LEIOMYOSARCOMA OF THE CHEEK. A CASE REPORT

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SUMMARY

Leiomyosarcomas are extremely rare in the oral cavity. So far, only five cases of primary leiomyosarcomas of the cheek have been reported. Here, we present an additional case of leiomyosarcoma of the cheek and discuss the histopathological characteristics, differential diagnosis and treatment modalities.

Key Words : Leiomyosarcoma, cheek, oral cavity

INTRODUCTION

Leiomyosarcomas are malignant neoplasms arising from smooth muscle cells. Although the majority of leiomyosarcomas develop in the uterus and gastrointestinal tract, they rarely originate within the oral cavity. To our knowledge, only five cases of primary leiomyosarcomas of the cheek have been reported so far. The purpose of our article is to report an additional case of leiomyosarcoma of the cheek and discuss its clinical and pathological characteristics.

CASE REPORT

A 45-year old male patient, who had been operated three times (1984, 1988, 1989) in two different hospitals, was referred to Ear, Nose and Throat Department of Hacettepe University in April 1990, with a recurrent soft tissue mass in his right cheek. It was learned that the lesion had involved the right cheek with extension to the right parotid region. The previous histopathological examinations had been interpreted as angiomyoma. The original blocks of these specimens could not be obtained for reexamination at our institute. A metastatic lesion of three centimeters in diameter in the left lung lower lobe superior segment, was also surgically removed in April 1990. It was reported as leiomyosarcoma metastasis.

On our initial examination, we detected a mass that involved the mucosal side of the cheek and extended to and filled the right parapharyngeal space. This lesion also continued in the preauricular region with a smooth surfaced elastic mass sized 6x7 cm with no ulceration on its surface. Physical examination revealed no palpable lymph node in the neck. Computed tomography disclosed a solid lesion located in the right cheek, destructing right mandibular ramus, maxilla and processus pterygoideus (Fig. 1). The mass pushed the parotid gland backwards, completely filling right pterygopalatine fossa, parapharyngeal space and Rosenmuller fossa. The biopsy of the lesion was evaluated as leiomyosarcoma at our institute.

The patient was given 52 Gy radiotherapy. In July 1990, a six centimeters metastatic lesion was discovered in the left lung. This time he was given chemotherapy (cyclophosphamide, vincristine, adriamycine and dacarbazine). There was no response to these treatments and the tumour kept on its growth filling the oral cavity almost totally. In November 1991 the patient underwent a tumour reduction operation in order to provide comfortable oral feeding. The histopathological examination of the specimen confirmed the diagnosis of leiomyosarcoma once again.

He was seen in the ENT department in April 1992 for the last time. He had multiple metastases bilaterally

in the lungs. His general condition was poor and he had another tumour reduction operation.

Histopathologic study of the two specimens obtained in our institute shared the same characteristics mentioned below. The microscopic slides were prepared from formalin-fixed tissue and stained with hematoxylin and eosin Masson's trichrome, vimentin and desmin. The tumour was composed of proliferative spindle cells with elongated cytoplasmic extensions arranged in bundles (Fig. 2). The nuclei of the tumour cells were cigar-shaped with blunt ends and mitotic count was 6 per 10 high power field (Olympus BH2 microscope, 10 oculer x 40 objective). There was no necrosis, but some areas were highly cellular and pleomorphic. Histochemically, neoplastic cells revealed red colour with Masson's Trichome. Immunohistochemically, some tumor cells showed vimentin and desmin positivity (Fig. 3). Electron microscopic examination exhibited myofilements with dense body formation and numerous pinocytotic vesicles (Fig. 4). As a result overall pathologic findings emphasize high grade malignancy.

