

CASE REPORT

Transoral approach to a huge neurofibroma of the parapharyngeal space: a case report

Parafarengeal bölgenin dev nörofibromasına transoral yaklaşım: Olgu sunumu

Enver ALTAŞ, M.D.,¹ Nesrin GÜRSAN, M.D.,² Harun ÜÇÜNCÜ, M.D.¹

Neurofibroma of the parapharyngeal space accounts for less than 2% of all parapharyngeal space neoplasms. A 38-year-old man presented with complaints of snoring and left-sided nasal obstruction. Medical history also revealed numerous huge skin neurofibromas and multiple café au lait spots all over the body. Pharyngeal endoscopic examination disclosed a firm, nonpulsatile submucosal mass, pushing the left pharyngeal region approximately 5 cm medially, and extending to the inferior part of the epiglottis. The tumor was removed by transoral excision without any subsequent complications. Histological and immunohistochemical examinations revealed a neurofibroma.

Key Words: Neurofibroma/surgery; pharyngeal neoplasms/surgery.

Parafarengeal bölge nörofibromaları bütün parafarengeal bölge neoplazilerinin %2'sinden azını oluşturur. Horlama ve burun sol kısım tıkanıklığı olan 38 yaşında erkek hastanın öyküsünde sütlü kahverengi lekeler ve çok sayıda dev cilt nörofibroması vardı. Farengeal endoskopik muayenede, epiglot alt kenarına kadar uzanan ve sol farengeal bölgeyi mediale doğru yaklaşık 5 cm iten, sıkı yapıda, nabazan vermeyen submukozal dev bir kitle görüldü. Tümöral yapı transoral eksizyonla çıkarıldı ve herhangi bir komplikasyon gelişmedi. Histopatolojik ve immünohistokimyasal incelemede tümörün nörofibroma olduğu görüldü.

Anahtar Sözcükler: Nörofibroma/cerrahi; farengeal neoplaziler/cerrahi.

Peripheral nerve sheath tumors are classified as benign (such as schwannoma, neurofibroma) and malignant (i.e. neurogenic sarcoma). Neurofibroma of the parapharyngeal space may occur as an isolated lesion or as part of von Recklinghausen's disease, an autosomal dominant disorder designated as type I neurofibromatosis.^[1] Neurofibromas of the parapharyngeal space have generally signs and symptoms related to tumor bulk such as nasal obstruction,

snoring, obstructive sleep apnea syndrome, eustachian tube dysfunction, dysphagia, odynophagia, hypoglossal palsy, Horner's syndrome and trismus.^[2-5] We present a patient with nasopharyngeal neurofibroma whose complaint was obstruction of the air passage of the left nasal cavity and snoring.

In general, there are various ways of approaching to the parapharyngeal space masses; transoral and external approaches (i.e. transcervical,

◆ Departments of ¹Otolaryngology and ²Pathology, Medicine Faculty of Atatürk University (Atatürk Üniversitesi Tıp Fakültesi, ¹Kulak Burun Boğaz Hastalıkları Anabilim Dalı, ²Patoloji Anabilim Dalı), Erzurum, Turkey.

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◆ Correspondence (İletişim adresi): Dr. Enver Altaş. Atatürk Üniversitesi Tıp Fakültesi, Kulak Burun Boğaz Hastalıkları Anabilim Dalı, 25240 Erzurum. Turkey. Tel: +90 442 - 236 12 12 Fax (Faks) : +90 442 - 236 13 01 e-mail (e-posta): enveraltas@hotmail.com

transparotid, transmandibular and combined approach). In the literature review, the widely accepted method of removal of the parapharyngeal masses was via an external approach, due to its good exposure. However, recently the transoral excision of parapharyngeal masses has been advocated because of its good exposure. But, transoral approach has various complications (i.e. upper respiratory tract hemorrhage, aspiration of hemorrhagic material into trachea etc.).^[6-9]

