is often used interchangeably and inappropriately to classify a large number of vasoformative tumors. Latest recommendation of vascular anomalies suggests hemangioma as vascular tumors which involute during childhood and non-involuting or rapidly involuting congenital hemangiomas. Hemangiomas include dilatation, proliferation, degeneration and malformation of the blood vessels.

Although, hemangima is a relatively common lesion of head and neck region, it is rarely associated with phleboliths. But when they are projected over the region, they may easily be confused with sialoliths, tonsilloliths, calcified lymph nodes, soft tissue tumors, calcification of arteries, metastatic tumors of thyroid and cystecercosis. The rare and unfamiliar presentations of these tumors thus lead to the common practice of inaccurate preoperative diagnosis and inappropriate treatment planning.

Here, we report a relatively rare case of hemangioma with unusual presentation at parapharyngeal space with multiple phleboliths.

Case Summary: A 13 years old patient presented through Outpatient Department of ENT with complain of progressive enlargement of painless lump on right side of the neck since birth of an otherwise healthy male child. Patient denied for any traumatic history or surgical intervention. Clinically, the patient was anemic and shortened in height. Other general physical and systemic examinations were unremarkable. Local examination revealed a soft mass of 13x12 cm in size over right side of the neck extending from tip of the mastoid to the mid-level of sternocleidomastoid muscle up to level III from top to bottom and from lateral border of cricoid cartilage to posterior triangle of neck anteroposteriorly. The lump was non-tender, non-pulsatile, fluctuant, compressible with normal temperature of overlying skin, smooth surface and regular margins, not adherent to overlying skin or underlying tissue. There was significant degree of restriction in neck movement towards right side. There was no audible bruit and ipsilateral cervical nodes could not be assessed. Contralateral side of the neck revealed no abnormality. Indirect laryngoscopy was done which showed slight bulging towards right oropharyngeal wall. On aspiration fresh blood was aspirated which was active and clotted after sometime. A presumptive diagnosis of cystic hygroma, brachial cleft cyst and hemangioma was made.

CASE REPORT

Parapharyngeal Space Hemangioma with Phleboliths: A Rare Entity

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ABSTRACT

Hemangiomas are common finding among head and neck angiomatous benign lesions especially among infants and young children but association with phleboliths is a relatively rare entity. Changes in blood flow dynamics within hemangioma results in thrombus and phlebolith formation. We report a relatively rare case of a large parapharyngeal space hemangioma in a 13 year old male child associated with multiple phleboliths on CT-scan imaging and successful surgical excision and the literature is reviewed.

Key words: Hemangioma, Parapharyngeal space, Phleboliths.

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Chest radiograph and routine hematological investigations were within normal limits. Doppler ultrasonography showed large well-defined complex lesion measuring 7.6x4.8 cm with multiple cystic spaces seen within it with no significant vascularity. Contrast enhanced-Computed Tomography and angiography revealed large lobulated well circumscribed heterogeneous soft tissue density mass with dilated tortuous minimally enhancing tubular structure in the right posterior triangle and superior division of anterior triangle of neck. Superiorly it is extending up to occipital region and inferiorly up to C5 vertebra. Anteriorly it is extending up to retro mandibular region and posteriorly up to nape of the neck. Laterally it is displacing the sternocleidomastoid muscles without evidence of infiltration. Medially, the lesion is abutting the larynx and thyroid gland. The carotid sheath and internal jugular vein are compressed. The lesion is supplied by the branches of external carotid artery and drain via tributaries of external jugular vein measuring up to 9.1x7.9x6.3 cm in anteroposterior, craniocaudal and transverse diameter respectively along with numerous phleboliths measuring up to 0.5cm. (Fig. 2)

It was planned to explore the mass under General Anesthesia. Patient was kept in left lateral position with shoulder bag to extend the neck. After scrubbing and draping, skin crease incisions was made from mid of the neck to posterior border of sternocleidomastoid muscles. Skin flap was raised. Tumor was identified underneath the sternocleidomastoid muscle. All the feeding vessels from external carotid artery were ligated. Blood loss during the procedure was minimal. Tumor was dissected out in toto, drain was placed and primary closure done in layers.

The resected mass was a single unencapsulated pale white to brownish round to oval cystic piece of tissue measuring 7x5x3cm. (Fig. 3) Histological examination revealed fibro collagenous adipose and lymphoid tissue with interconnecting channels of thick walled blood vessels lined by endothelial cells highly suggestive of venous hemangioma. Some of the blood vessels show thrombus formation with calcification. Associated poorly fixed lymph nodes showed reactive changes. Patient was discharged on forth postoperative day. On follow-up, the wound healed completely in two months, post-operative recovery was uneventful. On review after one year, the child was healthy and showed no evidence of recurrence.

