Case Report / Olgu Sunumu

Leiomyosarcoma of the soft palate

Yumuşak damak leiyomiyosarkomu

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Tumors of the smooth muscles are rarely seen, as the number of smooth muscles is low within the intraoral region. Leiomyosarcoma is a type of malign tumor originating from smooth muscles. The most common regions of leiomyosarcoma of the oral cavity are the maxilla and mandible. In this article, we present a leiomyosarcoma detected in a 20-year-old male patient who was admitted to the clinic with the complaint of a mass for about three months. The mass was located in the left half of the soft palate and it was resected *en bloc* with the mucosa. No recurrence was observed during the two-year follow-up period of the patient.

Key Words: Leiomyosarcoma; smooth muscle; soft palate.

İntraoral bölgede düz kas azlığı nedeni ile düz kas kaynaklı tümörler oldukça nadir görülür. Leiyomyosarkom düz kaslardan kaynaklanan bir malign tümör tipidir. Ağız içi leiyomyosarkomlarının en sık yerleşim yerleri maksilla ve mandibuladır. Bu yazıda yaklaşık üç aydır var olan kitle yakınması ile kliniğe başvuran ve leiyomyosarkom saptanan 20 yaşında bir erkek olgu sunuldu. Kitle yumuşak damağın sol yarımında yerleşik idi ve mukozası ile beraber *en blok* rezeke edildi. Hastanın iki yıllık takip süresinde nüks gözlenmedi.

Anahtar Sözcükler: Leiomiyosarkom; düz kas; yumuşak damak.

Tumors of the smooth muscles are usually seen in the uterus and rarely in the gastrointestinal system, skin and subcutaneous tissue.[1] The intraoral zone is not a usual place for a smooth muscle tumor to occur. In a review where 7748 smooth muscle tumors were classified throughout the body, intraoral smooth muscle tumors were reported with a 0.064% ratio.[2]

The reason for this low incidence in the oral cavity is, as might be expected, the small number of smooth muscles in the area.[3] However, the arterial tunica media, the smooth muscle of the ductus lingualis and the circumvallate papillae have been proposed as the source of the smooth muscle tumors observed within the oral cavity.[4-6]

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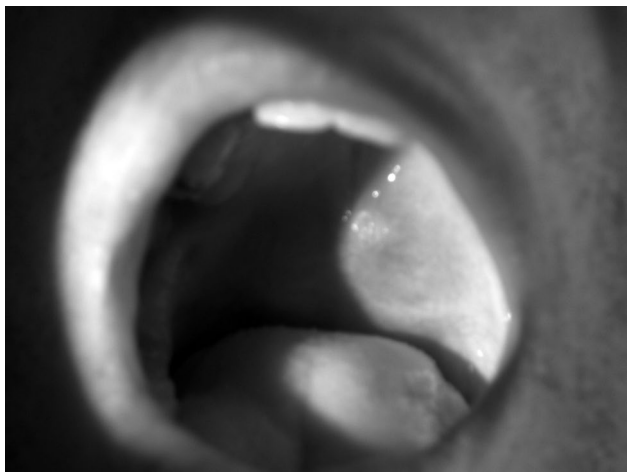

Figure 1. *View of the lesion.*

Compared to other areas of the body, the potential for malignancy of the intraorally located smooth muscle tumors is rather high. While the leiomyosarcoma (LMS)/leiomyoma (LM) ratio in the female genital system is 1/200, the same ratio is 20% in the intraoral region.[2,3]

A leiomyosarcoma originating from the soft palate is presented in this report.

CASE REPORT

During the physical examination of a 20-year-old male who presented to the clinic with a painless growth that had been in his mouth for approximately three months, a 1x1 cm mass with a darker color than the surrounding mucosa was detected on the left half of the soft palate (Figure 1). There was no risk factor pointing to an intraoral carcinoma in the patient's history other than cigarette smoking (1 pack/day for 5 years). Since no additional local or systemic pathologies (lymphadenopathy in the neck, mass in the lungs, etc.) were observed in the patient, the mass was extracted en bloc together with overlying mucosa and a safety margin of approximately 1 cm. The defect was left to heal by secondary intention. On histopathologic examination, tumor infiltration between the mucinous glands was observed. The tumor consisted of atypical spindle-like cells in intercrossing bundles. There was sparse mitotic activity in the tumor cells and they were immunohistochemically stained desmin-positive. No cytokeratin receptor expression was detected. When the morphological findings were evaluated together with immunohistochemical findings, the tumor was assessed as low-grade leiomyosarcoma (Figures 2, 3). The surgical borders were negative for malignancy. No recurrence was observed during the two-year follow-up period of the patient.

