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Case Report / Olgu Sunumu

Synovial sarcoma of the infratemporal fossa with intracranial extension

İntrakraniyal uzanım gösteren infratemporal fossanın sinoviyal sarkomu

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Synovial sarcomas (SS) account for 7-8% of soft-tissue cancers and 3-5% of all cases with head and neck involvement. Synovial sarcoma of the infratemporal fossa is very rare In this article, we report the fourth case of SS of infratemporal fossa and the first case with intracranial extension via the foramen ovale. A 31-year-old man admitted with a one-year history of intense pain in his right jaw. On physical examination, there was only hyperesthesia over the right mandible side. Computed tomography (CT) and magnetic resonance imaging (MRI) showed a mass in the infratemporal fossa and intracranial extension from the foramen ovale. The mass was surgically removed *en bloc*. Postoperative pathological examination reported the mass as a biphasic-type synovial sarcoma. The patient who received postoperative chemoradiotherapy had no recurrent disease for one year. The patient is still being followed in our clinic.

Key Words: Fossa infratemporalis; surgery; synovial sarcoma.

Sinoviyal sarkomlar (SS) yumuşak doku kanserlerinin %7-8'ini, baş ve boyun tutulumu olan kanserlerin ise %3-5'ini oluşturur. İnfratemporal fossanın sinoviyal sarkomu çok nadirdir. Bu yazıda, infratemporal fossanın dördüncü SS'si ve foramen ovale yoluyla intrakraniyal uzanım gösteren ilk olgusu sunuldu. Otuz bir yaşında erkek hasta bir yıldır devam eden çenenin sağ yarısında şiddetli ağrı yakınması ile başvurdu. Fizik muayenede mandibula sağ korpusunda yalnızca hiperestezi bulgusuna rastlandı. Bilgisayarlı tomografi (BT) ve manyetik rezonans görüntülemede (MRG) infratemporal fossadan kaynaklanan ve foramen ovale yoluyla intrakraniyal uzanım gösteren bir kitle tespit edildi. Kitle ameliyatla *en blok* olarak çıkartıldı. Ameliyat sonrası yapılan patoloji incelemesi, bifazik tip sinoviyal sarkom olarak bildirildi. Hastaya ameliyat sonrası kemoradyoterapi uygulandı ve bir yıllık takipte hastada herhangi bir nüks bulgusuna rastlanmadı Hasta halen kliniğimizde takip edilmektedir.

Anahtar Sözcükler: İnfratemporal fossa; cerrahi; sinoviyal sarkom.

Synovial sarcoma (SS) is a rare malignant soft-tissue tumor usually seen in the extremities of adults and typically over an age range of 14 to 35 years.[1] Only 3-5% of SS is located in the head and neck region, where it can present as a neck mass, hoarseness, dysphagia, and dyspnea.[2] Synovial sarcoma of

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the head and neck has a better prognosis than of the extremities. Despite multiple therapeutic approaches, the overall five-year survival rates of synovial sarcoma of the head and neck ranges from 47 to 82%.[3] Local recurrence occurs in 21-56%.[4] Cases of synovial sarcoma have been reported in the hypopharynx, larynx, parotid gland, sinuses, oral cavity, thyroid gland, and retropharyngeal space.[5] Although synovial sarcoma has been reported in the infratemporal fossa, no case had intracranial extension. Here, we report the first synovial sarcoma of the infratemporal fossa extending to the cranium via the foramen ovale.

CASE REPORT

A 31-year-old man presented with intense pain in his right jaw that had been present for one year. His pain progressed after facial injury in a traffic accident nine months earlier. On physical examination, there was hyperesthesia over the right mandibular body, but no other finding such as a sensory deficit or palpable mass.

Computed tomography (CT) showed a 21x23x31 mm, smooth-margined, calcified mass on the medial side of the right temporomandibular joint (TMJ) near the glenoid fossa, extending to the infratemporal fossa (Figure 1a, b). This caused a 3-4 mm defect in the skull base that extended to the intracranial area (Figure 1c) The differential diagnosis included synovial sarcoma, atypical meningioma, and neurogenic tumors.