CASE REPORT

A 38-year old man was referred to our hospital with a five-year history of snoring and a gradual obstruction of the air passage of the left nasal cavity. His medical history revealed von Recklinghausen's disease with numerous huge skin neurofibromas and multiple cafe-au-lait spots on the whole body. Pharyngeal endoscopic examination disclosed a firm, nonpulsatile submucosal mass, swelling of left pharyngeal region approximately 5 cm medially and from the basal cranium to epiglottis inferiorly. On oropharyngeal examination, the soft palate, uvula, tonsil, and glossopalatine arch were displaced medially because of pressure due to the mass. There were no external palpable neck swelling and other abnormalities detected during

general clinical examination. Hematological and biochemical investigations were all within the normal limits. T₁ and T₂ weighted magnetic resonance imaging (MRI) showed a well defined mass in the left parapharyngeal space without intracranial involvement, swelling of left pharyngeal region approximately 5 cm medially and approaching to epiglottis inferiorly and a well defined high-attenuated, 8x5x4 cm regularly oval shaped mass of fat attenuation was revealed in the left parapharyngeal region. In addition, MRI showed that the surrounding vital structures were not involved (Fig. 1a-c). Also, spindle shaped cells seen on peroral fine needle aspiration cytology suggested a neurofibroma. Thus, the characteristic tumor morphology on aspirates, a long term history, absence of neurological deficit and preservation of soft tissue planes were taken as strongly suggestive of a benign nerve sheath tumor.

Surgical technique

The mass was easily removed after a linear-vertical incision through the overlying mucosa and buccopharyngeal fascia and pharyngeal muscle dissection had been carried out, using a Boyle-Davis gag to allow sufficient exposure by transoral approach under general anesthesia. There was no evidence of