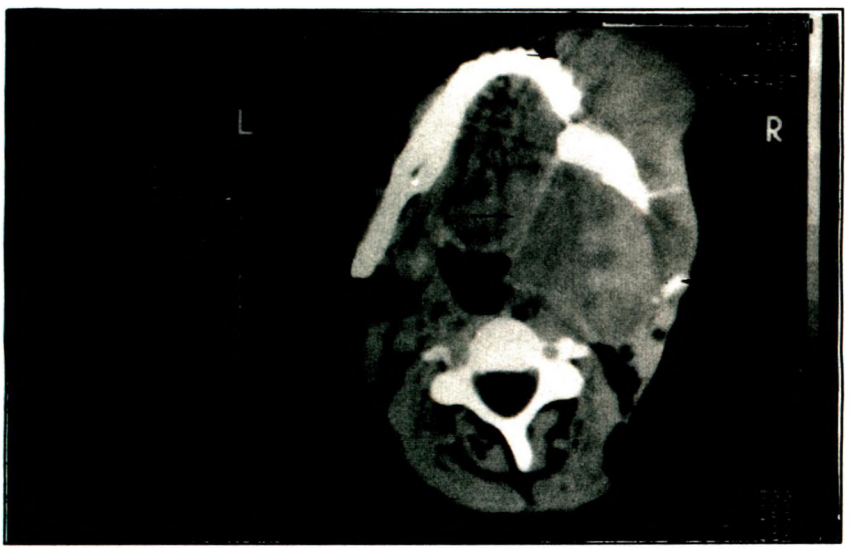

Fig. 1:
The solid lesion located in the right cheek, destructing right mandibular ramus, maxilla and processus pterygoideus, filling right pterygopalatin fossa and parapharyngeal space.

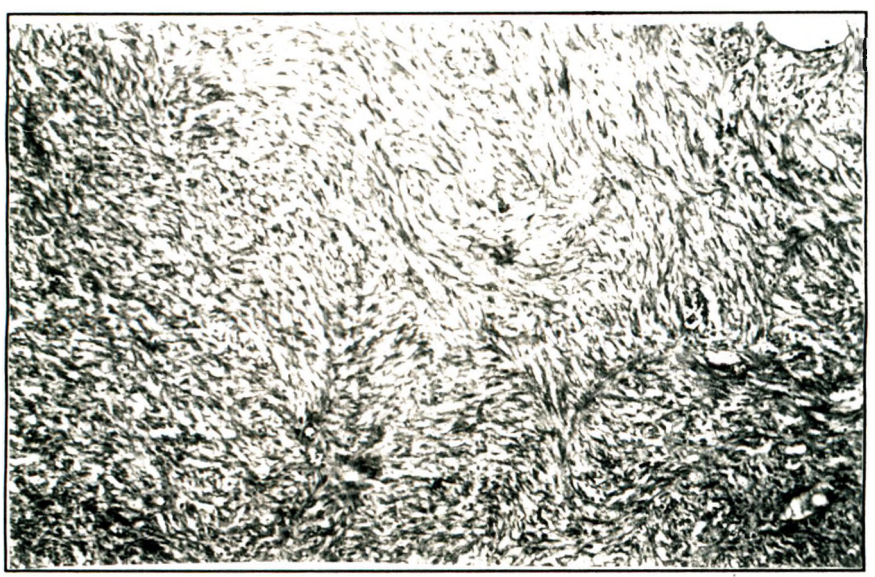

Fig. 2:
Proliferative spindle cells with elongated cytoplasmic extensions arranged in bundles (Haematoxylen-Eosin)