Magnetic resonance imaging (MRI) revealed a 33x25x19 mm calcified mass, medial to the right TMJ, extending to the infratemporal fossa causing a defect in the cranial bone and abutting the dura mater. The cerebral parenchyma was normal. The mass showed signal intensities similar to the gray matter on T_1- and T_2-weighted MRI and heterogeneous enhancement after contrast medium injection (Figure 2a-c) The initial diagnosis was an extracranial meningioma or chondroid lesion. No biopsy could be performed because of the location of the mass.

At surgery, a Montgomery incision was made and the facial nerve branches were identified and preserved. The deep lobe of the parotid gland was dissected from the angle of the mandible and the masseter was identified inferiorly and the TMJ capsule superiorly. The parapharyngeal area was observed clearly and the mass was located medial to the mandibular condyle. The deep lobe of the parotid was pulled aside with an ecarteur (Kocher-Langenbeck K452.05. Bahadır A.Ş. Samsun, Türkiye) and the mass was dissected and removed. No nerve or vessel damage was seen during dissection. Subsequently, microscopic examination of the midline skull base showed destruction of a 0.5x0.5 cm area of bone posterolateral to the foramen ovale, although the dura was intact. There was no cerebrospinal fluid leakage. Surgicel was applied to this area and the operation was completed.

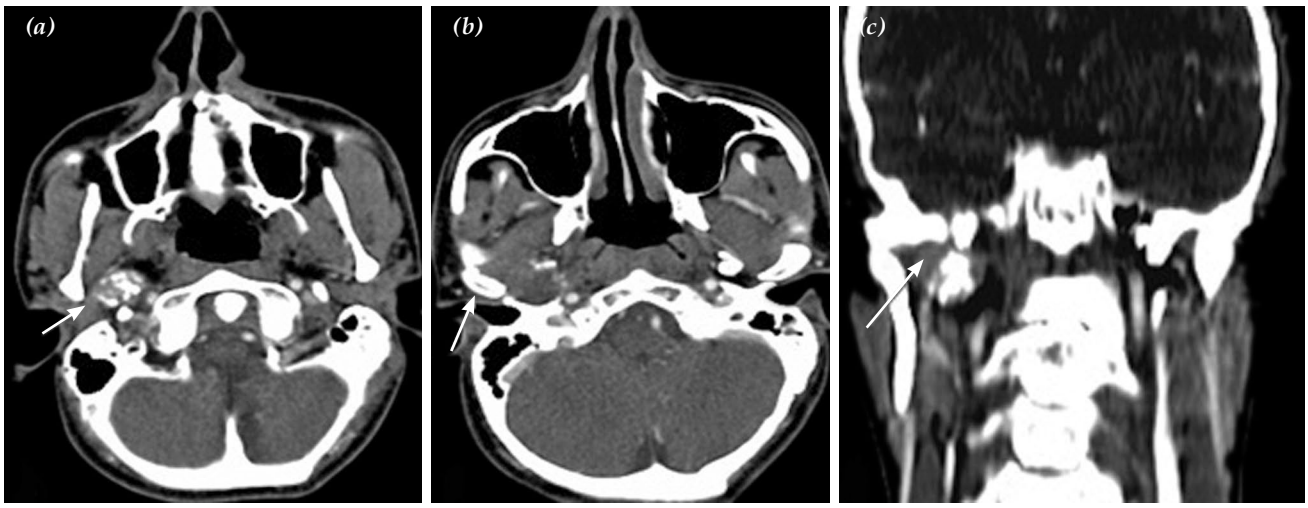

Figure 1. *Computed tomography.* ***(a)*** *Axial section after contrast injection shows a mass containing punctate calcifications in the right parapharyngeal space posteriorly, with a well-defined margin and average enhancement with the contrast medium (arrow).* ***(b)*** *The mass is near the right temporamandibular joint (arrow), displacing the mandibular head laterally.* ***(c)*** *Coronal multiplanar reconstruction sections show destruction of the bone of the middle cranial fossa and an indentation in the intracranial area (arrow).*

