***Case Report / Olgu Sunumu***

# Excision of recurrent synovial sarcoma of the infratemporal fossa

## Rekürren infratemporal fossa sinovyal sarkom eksizyonu

Fakih Cihat Eravcı, MD.,[1] Mehmet Düzlü, MD.,[1] Mehmet Ekrem Zorlu, MD.,[1]
Metin Yılmaz, MD.,[1] Mustafa Sancar Ataç, MD.[2]

[1]*Department of Otolaryngology, Medical Faculty of Gazi University, Ankara, Turkey*
[2]*Department of Oral and Maxillofacial Surgery, Gazi University Faculty of Dentistry, Ankara, Turkey*

***ABSTRACT***

Synovial sarcoma is a soft tissue sarcoma especially encountered in the lower extremities. The infratemporal fossa is quite a rare location. Since it is a closed location, combined approaches and multidisciplinary planning always need to be considered. This case emphasizes the high-grade character of synovial sarcoma, which causes it to recur often. The difficulty of clear surgical margins in the infratemporal fossa adds to synovial sarcoma a second challenging issue. Therefore, the need of complementary therapy is essential. In this case, we applied postoperative radiotherapy and we did not observe any sign of local, regional or distant metastasis in the one-year follow-up. In this article, we present a 68-year-old male patient together with totally excised synovial sarcoma in the infratemporal fossa by mandibular swing and transzygomatic technique information on the manifestation, imaging, histopathological features and postoperative complications of previous infratemporal fossa synovial sarcomas.

***Keywords:*** Infratemporal fossa; surgical approache; synovial sarcoma.

***ÖZ***

Sinovyal sarkom özellikle alt ekstremitelerde karşılaşılan yumuşak doku sarkomudur. İnfratemporal fossa oldukça nadir bir konumdur. Kapalı bir alan olması nedeniyle kombine yaklaşım ve multidisipliner planlama her zaman dikkate alınmalıdır. Bu olgu sıklıkla tekrarlamasına neden olan sinovyal sarkomun yüksek grade karakterini vurgulamaktadır. İnfratemporal fossada temiz cerrahi sınırın zorluğu, sinovyal sarkoma ikinci bir sıkıntılı sorun eklemektedir. Bu nedenle tamamlayıcı tedavi gereksinimi elzemdir. Bu olguda ameliyat sonrası radyoterapi uygulandı ve bir yıllık takipte lokal, bölgesel veya uzak metastaz izlenmedi. Bu yazıda, 68 yaşında bir erkek hastada mandibüler swing ve transzygomatik teknik ile total olarak eksize edilen infratemporal fossa sinovyal sarkom olgusu, önceki infratemporal fossa sinovyal sarkomların manifestasyonları, görüntüleme ve histopatolojik özellikleri ve ameliyat sonrası komplikasyon bilgileri ile birlikte sunuldu.

***Anahtar Sözcükler:*** İnfratemporal fossa; cerrahi yaklaşım; sinovyal sarkoma.

Synovial sarcoma (SS) is a rare malignancy, commonly derived from the soft tissues of the lower extremities. Head and neck involvement is seen in about 3 to 10% of cases, where the most common sites are the hypopharynx and cervical lymph nodes.[1-4] The infratemporal fossa (ITF) is an extremely rare location. The complex anatomy of the ITF and its closeness to vital structures has long made it a challenge for surgical intervention. We present a case

Available online at
www.kbbihtisas.org
doi: 10.5606/kbbihtisas.2016.09365
QR (Quick Response) Code

Received / *Geliş tarihi:* February 08, 2016 Accepted / *Kabul tarihi:* May 29, 2016

*Correspondence / İletişim adresi:* Fakih Cihat Eravcı, MD. Gazi Üniversitesi Tıp Fakültesi Kulak Burun Boğaz Anabilim Dalı, 06500 Beşevler, Ankara, Turkey.
Tel: +90 312 - 202 64 26 e-mail *(e-posta):* fceravci@gmail.com

of recurrent SS located in the ITF managed through transmandibular and transzygomatic approaches and review the literature.