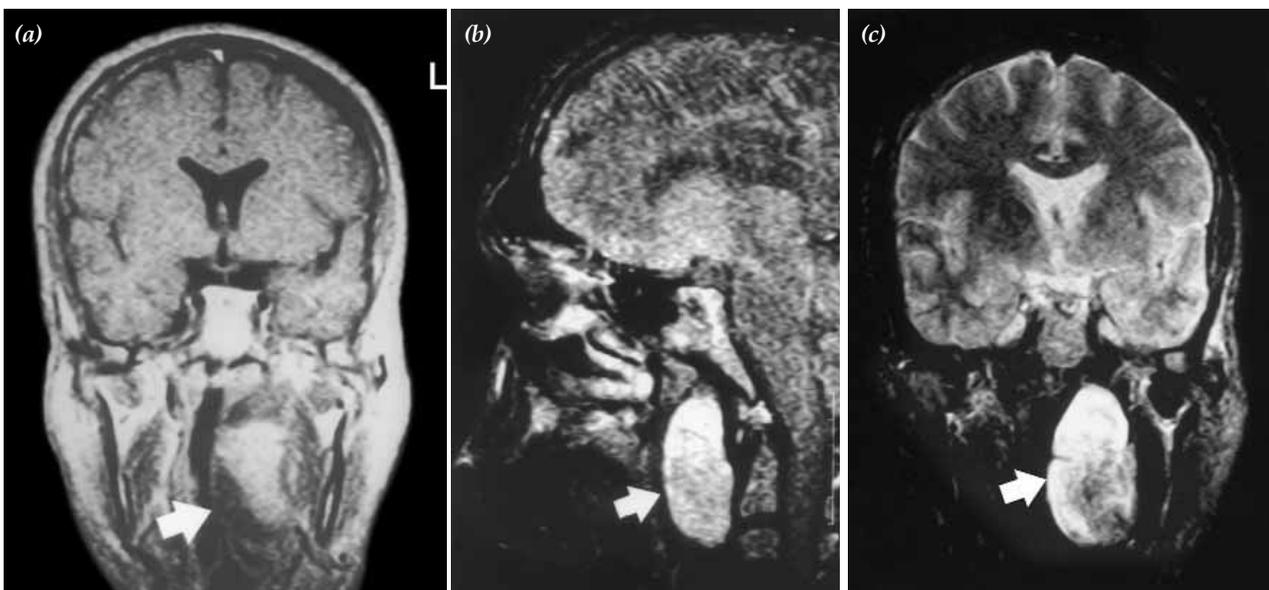


Fig. 1 - (a) T₁ weighed coronal MRI, (b) T₁ weighed sagittal MRI showing a parapharyngeal hypointensed mass and (c) T₂ weighed coronal MRI showing hyperintensed mass. The mass extends from the basal cranium to epiglottis inferiorly [a well defined highly-attenuated, regularly oval shaped mass of fat attenuation and hypovascularisation in the left parapharyngeal region (arrows)].

any attachment to surrounding structures. The mass was removed completely with less than 30 ml of blood loss. Intraoperative hemorrhage was controlled by bipolar electrocoater and no complication was observed. The mucosa and fascia were, then closed with intermittent 3/0 chromic catgut sutures. The gross specimen consisted of a regular, grayish-white spindle shaped solid mass (8x5x4 cm) having a gelatinous appearance on cross-section.

After total resection of the tumor, histopathological and immunohistochemical findings showed that it was a neurofibroma. Microscopic section showed interlacing bundles of elongated cells with wavy and dark-stained nuclei; i.e. the irregular spindle shaped tumor cells and small amount of mucoid material separating the cells and collagen each other. Cells were arranged in fascicles (Fig. 2). Immunohistochemically, the tumor showed immunoreactivity for S-100 protein, NSE and vimentin.

The patient was free from the initial complaints and remained well until the present time. The follow-up examinations at the 3rd, 6th, 12th, and 24th months and MRI check-ups did not reveal any signs of recurrence. We still follow-up the patient.

DISCUSSION

The cases with parapharyngeal neurofibroma have some symptoms and signs like nasal obstruction, snoring, obstructive sleep apnea syndrome,

eustachian tube dysfunction, dysphagia, odynophagia and trismus.^[3,4] Benign tumours rarely cause a neurological deficit as hypoglossal palsy, Horner's syndrome, which suggests a malignancy or jugular foramen involvement.^[5,7] The initial complaints were snoring and nasal obstruction in our case.

The differential diagnosis of a parapharyngeal neurofibroma includes parotid or minor salivary gland tumors, neuromas, glomus tumors, chordoma, soft tissue chondroma, chondrosarcoma, tongue-base hemangioma, solitary fibrous tumor, lymph nodes and lipomatous lesions.^[9-13] The majority of parapharyngeal space masses are benign. In Dankle et al's review,^[11] of 318 parapharyngeal masses, 45% were parotid, 15% were lymph node and 23% were neurogenous in origin, but only 5 cases (1.6%) were neurofibroma and 67 (21%) were malignant masses. Som et al.^[6] reported only 3 neurofibroma cases out of 104 parapharyngeal masses.

The parapharyngeal space with its complex anatomy is of great clinical importance. Because of its location deep within the neck, the space is difficult to examine by ordinary methods, but can be well demonstrated in the axial section by computed tomography and MRI.^[2] In addition, coronal sections are helpful in assessing the superior extent of tumor, base of skull erosion and intracranial extension.^[7] Also, we demonstrated the mass in the sagittal and coronal T₁ and T₂ weighted MRI. Magnetic reso-