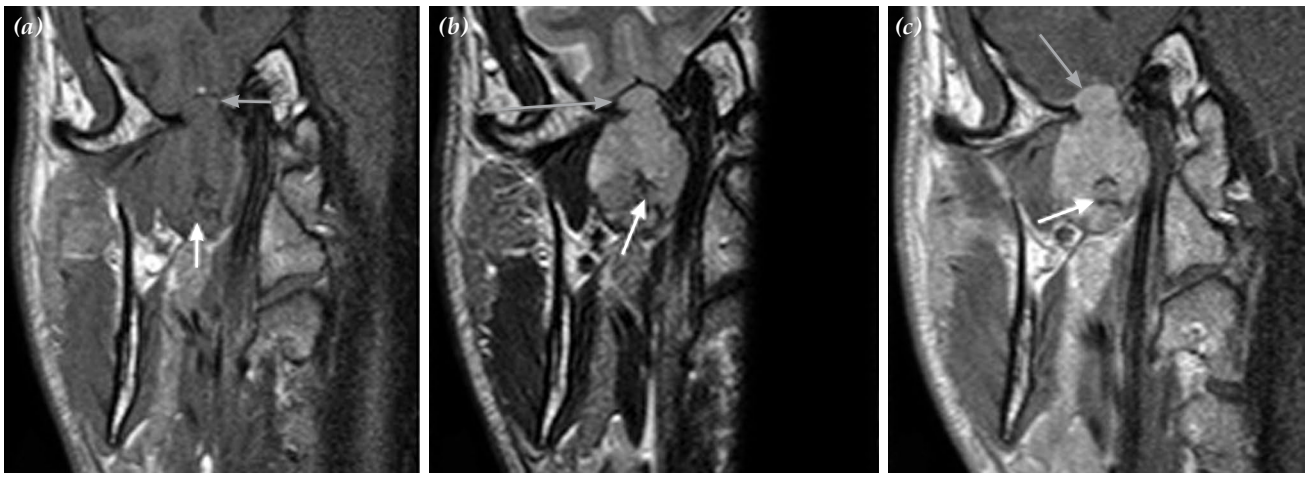

Figure 2. *Coronal magnetic resonance imaging.* ***(a)*** *T_{1a} sequences show a smooth mass isointense to the gray matter, with a halter configuration because of the extracranial component (black arrow).* ***(b)*** *T_{2a} sequence.* ***(c)*** *The mass was enhanced after contrast injection (black arrow). The calcified areas in the mass show low signal intensities in all views (white arrows).*

The postoperative pathology report was that the mass was a biphasic-type synovial sarcoma. The patient was referred to the head and neck tumor council postoperatively and had chemoradiotherapy postoperatively. Postoperatively, House-Brackmann grade 2 facial palsy was observed. On the fifth postoperative day, electroneurography (ENoG) showed 80% dysfunction and suggested that the palsy was caused by edema affecting the facial nerve. Electromyography (EMG) reported total denervation. Two months postoperatively, the facial palsy had disappeared and the EMG was normal.

One month postoperatively, MRI showed no mass in the right infratemporal fossa or medial to the TMJ. There was fluid and air in these areas, seen as slightly hyperintense in T_1-weighted images and bright hyperintense in T_2-weighted images. No residual mass was seen (Figure 3a-c).

The patient is still being followed one year postoperatively and there is no evidence of recurrence. No mass was seen on follow-up MRI year postoperatively (Figure 4).