## CASE REPORT

A 68-year-old man complaining of pain and a burning sensation in his left temporomandibular joint with impairment of mouth opening and closing was referred to our Otolaryngology Head and Neck Surgery Department from the Oral and Maxillofacial Surgery Department of Dentistry Faculty. As we learned from his past medical history, he had undergone surgery for a mass medial to the zygomatic bone 22 years previously. The definitive pathology report was malignant mesenchymal tumor. In the succeeding 10 years he had undergone five additional minor surgeries for recurrent subdermal preauricular masses. Physical examination revealed left frontal muscle paralysis, restriction of mouth opening and an incision scar in the temporal area from previous operations. Examination of the cranial nerves was all within normal limits except for the frontal branch of the facial nerve. No lymphadenopathy was palpated on neck examination. He underwent maxillofacial magnetic resonance imaging (MRI) with the suspicion of tumor recurrence. MRI revealed a 52x41x49 mm solid lesion located in the left infratemporal fossa with smooth margins and lobular contours (Figure 1). He underwent positron emission tomography-computed tomography (PET-CT) imaging for regional lymph nodes and systemic involvement, with no sign of regional or distant metastasis. For accurate diagnosis, an incisional biopsy was performed after a non-diagnostic fine needle aspiration biopsy result.

With written informed consent, a transmandibular and transzygomatic combined approach was utilized to enhance exposure (Figure 2). After a mandibular swing, the lobular mass located in the left pterygomaxillary fossa was observed transorally. In order to see the superior border of the mass, the miniplates which were fixed over the zygomatic process in a previous operation, were detached (Figure 2) for a transzygomatic approach. With these combined techniques the mass was dissected from surrounding structures and total excision was accomplished. Later, the miniplates over the zygomatic process were repositioned, the mandible was reapproximated and the operation was concluded. The definitive pathology revealed SS with spindle cells (Figure 3). Immunohistochemical studies showed epithelial membrane antigen (EMA) and vimentin positive staining. Pankeratin, S-100 and P63 markers for differential diagnoses showed a negative result. Two weeks after the operation, an adjuvant radiotherapy protocol was applied. A total of 6000 centigray (cGy) intensity modulated radiation therapy (IMRT) was applied to the location of the tumor bed

