Giant Parapharyngeal Schwannoma With A Massive Endocranial Extension

M Jovanovic, I Berisavac, M Markovic, V Bojovic, A Grubor, A Oroz, S Milenkovic

Citation

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Abstract

We describe a 36-year-old women referred to the ENT department presented with a large schwannoma which gradually increased in size and with a progressive unilateral facial parasthesia. It was detected by clinical and endoscopical examinations also with CT scan and MR imiging. A transoral endoscopic biopsy revealed a benign schwannoma. The patient first underwent a surgical transorally excision for parapharyngeal part of tumor. A massive intracranial component was completely removed by extracranial approach via lateral suboccipital craniotomy. Patient recovered without complication, and the follow-up 12 months later, however, showed no signs of process expansion. Early transoral/extrcranial complete excison can be currative procedure preserving cosmetics. The extent of tumor was such that it required of multidisciplinary planning of otolaryngologist and neurosurgeon.

INTRODUCTION

Schwannoma (neurinoma, neurilemmoma) is solitary tumor of Schwann cell origin. The Schwann cell surrounds peripheral nerve tissue and is believed to originate from neural crest [1]. In the parapharyngeal space, schwannomas may arise from the last four cranial nerves or the autonomic nerves, the vagus being the most common affected nerve [2]. Computed tomography (CT) and/or magnetic resonance imiging (MRI) are critical in assesment of the patients [3]. In this article we describe huge parapharyngeal benign schwannoma with enormous intracranial propagation. We descript a transoral approach to the supero-medial part of the parapharyngeal space combine with lateral suboccipital approach for endocranial tumor component. The clinical, diagnostic and treatment aspects of benign schwannoma will be discussed.

CASE REPORT

A 36-year-old women was admitted to ENT department with an incidentally discovered lesion in the right parapharyngeal space by oropharyngoscopy. Only a history of prior dental examination was revealed as the patient complained of thumbness of the right side of the face. A large, smooth, firm, submucosal swelling with relatevily sharp demarcation is seen medial to the right palatine tonsil extending toward the soft palate with medial displacement of the lateral epipharyngeal wall (Fig. 1).

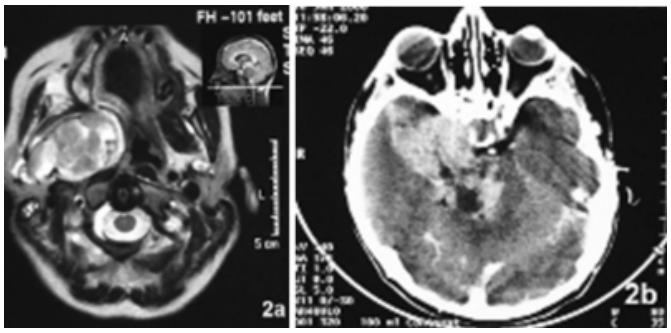
Figure 1

Figure 1

There was no abnormality on physical examination. Central facial nerve palsy was observed and right ear hearing loos was found by pure tone audiometry. Pre-operative (axial) MRI revealed a large homogeneously well-encapsulated mass predominantly in the right parapharyngeal space involving a pterygopalatine fossa (Fig. 2a). It was observed remarkable extension into the endocranium (Fig. 2b).

Figure 2

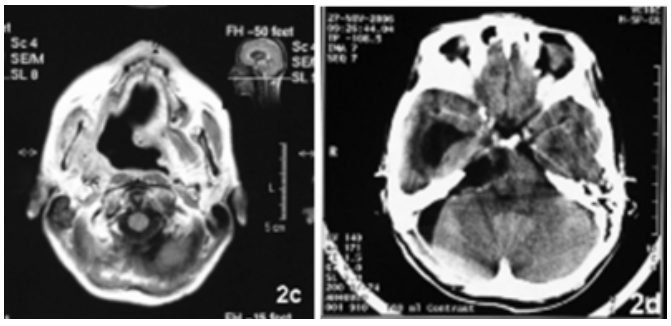
Figure 2a and Figure 2b



An angiogram showed well vascularity, but there were no discplacement of the common carotid artery and the internal jugular vein. As fine-needle aspiration cytology was negative a patient underwent a transoral endoscopic biopsy with final hystologic analysis confirming a benign schwannoma. Before the lesion was removed external carotid artery (ECA) was ligated by external neck approach. Furthermore, around the well exposed internal carotid artery (ICA) was placed loosely vascular loop in non-constricted fashion. Total resection of the parapharyngeal component of lesion was carried out en block with the endocranial nerves monitoring. CT or axial magnetic resonance image demonstrated a gross total resection of the parapharyngeal lesion (Fig. 2c) and endocranial part of tumor was excised via lateral suboccipital craniotomy (Fig. 2d).

Figure 3

Figure 2c and Figure 2d



Following removal, the upper pharyngeal cavity with defect of skull base were packed with Surgicel® and ribbon gauze soaked with antibiotics oitment. The pack was removed after 24 hours. The floor of the anterior fossa was not reconstructed.