Histologically, the tumor consisted of fibroblast-like spindle cells and epithelial cells arranged in solid nests, with pseudoglandular formation lined by flattened epithelioid cells (Figure 5, 6). The spindle cell component consisted of plump

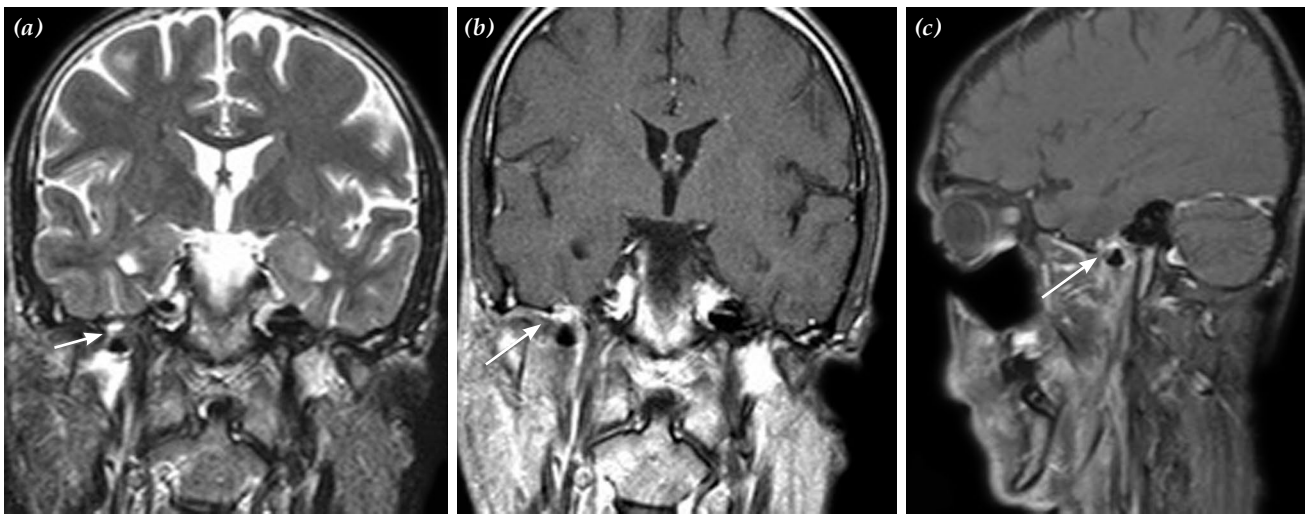

Figure 3. *Postoperative magnetic resonance imaging.* ***(a)*** *Coronal T_{2a} image.* ***(b)*** *Coronal T_{1a} post-contrast injection, and* ***(c)*** *Sagittal section post-contrast injection. All of the images show total excision of the mass, but focal thickening and contrast enhancement of the dura mater (arrows).*

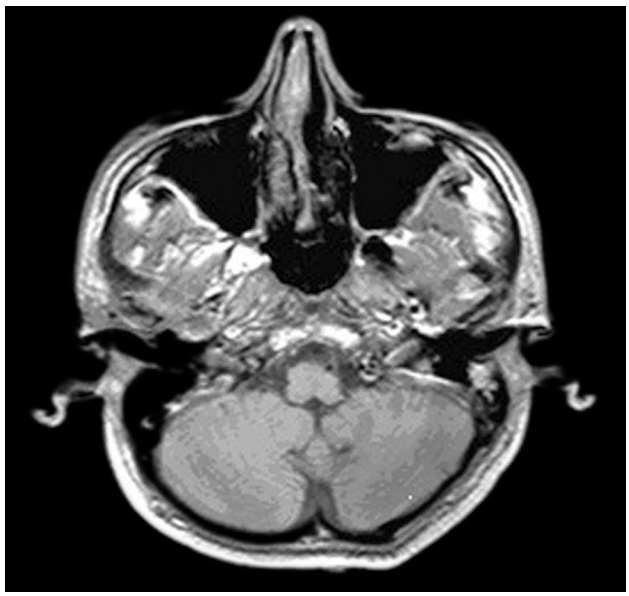

Figure 4. *Postoperative magnetic resonance imaging one year. No mass reported.*

spindle-shaped cells with a uniform appearance in a wavy or short fascicular pattern. The epithelial markers pan-cytokeratin and epithelial membrane antigen emphasized the structures of the epithelial component, whereas the spindle cells showed only rare, focal staining (Figure 7).

