

CASE REPORT

A Surprise Diagnosis of Parapharyngeal Cavernous Hemangioma in an Adult

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ABSTRAK

Ruang parafaring mengandungi banyak struktur penting. Oleh itu, pelbagai jenis patologi mungkin timbul daripada bahagian ini. Hemangioma ialah tumor vaskular yang jarang dan tidak merbahaya. Secara histologi, tumor ini boleh dikategorikan sebagai kapilari, kavernosus dan kapilari campuran. Terdapat pelbagai jenis pendekatan diagnostik dalam membuat diagnosis hemangioma kavernosus. Pengimejan seperti pengimejan resonans magnet (MRI) adalah lebih baik daripada imbasan tomografi berkomputer (CT). Walau bagaimanapun, dalam kes ini kami meneruskan dengan imbasan CT kontras di mana ia menunjukkan jisim peningkatan yang jelas di ruang parafaring kiri dengan berdekatan dengan arteri karotid. Kebanyakan hemangioma tanpa gejala boleh dirawat secara konservatif. Namun, ia bergantung kepada jenis tumor. Kaedah rawatan lain yang boleh digunakan untuk merawat hamangioma kavernous seperti steroid sistemik, sinaran, embolisasi pra operasi atau agen sklerosis. Secara kesimpulan, rawatan yang paling baik adalah dengan mengeluarkan keseluruhan tumor secara pembedahan. Dalam kes ini terdapat dilema diagnostik. Hemangioma parafaring harus menjadi salah satu diagnosis berbanding tumor-tumor lain untuk sebarang pembengkakan leher di ruang parapharyngeal. Pembedahan untuk mengeluarkan keseluruhan tumor ini dapat memberi hasil terbaik daripada ia tumbuh berulang kali tanpa embolisasi praoperasi.

Kata kunci: Hemangioma; kavernous; tumor parafaring; tumor vaskular

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ABSTRACT

Parapharyngeal space contains many vital structures thus various pathology may arise from this region. Hemangiomas are benign rare vascular tumors that are histologically categorised as capillary, cavernous and mixed capillary. We were reporting a rare case of a cavernous hemangioma in the parapharyngeal space. There are various diagnostic approaches in diagnosis of cavernous hemangioma. Imaging such as magnetic resonance imaging (MRI) is better than computed tomography (CT) scan. However, in this case we proceeded with contrasted CT scan where it showed well defined enhancing mass at the left parapharyngeal space with close proximity to the carotid artery. Asymptomatic cavernous hemangioma can be treated conservatively. Depending on the sites of the tumor, other treatment modalities that can be used such as systemic steroids, radiation, pre operative embolisation or sclerosing agents. Complete excision is the best in treating cavernous hemangiomas to prevent the recurrent of the disease. Hemangiomas should be considered as one of the differentials if a mass of soft tissue lesion with high vascularity been revealed by imaging. In this case, there was a diagnostic dilemma. Parapharyngeal hemangioma should be one of the differential diagnosis for any neck swelling over the parapharyngeal space. Complete excision of the tumor brings about the best favourable outcome, with a lower recurrence rate even without preoperative embolisation.

Keywords: Cavernous; hemangioma; parapharyngeal tumour; vascular tumour

INTRODUCTION

Parapharyngeal space is located in the upper neck above the hyoid bone. It is an inverted pyramidal shaped space that extends from the base of skull till the hyoid bone. Medially bounded by the pretracheal fascia, laterally by the pterygoid muscles and mandible. Anteriorly it is bounded by submandibular space and posteriorly by the retropharyngeal space (Kale et al. 2006). Parapharyngeal space contains many vital structures thus various pathology may arise from this region (Kashyap et al. 2015). Hemangiomas are benign rare vascular tumor that are

histologically categorised as capillary, cavernous and mixed capillary (Lad et al. 2017). We were reporting a rare case of a cavernous hemangioma in the parapharyngeal space.

CASE REPORT

We reported a case of, 61 years old woman presented with a 4 months history of painless left side neck swelling which was gradually increasing in size. She did not have any compressive symptoms. She had underlying multinodular goiter which she underwent subtotal thyroidectomy 20 years ago.

Examination of the neck revealed a mass measuring approximately 2.0 x 3.0 cm over the left angle of the mandible which was firm in consistency and was not pulsatile. There was medialisation of the left pharyngeal wall on flexible nasopharyngoscopy, however there was no narrowing of the airway.

