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OLGU SUNUMU

Synovial sarcoma arising from the hypopharynx: a case report

Hipofarenksten kaynaklanan sinovyal sarkom: Olgu sunumu

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Synovial sarcoma is a relatively rare soft tissue tumor. About 3 to 10% of cases arise in the head and neck, with the majority in the parapharyngeal region. We hereby report a 22-year-old male patient with synovial sarcoma confined to the hypopharynx. Treatment included wide surgical excision followed by radiotherapy. It is of interest that the lesion recurred 12 years after the initial primary surgery for synovial sarcoma at the same site. The patient has been disease-free for five years.

Key Words: Diagnosis, differential; hypopharyngeal neoplasms/pathology/surgery; recurrence; sarcoma, synovial/pathology/surgery.

Sinovyal sarkom yumuşak dokularda görülen nadir bir tümördür. Olguların %3-10'u baş-boyun yerleşimlidir, en sık parafarenjeal bölgede rastlanır. Bu yazıda, hipofarenkse yerleşik sinovyal sarkom saptanan 22 yaşında bir erkek hasta sunuldu. Olgu geniş cerrahi eksizyon ve radyoterapi ile tedavi edildi. Hastanın 12 yıl önce de aynı yerde görülen sinovyal sarkom nedeniyle ameliyat edildiği öğrenildi. Halen yaşamakta olan hastada beş yıldır tümör ve tedavisiyle ilgili bir sorun görülmedi.

Anahtar Sözcükler: Tanı, ayırıcı; hipofarenjeal neoplazmlar/patoloji/cerrahi; rekürens; sarkom, sinovyal/patoloji/cerrahi.

Synovial sarcoma of extraarticular localization is very rare.[1,2] Of all synovial sarcomas, 3% to 10% arise in the head and neck.[3] Because of the rarity of this tumor, there is no general agreement on the treatment modalities. Most of the reported cases are treated either with primary surgical excision alone or in conjunction with radiotherapy.

CASE REPORT

A male patient at the age of 22 years presented with a two-year history of sore throat, dyspnea, dysphagia for solid foods, dysphonia and cough. Within the last month, he had lost about three kilograms of body weight. Indirect laryngoscopy revealed a mass lesion, 4x3 cm in size, that originated from the posterior hypopharyngeal wall and almost completely filled the supraglottis with a very narrow airway left anteriorly. A biopsy of the lesion obtained under local anesthesia showed biphasic synovial sarcoma. He had a past history of tumor excision from the same side 12 years ago; although the result of the pathologic examination had initially been reported as adenocarcinoma, it turned out to be synovial sarcoma after total excision. It seemed

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that the former excision had been made transorally as there was no incision scar on the patient's skin. Postoperative radiotherapy recommended at that time had been refused by the patient's family.

On physical examination, no pathologic finding was found other than the mass lesion in the hypopharynx. Computed tomography (CT) showed a lesion at the level of the epiglottis, measuring 3x3 cm and comprising the airway. By magnetic resonance imaging (MRI) the mass lesion in the hypopharynx manifested a broad base on the prevertebral muscles and filled the airway significantly. The mass extended from the level of C_2 to the level of C_6 and showed no evidence of parapharyngeal or neurovascular invasion; however, signs of invasion to the left prevertebral region was noted (Fig. 1). In addition, there was a small calcified focus within the lesion. Further investigations by thorax CT, abdominal ultrasonography (USG), and bone scans showed no evidence of distant metastasis.

Under general anesthesia, and following tracheotomy, the tumor was excised via a transoral and transhyoid approach with wide surgical margins. The resulting defect in the posterior and left hypopharyngeal walls was reconstructed with a split-thickness skin graft from the left thigh and with the pectoralis major myocutaneous flap. No complications were encountered during the postoperative period. The patient was decannulated on the 12th postoperative day and was referred to radiotherapy for further treatment with 4800 cGy given to the tumor area and the neck.

