A Large Parapharyngeal Mass in A Five-Year-Old Girl: A Case Report

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Received: 21 Feb. 2022; Accepted: 26 Jan. 2023

Abstract- Lesions in the parapharyngeal space are rare and account for 0.5-1.5% of all head and neck tumours. Of these, venous malformations represent 1% of all parapharyngeal space tumors. In the current paper, we reported a 5-year-old patient who presented with a soft tissue swelling of the parapharyngeal space that was discovered by her parents after two weeks following adenoidectomy. The MRI revealed a tumour within the parapharyngeal space with a well-defined mass with low-signal intensity on T1 and high on T2. The internal flow voids also proposed phleboliths. Therefore, the tumour was removed by an intraoral approach. The post-op pathology result explicated a venous malformation.

Acta Med Iran 2023;61(3):187-190.

Keywords: Parapharyngeal space; Venous malformation

Introduction

The parapharyngeal space is a potential region beneath the skull base with a narrow connective tissue. This space is bordered superiorly by the base of the middle fossa and inferiorly by the hyoid bone. It is surrounded medially by the superior constrictor muscle, anteriorly by the pterygomandibular raphe (pterygomandibular ligament), posteriorly by the prevertebral fascia, and laterally by the deep lobe of the parotid gland. The styloid process splits the parapharyngeal space into a pre-styloid and post-styloid compartment. The pre-styloid space comprises the deep parotid lobe, minor salivary glands, lymph nodes, and fat contents. The post-styloid space contains the IX, X, XI, and XII cranial nerves, the cervical sympathetic chain, the internal jugular vein, and the internal carotid artery. This complex anatomy of the parapharyngeal space is responsible for various tumours. Tumours of the parapharyngeal space account for 0.5-1.5% of all head and neck tumours. Pre-styloidtumours commonly arise from the salivary origin, whilst the post-styloid space is most often affected by neurogenic tumours (1).

Among parapharyngeal tumors, 48% originate from salivary glands. The most common tumouris the pleomorphic adenoma, and 20% of those include lipoma, leiomyoma, teratoma, rhabdomyosarcoma, lymphangioma, and angiomatous malformation (2).

Imaging is crucial for the assessment of parapharyngeal space tumours. Magnetic resonance (MR), computed tomography (CT) with contrast, and contrast angiography in selected cases are essential for diagnosis. However, MR is the modality of choice for evaluating parapharyngeal lesions (1).

Five surgical approaches to tumours of the parapharyngeal space are: 1) transoral, 2) transcervical, 3) transparotid, 4) transmandibular (usually combined with transcervical), and 5) lateral skull base approaches (3). In the current paper, we reported a 5-year-old patient who presented with a soft tissue swelling of the parapharyngeal space that her parents found after two weeks following adenoidectomy.

Case Report

A five-year-old girl was referred to our tertiary

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centre with a history of snoring, hyponasal speech, and recent adenoidectomy with a pathologic result for follicular lymphatic hyperplasia. Her parents realised the swelling on the left side of her neck two weeks following the mentioned surgery-adenoidectomy.

On physical examination of the mouth and throat, an enlargement of the left parapharyngeal space with an intact mucosal surface was observed. A deviation of the uvula to the opposite side was also noted. Therefore, the patient underwent a thorough radiological assessment.

Color Doppler ultrasound revealed a 55×30 mm hypoechoic mass with internal echogenic foci , resulting in a compressive effect on the left palatine tonsil and medial displacement of the lateral pharyngeal wall and medial displacement of the lateral pharyngeal wall tonsil.

A 41×55 mm well-defined mass in the left parapharyngeal space with lobulated borders and patchy enhancement was observed on CT-scan. There was also some calcification in the lesion—phleboliths— in this imaging modality—neck CT scan with contrast (Figure 1).

MRI detected a well-defined mass with a low-signal intensity on T1 and high on T2. The internal flow voids were highly suggestive of phleboliths. Following Gadolinium injections, the lesion displayed a mild diffuse patchy enhancement. However, these findings highly suggest venolymphatic malformation (VLM) (Figure 2).

The patient underwent an intraoral surgical approach. A venous blood sample was initially aspirated from the mass throughout the surgery. Consequently, the mass was removed entirely from the left parapharyngeal space. At the end of the surgery, a safe airway was provided by tracheostomy. Decannulation was performed after three days following the surgery. Ultimately, the patient was discharged without complications four days following the operation.

Finally, the pathology results confirmed venous malformation with foci of necrosis and calcification.

