

Recurrent Lymphovascular Hamartoma of the Neck Spaces

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Citation

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Abstract

Lymphovascular hamartoma of the neck spaces is a rare occurrence. To the best of our knowledge, very few of them have been described in literature in the parapharyngeal and submandibular spaces. When it occurs in these spaces, it poses special diagnostic and therapeutic problems to both the pathologist and the surgeon since it can easily masquerade other bleeding tumors like angiofibroma. Careful correlative study of the histopathology and radiography along with high index of suspicion of this apparently innocent tumor like lesion is the key to the diagnosis. A wide open surgical approach and complete excision are required to prevent recurrences.

KEY MESSAGES

- Denovo lymphovascular hamartoma of the neck spaces is rare
- Masquerades bleeding tumors like angiofibroma
- Easily missed without a high index of suspicion
- Recurrences common if incompletely excised
- Wide open surgical approach required for complete excision

INTRODUCTION

A hamartoma may occur in any organ, but it most often involves the gastrointestinal tract.³ It remains a rare finding in the head and neck region and may hence lead to a misdiagnosis of a tumor in the presence of epithelial proliferation.¹¹ In spite of not being a neoplasm in the true sense, the clinicopathological behavior of a hamartoma is very much like that of a neoplasm and is known to recur. We describe one such interesting case that posed a diagnostic and therapeutic challenge to us, but we still managed it appropriately.

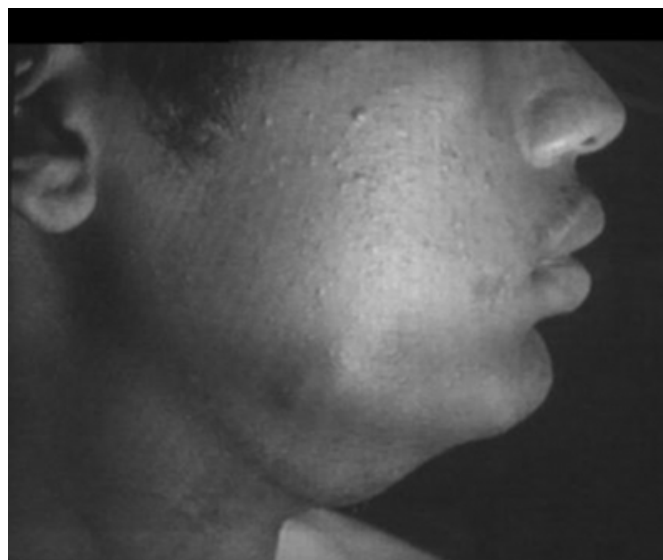
CASE REPORT

A 20 year old male patient presented to us with a recurrent swelling in the right side of the cheek and neck of 15 days duration. He also complained of difficulty in opening the mouth and difficulty in swallowing for the same duration. He had been operated twice before for similar complaints 4

years back. On inspection of the neck, a horizontal surgical scar was visualized parallel to the lower margin of the right side of the mandible. A diffuse firm swelling was also noticed on the right side of the neck extending from the parotid to the submandibular region crossing the mandible [Fig 1].

Figure 1

Figure 1: Lateral profile of the patient showing the swelling in the neck



In the oral cavity, a bulge was observed in the right lower buccal mucosa and the right tonsil was pushed medially. On palpation, the swelling was found to be non tender, mobile in all directions except superiorly and was not attached to the mandible. In the oral cavity, a bulge was observed in the

right lower buccal mucosa and the right tonsil was pushed medially.

CT scan of the head and neck revealed a well defined cystic lesion in the right submandibular region anterior to the sternocleidomastoid muscle and below the parotid gland extending to the parapharyngeal space. The inferior extent of the lesion was up to the superior border of the thyroid cartilage. The lesion had displaced the carotid vessels posteromedially, but had not involved them [Fig 2, 3].

Figure 2

Figure 2: Axial section of the post contrast CT scan of the head and neck showing the mass

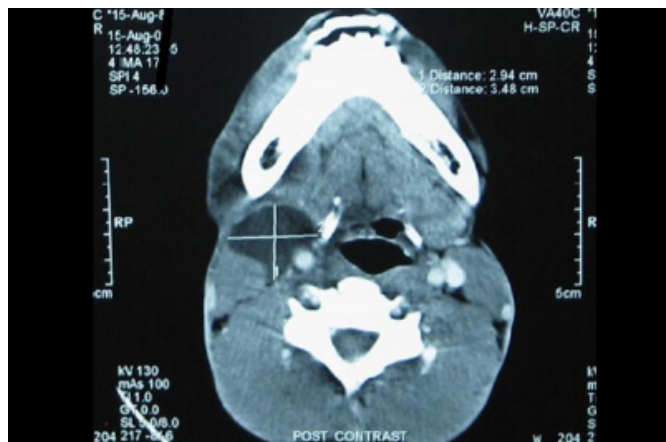
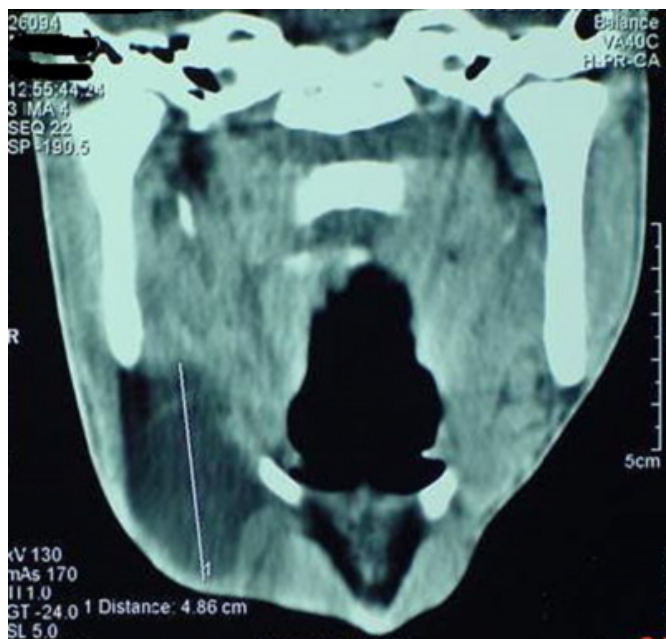