Transient disphagia appeared after the operation and nutrition of the patient was controlled with liquid diet. Subseqently a patient swallowing gradually improved with intensive jaws excercize. The symptoms and signs were significantly relieved without neurological sequelae. No evidence of recurrence was noted after one-year follow-up (Fig. 3).

Figure 4

Figure 3

DISCUSSION

Although parapharyngeal schwannoma are not common [4], the tumors with these size and such location could be considered as very rare lesions. The tumors with extracranial extension usually present as asymptomatic mass and often grow to a considerable size before the disease become obvious or clinical detactable [5]. Nevertheless there is no specific sign or symptom that identifies a neurogenic tumor of the parapharyngeal space. Sometimes pain and neurologic deficit could be suggestion of malignancy [6]. In our case tumor was mimikred by the disease of the teeth. The symptom that led patient to seek medical help was cheek parasthesia without neuralgia. Also, schwannoma only very rarely arise from primarily motor nerves [7]. Preoperatively, we could not determined the true nerve origin. More after, the tumor did however, passed through the parapharyngeal space and further spread locally upwards by bony erosion. Inspite of very destructive nature of tumor, no evidence of lymphoadenopathy was found on physical examination.

In making decision of therapeutic management it should be noted that the functional or vital prognosis could be impaired by local extension. The most appropriate surgical approach for large parapharyngeal tumor is the cervical approach, not

only in order to gain control of the large vessels but also to avoid injury of the other nerves in the area [8]. Also, a lesion of the size we present would probably require one stage procedure [9], but a more complex approach (mandibular or zygomatic bone resections) would be increased morbidity and potential for infection or brain herniation sequele [10].

Viewing the patients symptoms due to no raised intracranial pressure and based on the clinical and imiging findings which all suggested benign nature of a tumor (clearly circumscribed, space-occupying lesion) we decided to performed transorally/extracranial surgery. CT of the skull base was helpful for showing the bony destruction and for better orientation, but only the multiplane imaging provided by MRI can display the whole tumor and relations of the structure surrounding the tumor such as blood vessels and nerves.

Some authors believe that a transoral approach is cotraindicated for large paraphraryngeal lesions (more than 3 cm) because of potential risk of hemorrhage, damage of the cranial nerves, tumor spilage and decreased exposure [11]. But in our case, there was no adhesion between the tumor and surrounding tissue as suggested CT scan and MRI, so it was removed relatively easily. The exposure of the tumor further facilitated was easely by resection a part of a hard palate. Sometimes, mucosa retraction might be needded to better exposing intracranial portion of a tumor [12]. Intraoperative monitoring proved no permanent defects during the actual surgical manipulation. No significant complications were related to the approach itself. A visualization was felt to be adequate without need for conversion to a more extensive resection (mandibulotomy). Nevertheless, transorally extraction of tumor without bleeding remained chalenged assesment even after ligated ECA. Dispite a large midline defect we minimized developing of psedoencephalocele and also separated the tumor adequately the inadvertent of CFS leakege could be easely controlled. Intracranially, the large tumor (4 cm in the largest diameter) was predominantly located in PCU region. It was well encapsulated with clear clivage plane and colud be removed completely with no nearby vital structures damages occurred.

CONCLUSION

Clinical decisions of the treatment modalities must include as much quantitative information about clinical status of the patient as possible. We would advocate transoraly/extracranial assessment as safe procedure for complete removing both of parapharyngeal and endocranial section of tumor without morbidity. Although the parapharyngeal part of a tumor was very large as well as intracranial portion, our suggestion is to approach initally transoraly in cases of well encapsulated, medially placed lesions of the parapharyngeal space with benign nature on preoperative imiging.

CORRESPODING AUTHOR

Milan B. Jovanovic, Department of Otorhinolaryngology with Maxillofacial and Cervical Pathology, Clinical Hospital Centre "Zemun", Vukova 9, 11080, Zemun, Serbia e-mail: majov@eunet.yu Tel.: +381-11-3291851

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Author Information

Milan B. Jovanovic, MD, PhD
Department of Otorhinolaryngology with Maxillofacial and Cervical Pathology, Clinical Hospital Centre »Zemun«

Iva Berisavac, MD, PhD
Department of Neurosurgery, Clinical Hospital Centre »Zemun«

Marko Markovic, MD
Department of Neurosurgery, Clinical Hospital Centre »Zemun«

Vlada Bojovic, MD, PhD
Department of Clinical Neurology, Clinical Hospital Centre »Zemun«

Andrej Grubor, MD
Department of Otorhinolaryngology with Maxillofacial and Cervical Pathology, Clinical Hospital Centre »Zemun«

Aleksandar Oroz, MD
Department of Otorhinolaryngology with Maxillofacial and Cervical Pathology, Clinical Hospital Centre »Zemun«

Sanja Milenkovic, MD, PhD
Department of Clinical Pathology, Clinical Hospital Centre »Zemun«