DISCUSSION

Leiomyosarcoma is a type of malignant tumor originating from smooth muscles. It comprises 6% of all soft tissue sarcomas and 3% of cases are seen in the head and neck, oral cavity, sinonasal and submandibular regions.[7] Genetic factors, acquired immune system depression related to AIDS or the use of immunosuppressive agents, the Epstein-Barr virus can be listed among the etiological factors.[8]

Although intraorally-located LMS is rather rare,[3] the usual locations in the oral cavity are the maxilla and the mandible. In their review article

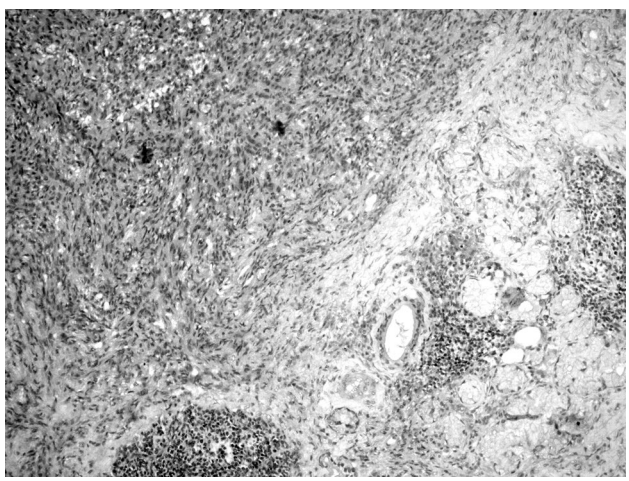

Figure 2. *View of the infiltrative tumor forming bundles (H-E x 200).*

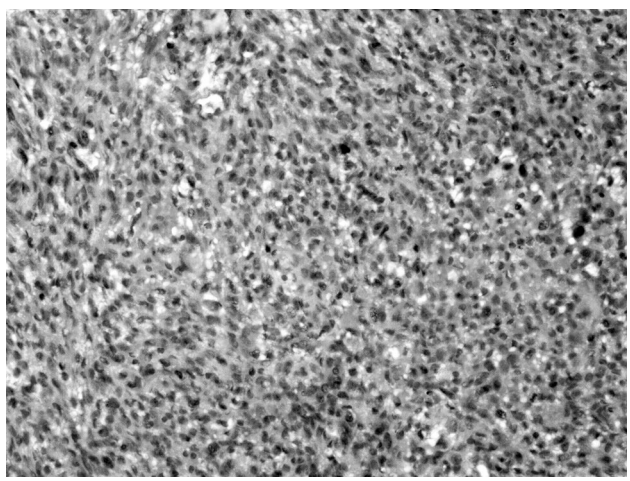

Figure 3. *View of the spindle-like cells and the sparse mitotic activity (H-E x 400).*

on smooth muscle tumors reported between 1884 and 1996 (139 leiomyomas, 68 leiomyosarcomas), Wertheimer-Hatch et al.[3] reported eight palatal LMS cases among a total of 68 leiomyosarcomas (12.7%). Since four palatal leiomyosarcoma cases have been reported since 1996,[8-11] our case is the 13th in the literature.

According to Wertheimer-Hatch et al.,[3] there is no difference between the sexes in terms of leiomyosarcoma incidence. The three age intervals where LMS is most likely to be observed are 50-59, 60-69 and 20-29 years of age. The patient in our case was a 20-year-old male.

The most common complaint in intraoral LMS is swelling and pain. It can also rarely cause problems in chewing and swallowing. Tenderness, rapid growth, toothache and loose teeth, necrosis, bleeding, itching, numbness, sensitivity, a feeling of pressure and exfoliation are the other symptoms.[3] In our case, the only complaint was a painless swelling. Mucocele, pleomorphic adenoma, hemangioma, fibroma, fibrous histiocytoma, fibrosarcoma, myosarcoma, neurilemmoma, neurofibroma, lipoma, giant cell granuloma, peripheral ossifying fibroma, squamous cell carcinoma, and advanced chronic periodontitis can be considered in the differential diagnosis.[3] Still, achieving the correct diagnosis without a biopsy is rather difficult. Nevertheless, distinguishing between benign and malignant tumors of the smooth muscle microscopically is also difficult and the Evans[12] malignancy criteria are employed for this purpose. These are increased cell size, increased irregularity of cell size and shape, lack of complete cell differentiation, presence of short and plump cells with oval nuclei and the presence of cells with hyperchromic and multiple nuclei with variable staining reactions.

The recommended choice of treatment for surface leiomyosarcomas is wide local excision. Limited local excision increases the local recurrence rate. For LMS, a surgical border of 1-2 cm is sufficient. A neck dissection can be applied because of the potential for regional and distant metastases.[7] In our case, since the dimensions of the lesion were small, a security margin of 1 cm has been considered to be sufficient and no re-excision has been planned. Leiomyosarcoma is thought to be resistant to radiotherapy[3] and the effect of postoperative radiotherapy is disputable.[3,13] Again, we did not plan any postoperative radiotherapy for the lesion because of its small dimensions.

In conclusion, LMS must be considered in the differential diagnosis of cases presenting with a painful palatal swelling.

Declaration of conflicting interests

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