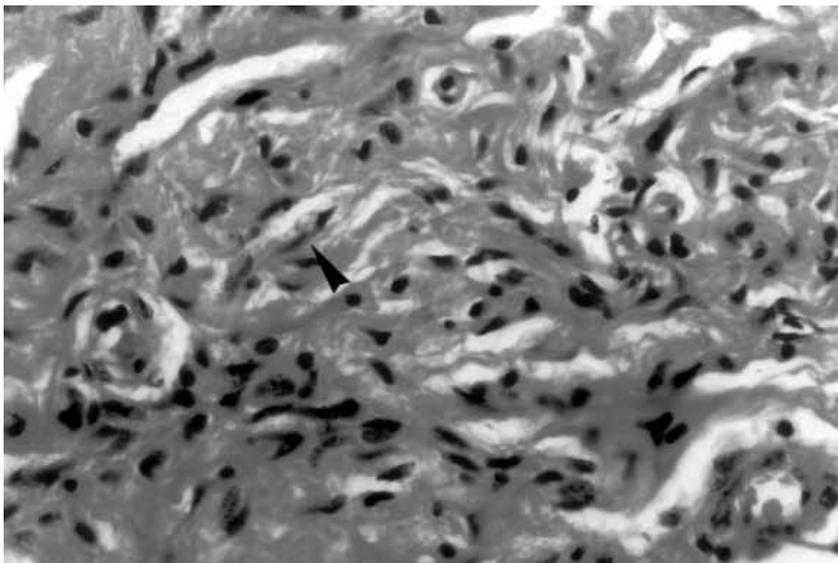


Fig. 2 - Histopathological findings: neurofibroma showing the irregular spindle shaped tumor cells (arrow head) (H-E x 200).

nance imaging showed that the surrounding vital structures were not involved and the mass was hypovascularized (Fig. 1a-c).

A complete resection of the tumor is needed for the treatment of neurofibroma, because neurofibromas may infiltrate extensively. A parapharyngeal space tumor presents a challenge to a surgeon with respect to surgical approach. In general, there are various ways of approaching the parapharyngeal space; through the mouth (transoral), through the neck (transcervical), behind the mandibula (transparotid), through mandibulotomy (transmandibular), and combined techniques. The accepted method of removal of the majority of parapharyngeal masses is via an external approach, in order to gain good exposure.^[6,14] Tandon et al.^[7] presented 15 patients with parapharyngeal neurofibroma operated through transcervical and transmandibular routes. Ten of 15 patients underwent a transcervical excision of the tumor. In only one case, Horner's syndrome developed after the operation. Tumor recurrence was detected in two cases; inadvertent injury to the vagus resulting in vocal cord paralysis. In five patients, a paramedian mandibulotomy was performed to extirpate large tumors (8 cm or more in diameter) extending into the retropharyngeal compartment and to the base of skull. Otherwise, they considered blind dissection in transcervical approach unsafe because of adhesions to the base of the skull or when separation of a medially displaced internal carotid artery was hampered by the rigid lateral wall of the parapharyngeal space. Also, they considered a transcervical approach adequate for tumors not impinging on the base of skull. Belli et al.^[15] presented 6 patients with benign primary tumors of the parapharyngeal space. They performed transmandibular and submandibular approaches, to avoid the risk of injury to the transparotid segment of facial nerve and risk of intraoperative bleeding and difficulties in radical enucleation of transoral approach, to allow an adequate exposure and a direct access to the parapharyngeal space. Dankle^[1] reported 5 patients with glomus intravagale, hemangioma, schwannoma, carotid body tumour, benign mixed tumor of the deep lobe of the parotid gland in 1987. He performed transmandibular approach to extirpate tumors. Postoperative course was uneventful in two cases. But, 3 cases had some complications 2 cases had unilateral vocal cord paralysis and 1 had weak-

ness of the marginal mandibular, hypoglossal, and recurrent laryngeal nerves. However, recently the transoral excision of parapharyngeal masses has been advocated because of its good exposure and little intraoperative hemorrhage.^[8] In our case, we used transoral approach because the surrounding vital structures were not involved and T₂-weighted MRI showed a hyperintensed and hypovascularized mass. The surgery was successful and no complication was observed.

We thought that the transoral approach for removal of a neurofibroma of the parapharyngeal may be an acceptable method in cases in whom the surrounding vital structures are not involved and a hypovascularized mass is present.

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