DISCUSSION

Synovial sarcoma constitutes 7-8% of soft tissue cancers and only 3-5% of these occur in the head and neck. Manifestations depend on the location and size of the tumor, but most commonly include hoarseness, dyspnea,

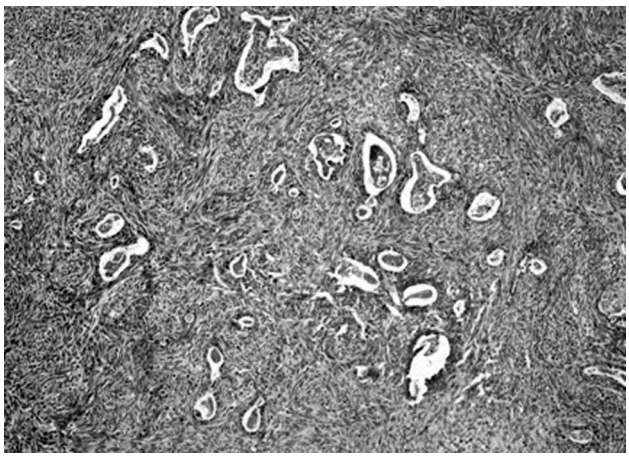

Figure 5. *Biphasic synovial sarcoma with prominent intraluminal eosinophilic secretions in epithelial structures (H-E x 100).*

dysphagia, neck mass, and dysphonia when it occurs in larynx, hypopharynx, or neck region, and with dysesthesia or hyperesthesia in the C3 dermatome when it arises in the infratemporal fossa, because of the proximity of the mandibular nerve exiting the foramen ovale.[2-5] Our patient presented with hyperesthesia over the right mandibular body.

Synovial sarcoma can occur at any site, but is frequently located near joints.[1] It does not originate from synovial tissue,[6,7] but from pluripotent mesenchymal cells near to or even remote from articular surfaces, tendons, tendon sheaths, juxta-articular membranes, and facial aponeuroses.[8] These lesions arise from malignant degeneration

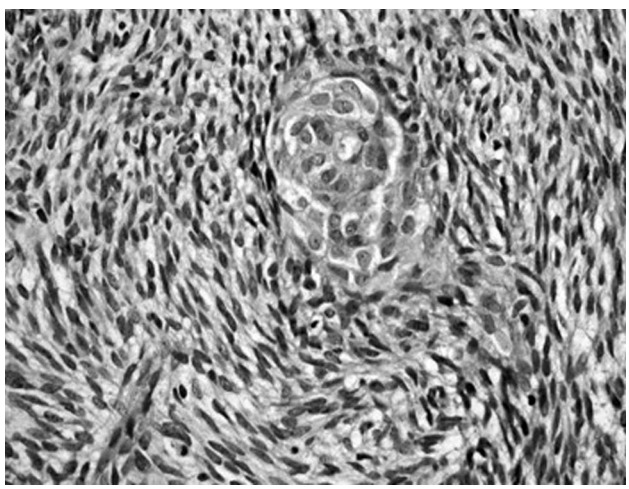

Figure 6. *High-magnificaton view of relatively uniform spindle cell component with pseudo-glandular epithelial structure (arrow) (H-E x 400).*

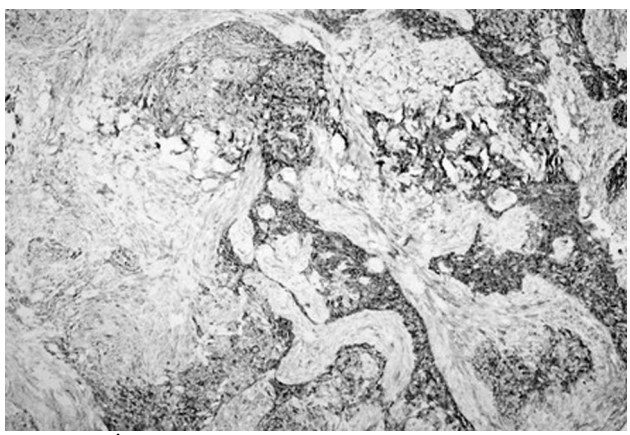

Figure 7. *İmmunohistochemically, the epithelial elements are strongly positive for epithelial membrane antigen, whereas focal immunoreactivity was observed in the spindle cell component (immunohistochemistry for EMA x 100).*

of these primitive mesenchymal cells.[9] In our case, the mass was near the TMJ, with no invasion or destruction of the joint.