Fig. 1 - *Sagittal magnetic resonance image of the hypopharyngeal mass.*

The histopathologic diagnosis was biphasic synovial sarcoma and the surgical margins were reported as tumor-free (Fig. 2a, 2b). The tumor was composed of two morphologically diverse types of cells that formed the characteristic biphasic pattern, namely epithelial cells resembling carcinoma and a sarcomatous component. Both the epithelial and spindle cells showed reactivity for cytokeratin (CK) and epithelial membrane antigen (EMA). However, the positivity in the spindle cell component was less intense and only scattered. The expression of vimentin was strong and diffusely positive in the spindle cells.

The patient has been alive and without evidence of recurrence five years after the operation.

DISCUSSION

Synovial sarcoma is most commonly found in adolescents and young adults, with a male-to-female ratio of about 2-3 to 1.[4-6] To date, not more than 100 cases of synovial sarcoma have been reported in the head and neck region.[7] In the head and neck, this tumour presents as less aggressive, with higher survival and lower recurrence rates.[7] To our knowledge, only one case of hypopharyngeal synovial sarcoma has been reported in Turkey.[5]

Patients with hypopharyngeal synovial sarcoma present most commonly with dyspnea, dysphonia and dysphagia due to tumor mass; pain is less frequent.[6] Radiologic evaluation (CT and/or MRI) is helpful in delineating tumour localization, extension, and airway restriction.[8] Calcification within the tumour is noted in 30 to 40% and is regarded as a good prognostic factor.[8,9]

Although there is no general agreement about their histogenetic origin,[2] synovial sarcomas in the head and neck are thought to arise not from synovial tissue, but from pluripotential mesenchymal cells, which undergo differentiation to an inner epithelial layer and an outer layer of connective tissue.[4] On gross examination, they are between 2 to 10 cm in diameter, and are usually firm and well-circumscribed with a pseudocapsule; the overlying skin or mucosa is usually of normal appearance.[4] Radiographically, they may appear benign in some cases.[8] Microscopically,

they are classified into monophasic and biphasic variants.[10] In the biphasic variant, the background stroma is made up of fibroblast-like cells packed tightly together, and the epithelial cells show gland-like formation.[3,4] The malignant nature of the stroma may not be recognized, and the tumor may be misdiagnosed as adenocarcinoma, which was the case in our patient 12 years before.[3,6] The demonstration of CK and EMA in the epithelial layer and vimentin in the stroma are almost pathognomonic for this tumor;[4] all three were positive in our patient. Soft tissue sarcomas of the head and neck are often misdiagnosed as poorly-differentiated tumors without the use of special immunohistochemical staining. An accurate pathologic diagnosis is of great significance for the therapeutic strategy.[11]

Management of hypopharyngeal synovial sarcoma requires wide surgical excision, because inadequate excision is associated with a high incidence of local recurrence (60-90%), usually within the first two years. Nearly all cases were treated by surgery alone or in combination with radiotherapy. Metastasis to regional lymph nodes is not common. Enlarged lymph nodes in some cases may not be an

(b)

Fig. 2 - **(a)** Biphasic pattern composed of epithelial glandular and cleft-like structures and sarcomatous areas (H-E x125). **(b)** Fibromyxoid stroma with hypercellular area around vessels (H-E x 125).

indication for neck dissection.[4,6] The size and extent of the tumor (primarily the depth) at the time of primary treatment are the most important prognostic factors, and radiotherapy is recommended especially in cases with microinvasion.[3-6] Furthermore, in a study of 14 cases of synovial sarcoma, localized disease was treated by intensive chemotherapy, with encouraging results.[12]

Hematogenous spread is far more common than lymphatic spread and results in distant metastases, which is the most important cause of mortality.[3,4] Therefore, is difficult to estimate both the prognosis and survival. The five-year survival rate is reported between 23% to 63%, with 10-year survival being much lower between 11% to 30%. The considerable difference between the 5- and 10-year survival rates empahizes the possibility of late recurrence and metastasis, especially to the lungs, hence, the importance of long-term follow-up.[4,6] Our patient had undergone surgery for synovial sarcoma at the same localization 12 years before, giving an idea about the late recurrence potential of the tumor. We believe that this case of synovial sarcoma may contribute to our understanding of its long-term recurrence potential.

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