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Lymphoma presenting as a parapharyngeal space tumour

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Abstract

Among all head and neck tumours, approximately 0.5% to 0.8% localize in the parapharyngeal space, with Non-Hodgkin's lymphomas in this region being exceptionally rare. They are documented in the literature either as isolated cases or in small case series. Here, we present a case involving a middle aged man who exhibited 4 month history of dysphagia and hoarse voice. Upon examination, showed a bulged left side of the soft palate and medialization of the pharyngeal wall. CT and MRI revealed a large lesion in the left parapharyngeal region pushing the pharynx anteromedially. Similar intensity enhancing lesion noted in nasopharynx. A transoral excision of parapharyngeal mass was performed, revealing a large B-cell non-Hodgkin's lymphoma. Typically, parapharyngeal space tumours are benign, and surgery is the preferred treatment. Patient was transferred to tertiary care oncology hospital for further treatment. This case underscores the rarity but potential presence of extranodal non-Hodgkin's lymphomas in the head and neck. Recognizing this pathology is crucial due to the different treatment approach it necessitates.

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Introduction

The parapharyngeal space, situated laterally to the pharynx, takes on an inverted pyramid shape that extends superiorly from the base of the skull to the greater cornu of the hyoid bone. It is enveloped anteriorly by the medial pterygoid muscle, posteriorly by prevertebral fascia, medially by pharyngeal constrictor muscles, and laterally by deep parotid fascia. This space contains crucial vessels and nerves, including the internal carotid artery, internal jugular vein, vagus nerve, sympathetic nerves, glossopharyngeal nerve and hypoglossal nerve. Anatomically, the styloid process divides the parapharyngeal space into a pre-styloid compartment anteriorly and a post-styloid compartment posteriorly, a division that proves significant in discerning parapharyngeal space tumours[1]. These tumours in the primary parapharyngeal space make up about 0.5% of all the neoplasms in the head and neck[2].

Clinical indications of parapharyngeal space tumours don't readily present themselves, leading to frequent delays in diagnosis. Furthermore, the deep-seated anatomical location of the parapharyngeal space poses challenges for preoperative biopsy-based histological diagnosis, often resulting in delayed initiation of malignancy treatment. Surgery typically serves as the primary treatment, with various approaches available, including the transoral, transparotid, and transcervical methods. In instances where achieving an adequate visual field proves challenging, midline mandibulotomy may become necessary[3]. Although recent advancements include endoscopic and robotic-assisted surgeries, these procedures are often characterized by a high level of complexity. This case report outlines the instance of relatively uncommon parapharyngeal space tumours, characterized by challenging diagnoses, accompanied by a comprehensive discussion of pertinent literature.

Case Report

54 year old male patient with diabetes presented with 4 months history of dysphagia and hoarse voice, with occasional aspiration of liquids, without any breathing difficulties. He was previously evaluated for nasal obstruction and snoring, where bulging in the nasopharynx was found and biopsy was taken and reported as chronic inflammation of respiratory epithelium, without any dysplasia, invasive malignancy or lymphoma. On examination, there was a bulging of left lateral oropharynx with normal overlying epithelium. Fiber optic nasolaryngoscopy revealed nasopharynx obliterated by a bulging mass more in left side with normal looking epithelium and also noted he had right sided vocal cord palsy with a phonatory gap.

He underwent contrast enhanced CT and MRI scans of head, neck and chest, and ultrasound scan neck and abdomen. MRI showed large lesion in the left parapharyngeal region measuring 3cm(AP) x 3cm(T) x 3.6cm(CC) displacing the pharynx anteromedially. Similar intensity enhancing lesion noted in nasopharynx measuring 5cm (T) x 3.4cm x 2.1cm (AP).