Figure 3

Figure 3: Coronal section of the post contrast CT scan of the head and neck showing the mass

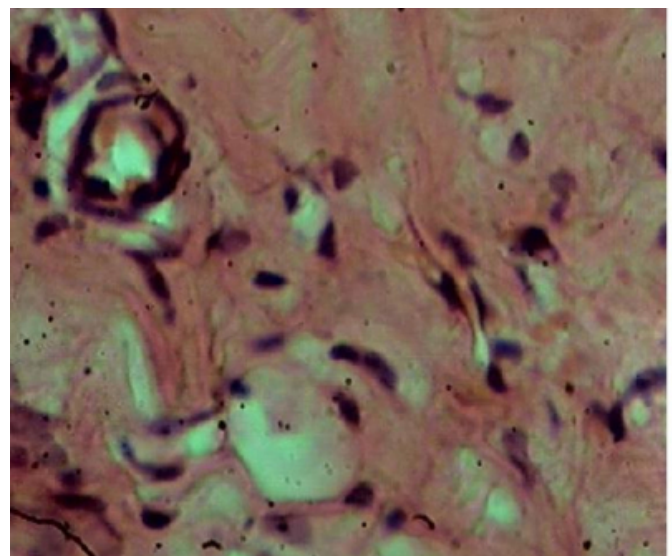


There was no evidence of internal septations or solid

elements within the lesion. There was also no evidence of cervical lymphadenopathy. Fine needle aspiration cytology of the mass was inconclusive. Hence a definitive excision of the tumor was planned using the external transcervical approach. There was extensive fibrosis of the soft tissues around the mass and dissection was difficult. The mass was found to be cystic and it bled profusely during excision. Bleeding was controlled by pressure packing and thermal cautery. Two units of whole blood were transfused. The mass was excised in toto from all its extensions and sent for histopathological examination [Fig 4]. Biopsy report read lymphovascular hamartoma. Fortunately the third time, no recurrence was observed 6 months after surgery.

Figure 4

Figure 4: Pictomicrograph of the excised mass [H & E stain, 40X]



DISCUSSION

The term 'hamartoma' was introduced by Albrecht in 1904¹, who distinguished between true neoplasms and tumor like lesions. Even though it resembles a tumor, it is actually a non neoplastic malformation or an in-born error of tissue development, often with an abnormal mixture of tissue indigenous to the region.³ A hamartoma may occur in any organ as it designates a focal overgrowth of mature normal cells and tissues at sites of identical cellular composition.²

The epithelial and mesenchymal hamartomas are uncommon in the head and neck region.³ In the head and neck, they have been described in the nasal cavity,⁴ nasopharynx,⁴ larynx,² oropharynx,⁷ hypopharynx,⁸ ear,⁹ Eustachian tube,¹⁰ and the deep neck spaces.³ Intracranial lesions like the extracerebral glioneural hamartoma extending into the parapharyngeal

space have also been described.³ Since denovo hamartomas are uncommon in the parapharyngeal space, they are easily missed or mistaken for other common tumors.

Symptoms caused by hamartomas of the neck are related to the affected site. They are not pathognomonic and may include dysphagia, trismus and swelling in the neck, that too when the size of the mass becomes significant. Hence there is often a delay in the patient presenting to the clinician. In advanced stages, these 'tumors' produce pressure symptoms in the neck even though they may not actually invade the structures.

Hamartomas have no capacity for continuous unimpeded growth, so their proliferation is self limiting.⁴ They have no tendency to regress spontaneously either. Nevertheless, they are prone for recurrences especially when excised incompletely. This mixed bag of characteristics has disputed the neoplastic origin of this uncommon lesion. Some authors believe that the hamartomas are benign neoplasms rather than just malformations.⁶ Hamartomas can be classified into mesodermal component containing type that are more frequent or the epithelial or glandular element containing type that are less frequent. Microscopically the lymphovascular hamartomas contain fibrocollagenous tissue, adipose tissue, skeletal muscle fibers, few nerve bundles, immature or primitive mesenchyme along with lymph channels and blood vessels interspersed between them as was seen in our case. This picture could masquerade an angiofibroma that is a true benign neoplasm, even though uncommon in that region. Besides angiofibroma, the differential diagnosis for this mass would include hemangioma, lymphangioma or a teratoma.

CT scan with contrast is an indispensable diagnostic tool in these cases to know the nature of the mass, consistency, extensions and attachments of the mass. CT angiography may additionally reveal the vascularity of the tumor.

Complete surgical excision through a wide open surgical approach is the treatment of choice to prevent recurrence. Repeated excisions and recurrences could lead to fibrosis and scarring of the tissues in the neck. External transcervical approach with or without mandibulotomy is preferred for complete access to all the extensions of the mass in the

parapharyngeal and submandibular regions. The thin walled blood vessels in the lesion are likely to bleed profusely during surgery and could make it difficult to excise the mass completely.

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