Contrast enhanced CT as in Figure 1 and 2 (in axial and coronal plane)

showed a well defined, heterogeneously enhancing left parapharyngeal mass measuring, 4.2 x 4.4 x 6.6 cm. Mass was located in post styloid region with infrahyoid extension. Mass was obliterating and displacing left parapharyngeal fat anteriorly with medialisation of left lateral pharyngeal wall consistent with scope findings. Left external carotid artery was in close

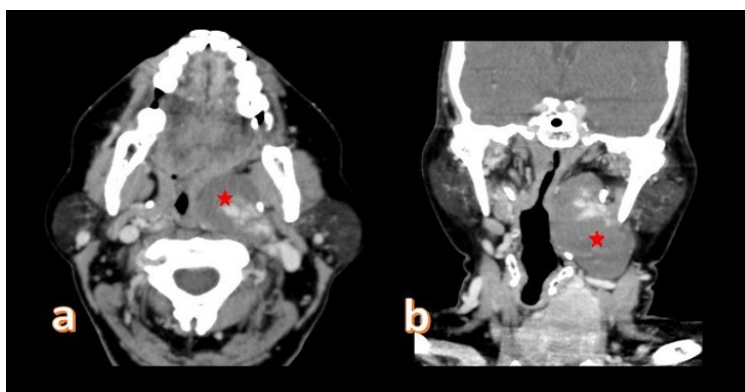


FIGURE 1: Contrast enhanced CT images in (a) axial (b) coronal plane; showed a well defined heterogeneously enhancing left post styloid parapharyngeal mass, obliterating and displacing parapharyngeal fat space, anteriorly. Evidence of medialization of left lateral pharyngeal wall, causing narrowing oropharyngeal lumen. Enlarged left thyroid lobe noted with clear demarcation between the mass

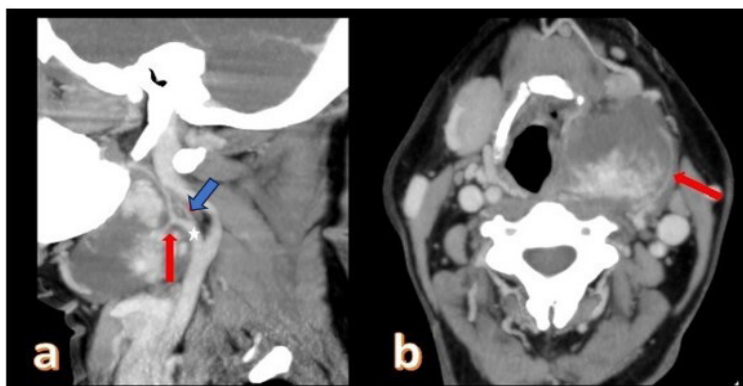


FIGURE 2: Contrast enhanced CT images (a) sagittal (b) axial plane; mass was encased and supplied by lingual artery (long red arrow); a branch of external carotid artery. Left ascending pharyngeal artery (short red arrow) was seen posteriorly with distal branch which was not well visualised

proximity with the mass while left lingual artery was seen encasing and supplying the mass, anterolaterally.

Fine needle aspiration of left neck swelling was unsatisfactory. Patient then proceeded with ultrasound guided fine needle aspiration of the left parapharyngeal mass which was also reported as unsatisfactory.

In view of the unsatisfactory cytology findings, a decision was made to proceed with complete excision of the mass via transcervical with diagnostic and therapeutic approach. Intraoperatively the tumor was found well capsulated lobe, soft in consistency with size measuring 8 x 5 cm (Figure 3). There was a feeder



FIGURE 3: Highly vascularised, well encapsulated mass, measuring 8 x 5 cm

arterial vessel possible intent from lingual artery which the tumor was ligated. Tumor was removed in total with minimal intraoperative bleeding which was about 100 mls. Surgery was performed using harmonic scalpel.

Histopathological examination revealed the excised tumor to be a cavernous hemangioma. Microscopically, the lesion is composed of dilated vascular channels lined by benign endothelial cells. Thrombosed blood vessels were present with surrounding histiocytes surrounding and minimal lymphocytes were noted in the slightly fibromyxoid stroma as in Figure 4.

Post operatively, patient had deviation of tongue to the left, as a result of left hypoglossal nerve

paresis which gradually resolved after 2 months. Patient had no evidence of recurrence at 16 months after excision.

