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INTRODUCTION

Neurofibromatosis type 1 is an autosomal dominant multisystem genetic disorder characterized by cutaneous signs such as skin neurofibromas and café au lait spots. Benign neurofibroma of the parapharyngeal space may occur as part of type 1 neurofibromatosis [1]. Due to the complex anatomy of the parapharyngeal space, various surgical approaches have been devised for the excision of tumors in this area. In general, external approaches are more favored due to better surgical field exposure. Here, we present a rare case of a huge parapharyngeal space neurofibroma treated surgically through a transoral approach.

CASE REPORT

A 29-year-old male with underlying neurofibromatosis type I presented with complaint of muffled voice and dysphagia for the past 3 years. However, there was no odynophagia, dyspnea or aspiration symptoms. The dysphagia resulted in loss of weight of about 8kg over a 9 months period.

Examination revealed multiple huge cutaneous neurofibromas with café au lait spots all over his body. Neck examination revealed a right neck swelling which was firm, mobile and non-tender. Intraorally, there was medialization of the right lateral pharyngeal wall and tonsil with normal mucosa overlying the mass (Figure 1). The mass was firm and non-pulsatile. Flexible laryngoscopy revealed a right oropharyngeal mass extending until the vallecula which displaced the epiglottis posteriorly. Airway was otherwise patent and vocal cords were mobile bilaterally. MRI neck reported a well circumscribed lesion at the prestyloid region of the right parapharyngeal space pushing the internal carotid artery posterolaterally. The lesion did not encase or abut any major blood vessels.

Excision of the right parapharyngeal mass was performed via a transoral approach of which an 80mm x 40mm x 35mm well encapsulated, firm mass was removed in total (Figure 2). This was done via a vertical incision made lateral to the right anterior pillar and once the mass was identified, it was dissected along the fascial plane using harmonic scalpel. The wound was sutured (Figure 3) and nasogastric tube inserted for feeding. HPE was reported as neurofibroma. Post-operatively, there were no complications.

DISCUSSION

Parapharyngeal space tumors account for less than 1% of all head and neck neoplasms. Of these tumors, 80% are benign. Both benign and malignant tumors may arise locally from any of the structures in the parapharyngeal space. It can also be an extension from adjacent regions or occur as metastases from distant malignancies [2,3]. Neurofibromas account for approximately 10% of parapharyngeal space tumors [1].

Most parapharyngeal space masses are treated by surgical excision but due to the complex anatomy, resection of the tumor is challenging. The four basic approaches are transcervical, transparotid, mandibular swing and transoral approaches [2]. Most surgeons prefer the transcervical approach due to better surgical field exposure. Transoral approach is the most controversial one due to cited limited exposure, risk of tumor spillage, increased risk of bleeding, neurovascular injury and risk of incomplete removal. According to Olsen, the transoral approach is considered to be unsafe since it is correlated with many post-operative complications such as hemorrhage, fistulas, dehiscence, and nerve damage [4].

Therefore, the surgeon should possess good surgical skills and experience in order to perform transoral approach for large parapharyngeal space tumors. Even though the lesion in our case was large, it was located at the prestyloid region which provided easier access compared to poststyloid region. The mass was not of vascular origin and was benign. This is evident by the well circumscribed feature of the mass on MRI with no surrounding extension or invasion of surrounding structures. Goodwin et al. [5] concluded that transoral approach provided an adequate access to the parapharyngeal space. When compared to transcervical approach, it is associated with lower post-operative complication rates and showed some advantages despite certain limitations. The main limitation would be the limited surgical field exposure. Betka et al. [6] advocate the transoral approach due to its superior visual control of dissection over the medial and upper parts of the tumor, short surgical and hospitalization times, and good functional and cosmetic outcome. However, the cases must be carefully selected preoperatively.

With the advancement of technology, robotic surgery has also been shown to produce good results by overcoming the visual and instrumental limitations of the conventional transoral approach. However, this is limited by the availability of equipment and expertise [7].

CONCLUSION

Parapharyngeal space tumors are located in a complex anatomical position in our neck and surrounded by various vital structures. Therefore, the choice of surgical approaches must be based on careful consideration of the tumor's size, its location, vascularity and likelihood of malignancy. It has been shown that through careful selection of cases in addition to proper surgical expertise and skills, it is possible to perform the transoral approach to excision of parapharyngeal space tumors safely.

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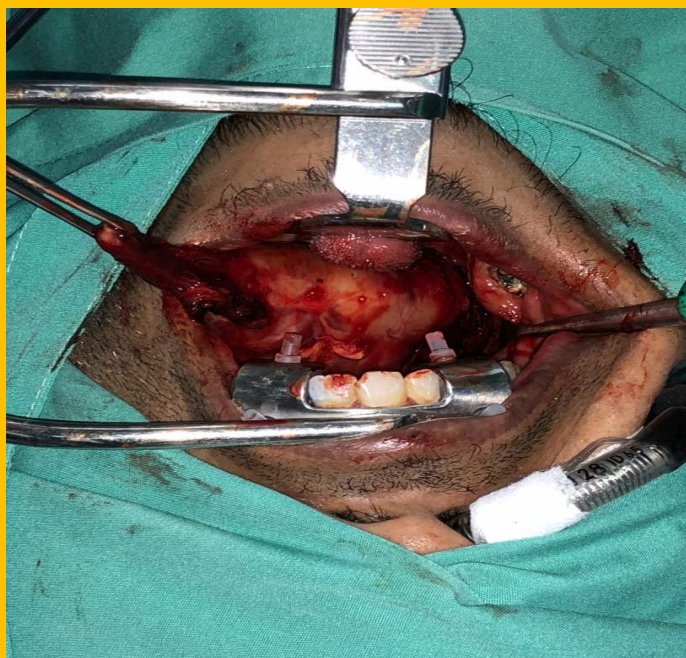


Figure 1: Intraoral view of the huge right parapharyngeal mass on traction using forceps



Figure 2: Measurements of the excised right well encapsulated parapharyngeal neurofibroma



Figure 3: View of the lateral oropharyngeal wall post excision of the right parapharyngeal neurofibroma