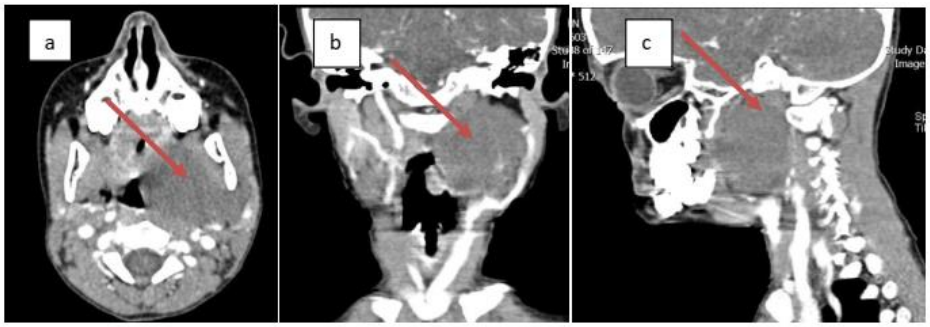

Figure 1. These CT images reveal a soft tissue density similar to muscle. a)axial, b) coronal, and c) sagittal plane

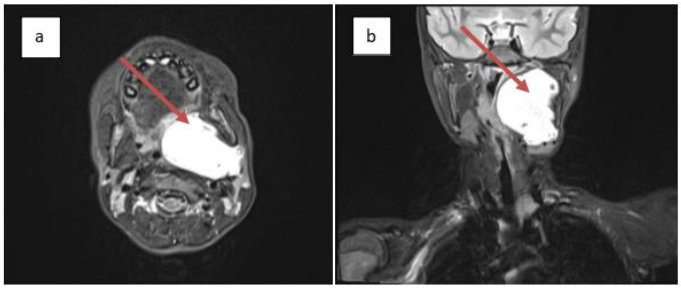

Figure 2. On T2-weighted images, a soft tissue mass in the parapharyngeal space is observed; a) axial and b) coronal

Discussion

Parapharyngeal space masses are uncommon. They comprise only 0.5-1.5 % of all head and neck tumours. Of these, VLMs represent 1% of all parapharyngeal space masses. VLMs represent 1% of all parapharyngeal space masses (4). Like other masses of this space, VLMs manifest in the intraoral or cervical region. Usually, they may be discovered incidentally on imaging studies for other clinical reasons.

Venous malformations (VMs) typically represent an unencapsulated, circumscribed, or trans-spatial lesion and may contain phleboliths. In such lesions, low flow on imaging is observed (5). The International Society for the Study of Vascular Anomalies (ISSVA) classifies vascular anomalies into two categories, i.e., tumours and

malformations. The most common vascular tumour is infantile hemangioma, whereas VM is the most common vascular malformation (6).

The VM may represent a well-circumscribed, mildly lobulated enhancing mass that is isodense to muscle on CT and markedly T2 hyperintense on MR imaging. Until now, very few VM cases with imaging characteristics of primary parapharyngeal space have been reported. These reports have frequently described these lesions as "hemangiomas, " reflecting a controversial ongoing usage of older nomenclature (7).

In those reports, on MR imaging, all VMs were well-defined and comparable with skeletal muscle markedly hyperintense on T2WI and isointense to minimally hyperintense on precontrast T1WI. The signal voids may manifest in cases with phleboliths. In a study of head and neck VMs, phleboliths were reported in only 28.6% of cases (8). In this study, postgadolinium T1WI revealed highly variable enhancement patterns. Although this is frequently patchy and delayed, demonstrating sequential filling of the lesion with slow washout was observed (7).

VMs appear as lobulated hypoechoic masses at sonography. with small internal venous channels. On spectral Doppler ultrasound, waveforms are venous, and a curvilinear hyperechoic centre with posterior shadowing might be observed if phleboliths are present (9).

In our case, almost the entire characteristics of VM were displayed in pre-operative radiologic studies. These features included hyperintensity on T1 MRI. Internal flow voids are suggestive of phleboliths (Figure 3), soft tissue density similar to muscle with calcified centres suggestive of phleboliths on CT scanning (Figure 1), and a mass containing venous and calcified foci suggestive of phleboliths canals on colour-doppler ultrasound.

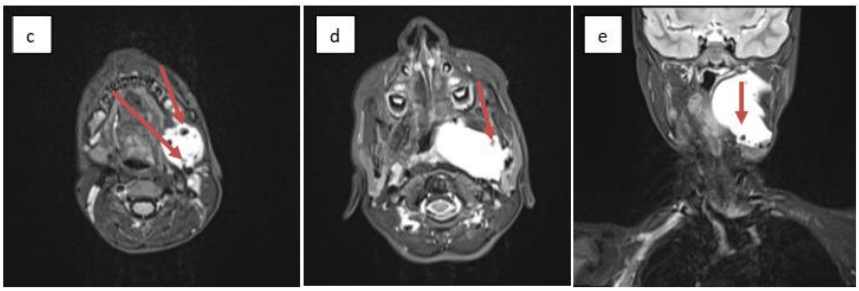

Figure 3. Internal flow voids are suggestive of phleboliths on MRI (red arrows). c) and d) axial, and e) coronal view

Because of the rareness of parapharyngeal space venous malformation and diagnostic overlap with other more common lesions in this space, such as pleomorphic adenoma, this diagnosis should be considered in differential diagnoses, particularly in pediatric patients. Recognition of typical imaging characteristics of venous malformation in pre-operative radiologic examinations may guide us in determining the best surgical approach.

Acknowledgements

The authors thank Dr Ali Kazemi for reporting all radiologic images and Dr TaherehYousefi for reporting the results of the examination of the specimen.

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