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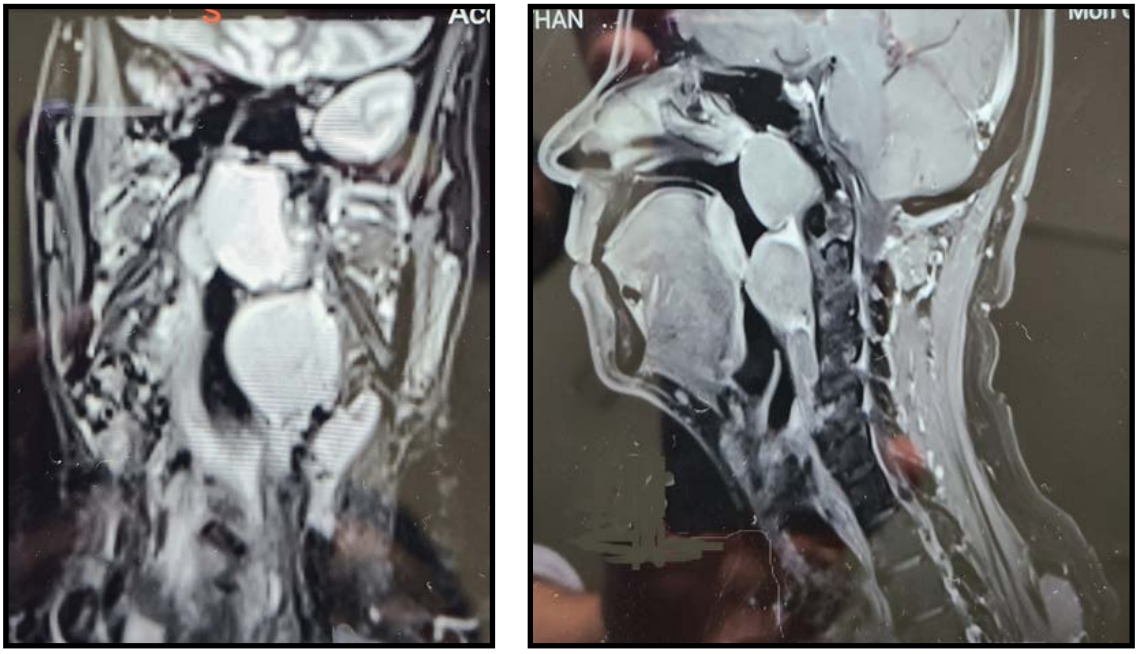

Figure 1. Coronal and sagittal sections of MRI shows two separate lesions in the nasopharynx and left side parapharyngeal space

Parapharyngeal tumour was resected trans orally (endoscopic assisted) and sent for histology. Specimen was reported as diffuse B Cell lymphoma. He was transferred to tertiary care oncology hospital for further management.

Discussion.

Parapharyngeal space tumours, accounting for 0.5% to 0.8% of all head and neck tumours, pose a diagnostic challenge due to their few and late symptoms, Parapharyngeal space tumours exhibit a variety of histological tissue types[3], making preoperative histological diagnosis challenging; hence, reliance on differentiation through CT or MRI becomes essential[4]. The parapharyngeal space is broadly partitioned by the styloid process into an anterior pre-styloid compartment and a posterior post-styloid compartment. Typically, we distinguish parapharyngeal tumours based on the structures present in each area. In the pre-styloid compartment, parotid gland tumours are most common, alongside lipomas, lymphomas, and their malignant counterparts elsewhere. Conversely, in the post-styloid compartment, neurogenic tumours take precedence, including paragangliomas, connective tissue tumours, carotid aneurysms, lymphomas, and their malignant counterparts elsewhere [1]. When it comes to neurogenic tumours, the differentiation of originating nerves relies on the positional relationship between the internal jugular vein and the internal external carotid arteries.

Parapharyngeal space tumors, accounting for 0.5% to 0.8% of all head and neck tumors, pose a diagnostic challenge due to their few and late symptoms, often presenting as asymptomatic masses[5, 6]. When symptoms manifest, dysphagia, foreign body sensation, and pain are common indicators. In our case, the initial symptoms emerged in the advanced phase of tumor growth, coinciding with nasopharyngeal extension. The majority (70-80%) of parapharyngeal space neoplasms are benign, with pleomorphic adenoma being the most prevalent, followed by miscellaneous benign tumors, paragangliomas, and neurogenic tumors.

Despite advanced diagnostic tools like multiplanar CT, MRI, angiography, and fine needle aspiration cytology (FNAC) or superficial biopsy, diagnosis remains challenging[7]. Radiological features specific to parapharyngeal space lymphomas are lacking, making imaging valuable for assessing extent rather than providing a definitive diagnosis. FNAC, while limited by nondiagnostic samples (25%) due to bleeding and technical challenges, does not significantly contribute to parapharyngeal tumor differential diagnosis. Histopathology and immunohistochemistry stand as the gold standard for diagnosis.

Distinguishing lymphomas from other parapharyngeal tumors is crucial for treatment planning. Surgical intervention is typically recommended for parapharyngeal tumors, but for lymphomas, radiation therapy serves as the primary treatment modality for localized disease (stages I and II), particularly for low-grade lymphomas. Advanced disease and intermediate to high-grade lymphomas may require a combination of chemotherapy with or without radiation. Surgery is mainly reserved for diagnostic purposes.

Key Messages /Conclusion

In conclusion, while extranodal non-Hodgkin's lymphomas in the head and neck are rare, recognizing them is vital due to their distinct treatment approach.

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