**DISCUSSION**

Vascular malformations and hemangiomas are one of the most frequently encountered angiomatous lesions of head and neck region that possess significant morbidity and even mortality in both children and young adults. Current classification proposes the concept of hemangioma as a benign tumor of infancy having a rapid growth phase with endothelial cell proliferation followed either by gradual involution or non-involution. Whereas vascular malformations are structural anomalies of blood vessels without endothelial cell proliferation and persist throughout life\(^2\). From the findings of the history and presentation of the above case, it was categorized as non-involuting congenital hemangioma.

The common practice of conservative treatment of hemangioma has led to the adoption of “watchful neglect” as a common practice while managing this disorder\(^5\). This prolonged stasis of blood in blood vessels leads to changes in blood flow dynamics within hemangiomas resulting in thrombus and phlebolith
Parapharyngeal space hemangioma with phleboliths: a rare entity

formation. Phleboliths are calcified thrombi occur in venules, veins or sinusoidal vessels and characteristic findings of venous or cavernous hemangioma usually multiple in number, small and ovoid in shape with radiologic hallmark of a laminated concentric ring (onion-like) appearance distributed in a laminated pattern.

After extensive search of literature, about 23 cases of head and neck hemangiomas with phleboliths were properly documented and out of those only two of the reported cases were of parapharyngeal origin. Tumors of parapharyngeal space possess prime importance to head and neck surgeons not only because of its complex anatomy and close proximity to vital anatomical structures but also due to the various complex surgical approaches to it.

The pathology is commonly observed among Caucasian females predominantly towards left side with most preferably involving orofacial structures include lips and tongue followed by gingiva, mandible, palate, buccal mucosa and floor of the mouth and salivary glands with almost 80% of hemangiomas occurring as a single tumor. Our patient was a South-Asian young male having a single tumor on right side involving parapharyngeal space but did not have any clinical or radiographic evidence suggestive of salivary gland involvements which are otherwise most common site for hemangiomas. So, we had difficulty with the preoperative diagnosis due to the absence of typical symptoms or signs of a vascular lesion in the parapharyngeal space such as pulsation or bruit.

Venous and cavernous hemangiomas are two of the vascular malformations that do not regress with time and also the history of the presented case suggests the same. Venous hemangioma involving the parapharyngeal space is an extremely rare vascular tumor especially when it is infantile. Both of the hemangiomas contain tortuous vascular channels with stagnant blood flow. This change in the blood flow dynamics result in thrombus and phlebolith formation which is common in both cavernous and venous hemangiomas but an extremely rare finding when involving cervical and orofacial regions.

CT scan with contrast is an excellent imaging technique for revealing phleboliths. Although, MRI is very useful in detecting vascular lesions, the detectability of phleboliths on CT image is superior to that of MRI. It is suggested that plain X-ray films may also reveal calcified lesions. Doppler ultrasound, contrast-enhanced CT scan and angiography were used in present case to diagnose the condition as parapharyngeal venous hemangioma with multiple phleboliths.

It is very important to differentiate venous hemangioma from cavernous hemangioma for the purpose of management but it is sometimes difficult because of their similar characteristics either clinically or radiologically. However, studies have demonstrated the high sensitivity of perfusion and blood pool scintigraphy in detecting head and neck hemangiomas and could also be able to differentiate between cavernous and venous hemangioma. Histopathology is definitive and confirmatory to differentiate between the two. Venous hemangiomas contain dilated vessels with thick, fibrous walls, whereas cavernous hemangiomas have capillary-sized vessels lined by flat endothelial cells.

Management of vascular lesions is based upon location, extent, flow characteristics, symptoms, functional compromises and cosmetic concerns. As majority of the lesions undergo spontaneous involution and about 50% of hemangiomas completely regress by the age of 5 years, definitive treatment is not recommended when discovered in early childhood. The hemangiomatous lesions considered for treatment are grossly enlarged, potentially life threatening and compromising vital or functional prognostics; that entail in severe complication and result in esthetic or functional after-effects.

Excision of the large tumors in parapharyngeal space is challenging because of the potential risk of severe hemorrhage and nerve injury. Generally, hemangiomas in the parapharyngeal space, can be resected through transcervical approach. When surgery is impossible, the use of corticosteroids, cryotherapy, feeding vessel ligation, embolization, and use of sclerosing agents are other alternatives. For the presented case, surgical excision was the best choice and so we opted for that because the tumor was grossly enlarged bulging towards right oropharyngeal wall compromising the airway and has not been regressed since 13 years.

CONCLUSION

Venous hemangioma with numerous phleboliths is an extremely rare finding and should be considered as differential diagnosis of radiopaque lesions of parapharyngeal space. It is important that diagnosticians and radiologists should be involved when presumptive diagnosis and treatment plan is necessitated for successful management.

REFERENCES