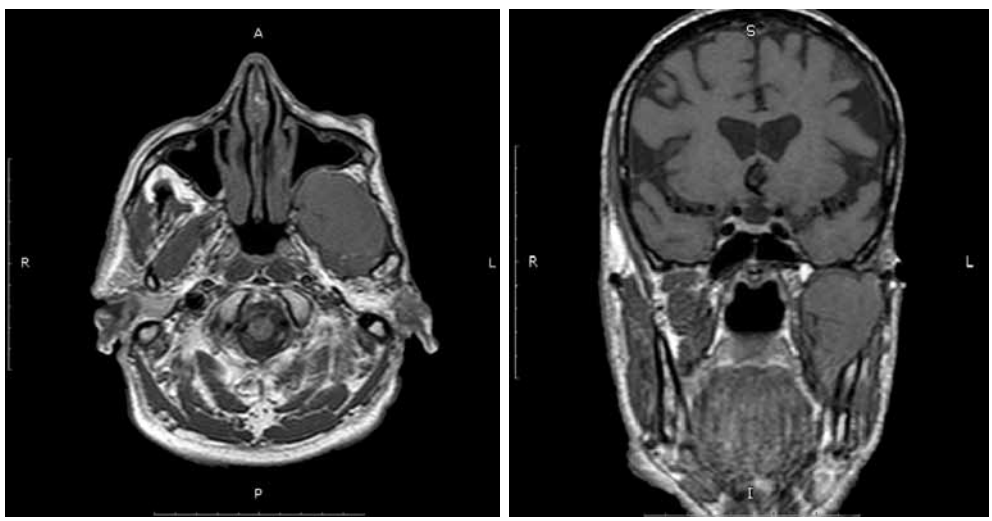


***Figure 1.*** *Preoperative infratemporal fossa mass on axial and coronal magnetic resonance imaging sections.*

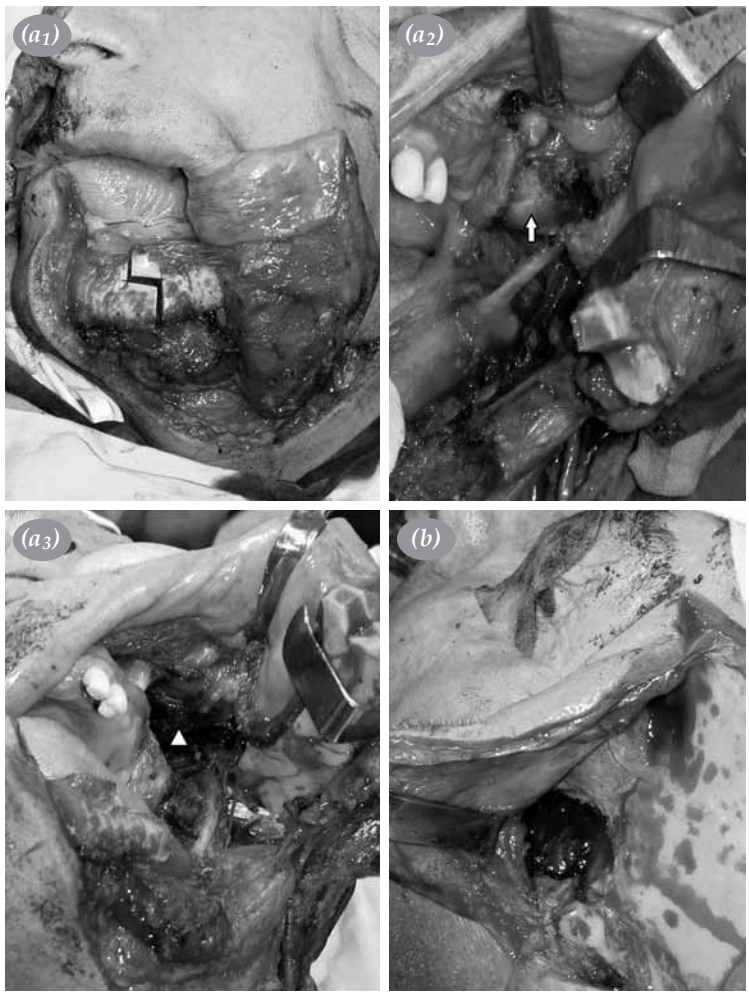


***Figure 2.*** **(a)** *Mandibular swing technique* ***(a1)*** *mandibular cut,* ***(a2)*** *infratemporal fossa mass before dissection (arrow),* ***(a3)*** *infratemporal fossa after excision of the tumor (arrow head) and* ***(b)*** *transzygomatic surgery technique.*

and neck for thirty days. After the RT protocol, an orocutaneous fistula developed in the submental area. This was closed with extraction of the miniplate in the mandibular swing area and antibiotic treatment. There was no sign of recurrence on MRI on one-year follow-up (Figure 4).

## DISCUSSION

The close relation of the infratemporal fossa to vital structures and its concealed location has made it a challenge for a long period. Surgical approaches have enriched the understanding of the anatomy and provided improvement in technology. With the widespread use of imaging studies, incidental cases identified at a small size have increased and with the use of endoscopic modalities the treatment options have moved to a higher level. Because of the concealed localization, all the surgical approaches defined to date are worthwhile and in almost all cases combined approaches are required. Total excision with acceptable morbidity and prolonged survival is possible using diverse approaches and combinations specific to the case.[1]

The ITFs surrounding bony limits are defined superiorly by the sphenoid greater wing and squamosal part of the temporal bone, medially by the sphenoid bones and lateral pterygoid lamina, anteriorly by the maxilla posterior wall, laterally by the zygomatic arch and ascending ramus of the mandible. The ITF connects to the

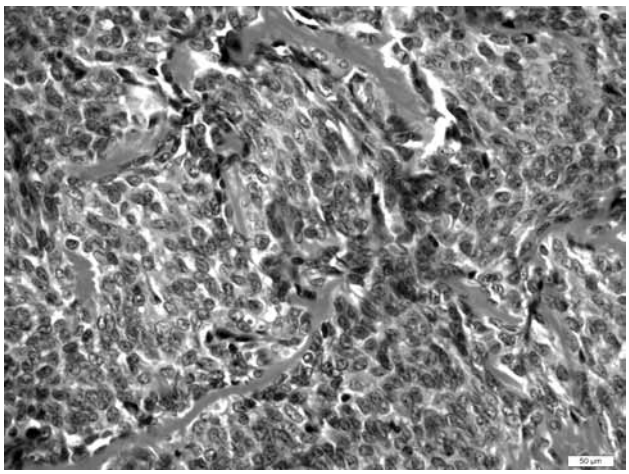

***Figure 3.*** *Definitive pathology: malignant spindle cells, split by collagen bands (H-E x 40).*

temporal fossa superolaterally and connects to the parapharyngeal area posterior to the medial pterygoid muscle. The internal jugular vein and internal carotid artery form the posterior limit and the pterygoids and temporalis muscle are inside the ITF. Added to this are the trigeminal nerve maxillary and mandibular branches and the maxillary artery running in this location.[2]