Synovial sarcoma has monophasic and biphasic patterns, involving epithelial and spindle cells.[10] The monophasic pattern is subdivided into the monophasic spindle and epithelial cell types.[1] The tumor tissue consists of sarcomatous stroma and glandular space. Synovial sarcomas may be mistaken for fibrosarcomas when fibrosarcomatous spaces predominate, and they may be confused with metastatic adenocarcinoma when glandular spaces predominate.[11] Sometimes it can be difficult to distinguish the monophasic type from other sarcomas, such as fibrosarcomas, malignant schwannomas, hemangiopericytomas, and leiomyosarcomas.[12] The tumor in our case contained fibroblast-like spindle cells and epithelial cells arranged in solid nests, with pseudoglandular formation, lined by flattened epithelioid cells, and the spindle cell component consisted of plump spindle-shaped cells of uniform appearance in a wavy or short fascicular pattern.

In reported imaging studies, synovial sarcoma ranged from to 2 to 9 cm.[6,13] Some authors have argued that the prognosis depends to the size of the tumor, and is better if the tumor is less than 4 cm in diameter.[14] The tumor in our case measured 21x23x31 mm on CT and 33x25x19 mm on MRI.

Classically, CT shows a multilocular tumor with smooth margins and heterogeneous enhancement after contrast injection.[6] The tumor may contain calcifications, hemorrhage, and necrosis. Calcification indicates a better prognosis in synovial sarcoma.[15] The tumor in our case was a smooth-margined, punctate-calcified mass arising from the infratemporal fossa and extending to the cranium on CT.

Magnetic resonance imaging revealed the synovial sarcoma to be of intermediate intensity on T_1-weighted sequences and variable intensity on T_2-weighted sequences, with heterogeneous enhancement after contrast injection. Hemorrhage, fluid-fluid levels, and cyst formation are also features of this disease.[16] On MRI, we observed a calcific mass that showed signal intensity similar to the gray matter in T_1- and T_2-weighted MRI, and heterogeneous enhancement after contrast injection.

Surgery is the main therapy for synovial sarcoma, but can be supplemented with chemotherapy and radiotherapy.[17] Different approaches have been reported, such as the trans-orbitozygomatic approach with a mandibular osteotomy.[18] We used a transparotid approach and did not need to perform a mandibular osteotomy to approach the midline skull base. Previous articles suggest removing the mandibular condyle and TMJ.[18] We did not observe any invasion or destruction, so we preserved all bone structures.

Postoperative radiotherapy has been advocated to improve local control rates in the head and neck; adjuvant chemotherapy may prevent or delay the occurrence of distant metastases.[19] Local recurrence occurs in up to 80% without radiotherapy, but decreases to 28-49% with adjuvant radiotherapy.[20] Generally, lymph node dissection is not necessary when the lymph nodes are negative. Ifosfamide has been given in some cases with good results; however, the cost effectiveness of chemotherapy is still not clear because of the rarity of synovial sarcoma.[17]

In conclusions, we presented the fourth case of synovial sarcoma of the infratemporal fossa reported in the English literature; unlike the previous cases, this showed intracranial extension. We believe that the publication of further case reports will lead to standard methods for diagnosing and treating synovial sarcoma.