DISCUSSION

Parapharyngeal tumors include benign or malignant salivary gland neoplasms, neurogenic tumors and other miscellaneous tumors. Pleomorphic adenomas, paraganglionic and schwannomas are found in the prestyloid space. Based on studies done, out of 16 patients, 14 underwent surgery, 78.6% were reported as benign and 21.4% had malignant tumors. Out of this benign, 50% were reported as schwannoma (n=11), 18% as paraganglionoma (n=2) and 8% as hemangioma (n=3) (Kuang et al. 2019).

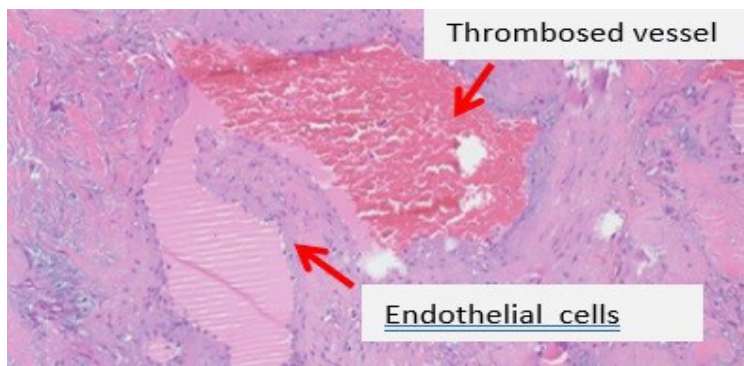


FIGURE 4: Histopathology shows irregular vascular channels which are filled up with blood, space lined by a single layer of endothelial cells and thrombosed vessels (hematoxylin and eosin, original magnification x10 under high power field)

Tumors reported in post-styloid space include neurogenic, metastatic masses, and tumors of ectopic salivary glands.

Cavernous hemangioma is a rare vascular-related neoplasm formed by abnormal dilation of venous blood vessels. In a study conducted by Watson and McCarthy in 1940, 56% of all hemangiomas were seen to occur in the head and neck region. Out of these cases, 85% of cases were reported in newborns and first-year infants (Kale et al. 2006; Kashyap et al. 2015). Hemangiomas are classified into three histopathological types; capillary, cavernous, and mixed type. The capillary type is usually surrounded by tissues that undergo proliferation. The cavernous types, in this case, are characterised by mitotic activity with well-capsulated vascular structures. The mixed type resembles both mitotic and proliferative activity (Kiyamaz et al. 2010).

There are various diagnostic approaches in the diagnosis of cavernous hemangioma. One of the best modalities imaging to diagnose

hemangioma is magnetic resonance imaging (MRI). MRI demonstrates hemangioma with excellent contrast between the normal tissue and lesion, shown by Itoh et al. (1986). According to Murata et al. (1997), scintigraphy has been demonstrated to have high sensitivity for detecting head and neck hemangiomas. Ultrasound Doppler is a useful imaging tool for soft tissue vascular anomalies. However, it has limitations for lesions that lie deep in the para-pharyngeal space. Berg et al. (1986) revealed computed tomography (CT) scan has sufficient accuracy to predict the histological diagnosis of the lesion but CT is less precise than MRI for determining the extent of disease as it has lower soft tissue sensitising (Vogl et al. 2010). Without having a vascular tumor as our primary diagnosis, based on our clinical findings where the mass was firm in consistency on palpitation. We proceeded with a CT scan rather than MRI as our standard imaging tool.

Asymptomatic cavernous hemangioma can be treated conservatively. Depending on the

sites of the tumor, other treatment modalities can be used such as systemic steroids, radiation, pre-operative embolisation, or sclerosing agents. Complete excision is the best in treating cavernous hemangiomas to prevent the recurrence of the disease. Wolf et al. (1985) and Tang et al. (2002) reported a local recurrent rate of 18% after incomplete surgical excision of the lesion.

In histopathological examination, 4 types of immunohistochemical staining were used, CD34 and smooth muscle actin (SMA) which was positive but CKAE1/AE32 and CD 68 were negative. CD 34 is a transmembrane glycoprotein expressed on early lymphohematopoietic stem cells, progenitor cells, and endothelial cells. SMA marker is a sensitive marker of myoepithelial differentiation (Declercq et al. 2012). This staining favors a hemangioma type of lesion in this patient (Mieke et al. 2022).

CONCLUSION

Parapharyngeal cavernous hemangioma is a rare entity. From this case we learnt that there was a diagnostic dilemma. Hemangiomas should be considered as one of the differentials if a mass of soft tissue lesion with high vascularity has been revealed by imaging. Complete excision of the tumor brings about the best favourable outcome with lower recurrence rate even without preoperative embolisation.

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