The most common presentation is swelling and asymmetry in the face, followed by trismus, pain (ear, face), visual problems, conductive hearing loss and epistaxis. Reported symptoms in the ITF SS mimicking other ITF masses include painless cheek swelling, local pain, restriction of mouth opening and migraines. The most importantly encountered symptom is local pain, as seen in our case.[2,3] If not identified incidentally, lesions of large size can be encountered. While our tumor size was 47 mm, the mean tumor size in the literature is 5.79 cm. Intracranial extensions with cranial nerve deficits can be seen because of their proximity. In these cases, the critical point is involvement of the cavernous sinus. Computed tomography and MRI give important information about bone erosion, muscular invasion and lesion type. In our case, fine needle aspiration cytology (FNAC) gave no definitive diagnosis preoperatively. Although ultrasonography-assisted fine needle aspiration biopsy is effective with ITF masses, only one of five ITF SS are diagnosed with FNAC. Therefore, preoperative FNAC use is controversial.[2]

The ITF is often affected by masses derived from neighboring locations, such as the nasopharynx, parotid gland, middle cranial fossa and external auditory canal. Secondly, primary tumors of the region and metastases are rarely seen.[2] Histologically benign and malignant tumors are encountered. The types of tumors make up a long list, including adenoid cystic tumor, meningioma, benign and malignant peripheral nerve tumors, lipoma, hemangiopericytoma, giant cell tumor, relapsing benign parotid tumors, histofibroma, rhabdomyosarcoma, fibrosarcoma and SS.[4-8] Infratemporal fossa metastatic lesions noted in the literature are malignant meningioma, adrenal tumor and papillary thyroid cancer.[1]

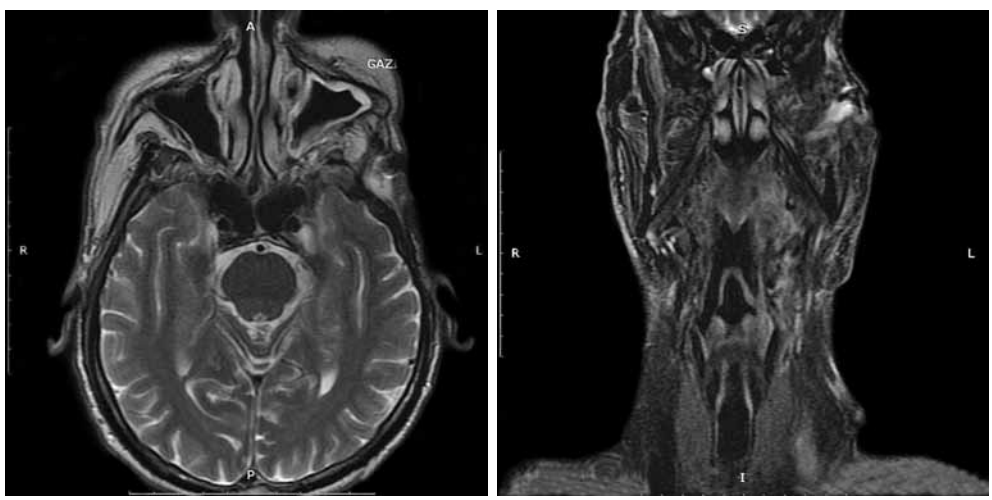


***Figure 4.*** *First year postoperative magnetic resonance imaging.*

Synovial sarcoma is a rare cancer type, which is primarily derived from the soft tissues of the lower extremities. Due to the high rate of local recurrence, distant metastases and rapid progression, treatments are challenging. Recurrence affecting the head and neck is seen in about 3 to 10% of cases, where the most common sites are the hypopharynx and cervical lymph nodes. The ITF is an extremely rare localization. Tumors are not derived from synovial cells but from periarticular mesenchymal tissues. Just like benign lesions, the SS growth pattern can be expansive with smooth boundaries. When imaging studies reveal septations, hemorrhages, calcification and cystic components, SS must come to mind. Previous ITF SS imaging studies show heterogeneous mass and septations, as in our case. Added to this there are examples of infiltration to the surrounding soft tissue, bony structures and foramen ovale extensions.[6] Similar to previously reported cases, we did not see lymphadenopathy in the neck.