Declaration of conflicting interests

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REFERENCES

1. Pai S, Chinoy RF, Pradhan SA, D'Cruz AK, Kane SV, Yadav JN. Head and neck synovial sarcomas. J Surg Oncol 1993;54:82-6.
2. Bertolini F, Bianchi B, Pizzigallo A, Tullio A, Sesenna E. Synovial cell sarcoma of the neck. Case report and review of the literature. Acta Otorhinolaryngol Ital 2003;23:391-5.
3. Roth JA, Enzinger FM, Tannenbaum M. Synovial sarcoma of the neck: a followup study of 24 cases. Cancer 1975;35:1243-53.
4. Taylor SM, Ha D, Elluru R, El-Mofty S, Haughey B, Wallace M. Synovial sarcoma of the pericricoidal soft tissue. Otolaryngol Head Neck Surg 2002;126:428-9.

5. Gallia GL, Sciubba DM, Hann CL, Raman SP, Westra WH, Tufaro AP, et al. Synovial sarcoma of the frontal sinus. Case report. J Neurosurg 2005;103:1077-80.
6. Hirsch RJ, Yousem DM, Loevner LA, Montone KT, Chalian AA, Hayden RE, et al. Synovial sarcomas of the head and neck: MR findings. AJR Am J Roentgenol 1997;169:1185-8.
7. Sigal R, Chancelier MD, Luboinski B, Shapeero LG, Bosq J, Vanel D. Synovial sarcomas of the head and neck: CT and MR findings. AJNR Am J Neuroradiol 1992;13:1459-62.
8. Carrillo R, Rodriguez-Peralto JL, Batsakis JG. Synovial sarcomas of the head and neck. Ann Otol Rhinol Laryngol 1992;101:367-70.
9. Cihak RA, Lydiatt WM, Lydiatt DD, Bridge JA. Synovial sarcoma of the head and neck: chromosomal translation (X;18) as a diagnostic aid. Head Neck 1997;19:549-53.
10. Dei Tos AP, Dal Cin P, Sciot R, Furlanetto A, Da Mosto MC, Giannini C, et al. Synovial sarcoma of the larynx and hypopharynx. Ann Otol Rhinol Laryngol 1998;107:1080-5.
11. Quinn HJ Jr. Synovial sarcoma of the larynx treated by partial laryngectomy. Laryngoscope 1984;94:1158-61.
12. Nadig SK, Love MH, Brooker DS, Hall SJ. Synovial sarcoma in the retropharyngeal space. J Laryngol Otol 2002;116:224-6.
13. Bukachevsky RP, Pincus RL, Shechtman FG, Sarti E, Chodosh P. Synovial sarcoma of the head and neck. Head Neck 1992;14:44-8.
14. Duvall E, Small M, Al-Muhanna AH, Maran AD. Synovial sarcoma of the hypopharynx. J Laryngol Otol 1987;101:1203-8.
15. Mamelle G, Richard J, Luboinski B, Schwaab G, Eschwege F, Micheau C. Synovial sarcoma of the head and neck: an account of four cases and review of the literature. Eur J Surg Oncol 1986;12:347-9.
16. Sigal R, Chancelier MD, Luboinski B, Shapeero LG, Bosq J, Vanel D. Synovial sarcomas of the head and neck: CT and MR findings. AJNR Am J Neuroradiol 1992;13:1459-62.
17. Bilgic B, Mete O, Oztürk SA, Demiryont M, Keles N, Basaran M. Synovial sarcoma: a rare tumor of larynx. Pathol Oncol Res 2003;9:242-5.
18. Lai V, Farrag TY, Cao D, Aygun N, Boahene K, Tufano RP. Synovial sarcoma of the infratemporal fossa. Am J Otolaryngol 2007;28:444-7.
19. Helmberger RC, Stringer SP, Mancuso AA. Rhabdomyoma of the pharyngeal musculature extending into the prestyloid parapharyngeal space. AJNR Am J Neuroradiol 1996;17:1115-8.
20. Doval DC, Kannan V, Mukherjee G, Shenoy AM, Shariff MH, Bapsy PP. Synovial sarcoma of the neck. Eur Arch Otorhinolaryngol 1997;254:246-50.