To the best of our knowledge, there have been eleven case reports of ITF SS in the English literature to date. Because of their rare occurrence, all the cases in the literature need to be considered to understand the clinical behavior and management of the tumor. Although the ITF SS reports were predominantly in women, our present case was in a man. Presented ITF SS cases show a mean age of 39.7 years, while in our case it was first determined at 46 years old.[6]

The prognosis can vary depending on the stage and histological type at the time of diagnosis. In contrast to other sarcomas, SSs consist of carcinoma-like epithelial cells and fibrosarcoma-like spindle cells. Two cell components of SS are categorized according to the differentiation stage and the relative predominance of these characteristics. The histological patterns are biphasic, monophasic epithelial, monophasic fibrous and undifferentiated forms. While the epithelial and spindle component are seen together in biphasic SS, if the spindle component is seen alone it is defined as a monophasic fibrous type while if the epithelial cell group is seen alone it is defined as a monophasic epithelial type. Mostly the monophasic fibrous type and biphasic type are encountered. Our case consisted solely of a spindle component and was thus categorized as a monophasic fibrous type. A review of ITF SS cases shows four cases of biphasic SS, three cases of monophasic SS and one unspecified case, while the others were not reported.[6] In addition, concerning immunohistochemical analysis at diagnosis: epithelial markers EMA and cytokeratin, mesenchymal marker CD99 and vimentin can be determined and confirmed by histopathologic analysis. In our case, immunohistochemical studies showed EMA and vimentin positive staining. For differential diagnosis, fibrosarcoma, squamous cell cancer and malignant nerve sheet tumors must be kept in mind. Pankeratin, S-100 and p63 markers were studied for differential diagnosis and showed negative results.

Although there is no consensus on treatment approach, suggestions are total surgical resection complemented with radiotherapy, chemotherapy or combined therapy. There is no optimal and standard management for head and neck SSs so these case reports are useful for better management. Prognoses of soft tissue sarcomas depend on negative resection margins and the SS five-year survival rate is about 57%. With head and neck involvement, the survival rate increases to 72% but depends on location and size. Total excision of ITF located SS is challenging because of vital structures, and additive treatments are required.[4-6]

Along with understanding the anatomy, various surgical approaches developed for ITF masses recommend total excision with maximal exposure and minimal morbidity. If needed, these approaches can be combined. Multidisciplinary collaboration is needed pre-, peri- and postoperation when intracranial, intraorbital, or temporomandibular extensions are present. Although it has the disadvantage of sacrificing hearing and causing facial weakness, Fisch's contribution is helpful in combining the experience of otologic and neck procedures. A combination of the preauricular transzygomatic and transmandibular approach with a facial degloving approach will maximize exposure for excision. Additionally, coronoid process excision can contribute to further exposure as needed. The maxillary swing procedure, which Wei et al. first defined, is an option especially for reaching masses that extend to parapharengeal and oro-

nasopharengeal locations.[9-13] Midfacial swing can be utilized to reach midline structures such as clivus masses. Orbital swing is indicated for retrobulbar tumors inferior to the orbital nerve and superior clivus tumors.[9,12] Subtemporal, intratemporal and orbitozygomatic approaches are useful for superior settled ITF tumors.[14-17]

Although the endoscopic option must always be on hand to combine as needed, it also has contraindications for ITF lesions, such as massive intracranial extension, masses surrounding the internal carotid, or widespread to the masticator space. It is harder to expose and reach the lateral masses using endoscopy with ITF localization. Main vascular bleeding, facial numbness, dry eye syndrome, permanent conductive hearing loss resulting from glue ear, and temporary mastication problems can be encountered after endoscopic surgery. Instead of considering this a conservative and less radical option, it should be considered a complementary option.[18] Though the endoscopic approach is used effectively for sinonasal tumors,[19] ITF extension restricts its utilization. The probability of utilizing open surgical approaches is dominant for ITF masses, especially for total resection.

With ITF tumors, in spite of total dissection, vital structures limit the free margin of excision. Therefore, recurrences commonly occur, and postoperative treatment is needed in most cases.[11] In our case, because of recurrence and ITF localization, adjuvant radiotherapy was applied.

Cranial cerebrospinal fluid leak, temporary facial paralysis, bleeding, hematoma and abscess can be seen postoperatively.[1] In our case local healing impairment occurred after adjuvant therapy.

In conclusion, with ITF location all defined surgical approaches should be weighed, and in contrast to the high number of options, a free margin is still challenging. Because of the free margin problem and high recurrence rate in ITF SS, complementing therapies must be considered.

**Declaration of conflicting interests**

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

**Funding**

The authors received no financial support for the research and/or authorship of this article.

## REFERENCES

1. Givi B, Liu J, Bilsky M, Mehrara B, Disa J, Pusic A, et al. Outcome of resection of infratemporal fossa tumors. Head Neck 2013;35:1567-72.
2. Tiwari R, Quak J, Egeler S, Smeele L, Waal IV, Valk PV, et al. Tumors of the infratemporal fossa. Skull Base Surg 2000;10:1-9.
3. Bryant L, Goodmurphy CW, Han JK. Endoscopic and three-dimensional radiographic imaging of the pterygopalatine and infratemporal fossae: improving surgical landmarks. Ann Otol Rhinol Laryngol 2014;123:111-6.
4. Nomura F, Kishimoto S. Synovial sarcoma of the temporomandibular joint and infratemporal fossa. Auris Nasus Larynx 2014;41:572-5.
5. Gheisari R, Roozbehi A. Malignant peripheral nerve sheath tumor of the infratemporal fossa. J Craniofac Surg 2010;21:596-8.
6. Wushou A, Zhao YJ, Shao ZM. Synovial sarcoma of the infratemporal fossa: A case report. Oncol Lett 2014;8:2165-70.
7. Dhawan A, Shenoy AM, Chavan P, Sandhu S, Sriprakash D. Synovial sarcoma of the infratemporal fossa with extension into the oral cavity--a rare presentation and literature review. J Oral Maxillofac Surg 2012;70:2923-9.
8. Jeyanthi K, Karthikeyan R, Sherlin HJ, Anuja N, Ramani P, Priya P, et al. Pleomorphic adenoma in the infra-temporal space: the first case report. Head Neck Pathol 2007;1:173-7.
9. Moreira-Gonzalez A, Pieper DR, Cambra JB, Simman R, Jackson IT. Skull base tumors: a comprehensive review of transfacial swing osteotomy approaches. Plast Reconstr Surg 2005;115:711-20.
10. Maheshwari GU, Chauhan S, Kumar S, Krishnamoorthy S. Access osteotomy for infratemporal tumors: Two case reports. Ann Maxillofac Surg 2012;2:77-81.
11. Chatni SS, Sharan R, Patel D, Iyer S, Tiwari RM, Kuriakose MA. Transmandibular approach for excision of maxillary sinus tumors extending to pterygopalatine and infratemporal fossae. Oral Oncol 2009;45:720-6.
12. Roh JL. Removal of infratemporal fossa schwannoma via a transmandibular transpterygoid approach. Eur Arch Otorhinolaryngol 2005;262:428-31.
13. Otremba M, Adam S, Omay SB, Lowlicht R, Bulsara KR, Judson B. Maxillary swing approach for extended infratemporal fossa tumors. Laryngoscope 2013;123:1607-11.
14. Honda K, Asato R, Tanaka S, Endo T, Nishimura K, Ito J. Vidian nerve schwannoma with middle cranial fossa extension resected via a maxillary swing approach. Head Neck 2008;30:1389-93.
15. Amin AA. Maxillary swing approach for surgical resection of recurrent nasopharyngeal tumors. J Egypt Natl Canc Inst 2007;19:219-23.
16. Guo Y, Guo C. Maxillary-fronto-temporal approach for removal of recurrent malignant infratemporal

fossa tumors: Anatomical and clinical study. J Craniomaxillofac Surg 2014;42:206-12.
17. Timoshenko AP, Asanau A, Gavid M, Colin V, Martin C, Prades JM. Preauricular transmandibular and transzygomatic approach for tumors of the infratemporal fossa revisited. ORL J Otorhinolaryngol Relat Spec 2013;75:250-5.
18. Battaglia P, Turri-Zanoni M, Dallan I, Gallo S, Sica E, Padoan G, et al. Endoscopic endonasal transpterygoid transmaxillary approach to the infratemporal and upper parapharyngeal tumors. Otolaryngol Head Neck Surg 2014;150:696-702.
19. Forer B, Lin LJ, Sethi DS, Landsberg R. Endoscopic Resection of Sinonasal Tract Schwannoma: Presentation, Treatment, and Outcome in 10 Cases. Ann Otol Rhinol Laryngol 2015;124:603-8.