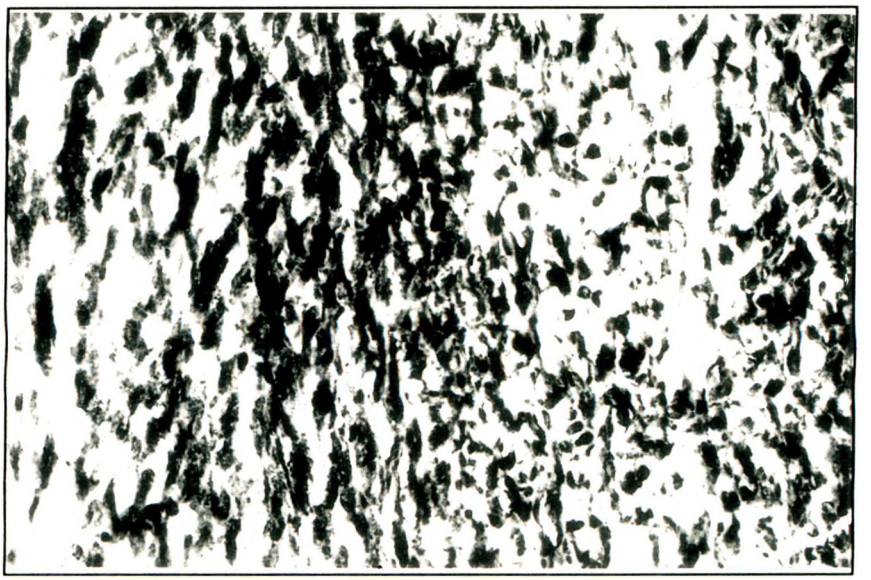

Fig. 3:
Vimentin and desmin positivity in tumoural cells

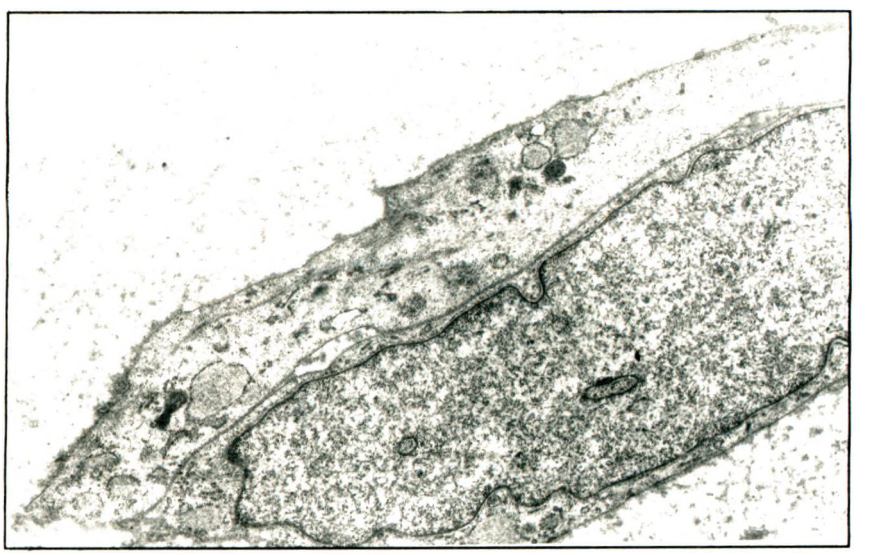

Fig. 4:
Myofilaments with dense body formation and numerous pinocytotic vesicles (Electron microscopy)

DISCUSSION

Leiomyosarcomas comprise about 2.3 - 5.3% of all malignant soft tissue tumours (1). They have equal sex incidence and the ages of 40% of the cases vary between one and 29 years (2).

They usually present as a slowly enlarging, discrete, smooth, firm, subcutaneous or submucosal unulcerated painless mass (3). Pain and tenderness is a more recognised feature of leiomyomas (4).

The most common site of smooth muscle tumours is the uterus (5). An extensive review of 7748 smooth muscle tumours by Farman and Kay (6) revealed that 95% occurred in the female genitourinary tract and they also noted that only two of these patients had tumours in the oral cavity. On the other hand, O'Day at al. (7) stated that only three of the 219 (1.4%) malignant mesenchymal tumours located in the oral cavity were leiomyosarcomas. Poon et. al. (2) were able to find only 25 cases of oral leiomyosarcomas in an extensive literature survey from 1908 to 1987. Only five of these occurred in the cheek.

The reason for this rarity is believed to be due to paucity of the smooth muscles in the oral cavity (2). Recent reports suggest that smooth muscle elements of blood vessels walls, the circumvallate papilla, aberrant hair follicles in the cheek, and the primitive mesenchyme can be the source of these tumours in the oral cavity. Miles and Waterhouse (8) also

suggested that accessory salivary glands which are located in the submucosa of this region can act as a source of smooth muscle tumour.

Enzinger et. al. (9) classified benign smooth muscle tumours as cutaneous leiomyoma (leiomyoma cutis), angiomyoma (vascular leiomyoma), and leiomyoma of deep soft tissues. In our case pathological examination of the first three operations which had been performed in different hospitals, were evaluated as angiomyoma. Duhig and Ayer (10) suggested that leiomyomas should be considered as vascular malformations, and it is easy to reach a wrong conclusion of angiomyoma instead of leiomyosarcoma. In the differential diagnosis other soft tissue sarcomas such as fibrosarcoma, synovial sarcoma and malignant schwannoma should be considered. From this point of view histochemistry and immunohistochemistry and electron microscopy are supportive techniques. In the presented case, red colouration with Masson's trichrome, diffuse vimentine positivity, focal desmin positivity with immunohistochemistry and pinocytotic vesicles with electron microscopy suggest smooth muscle differentiation. Therefore the possibility of misinterpretation always has to be kept in mind.

It is also important to differentiate leiomyoma from leiomyosarcoma. The metastatic lesion in the lung and the excised lesion in the cheek were reported as leiomyosarcoma whereas original diagnosis was leiomyoma.

According to Haediche and Kaban (5), oral leiomyoma and leiomyosarcoma share many characteristics histopathologically and it is sometimes extremely difficult to make a precise differentiation between them unless regional lymph node involvement or metastasis is present. Stout and Hill (4) stated that, the frequency of mitotic activity is the basic criterion to determine the state of neoplasm. One or more mitoses per five highpower fields, is considered to be associated with malignancy, but mitotic activity may only be present in some parts of the tumour. Therefore, the entire tumour must be meticulously examined. Also in contrast to the benign leiomyomas, malignant sarcomas tend to show nuclear pleomorphism and cellular atypia.

Successful treatment of oral leiomyosarcomas is based on early wide excision (3, 7, 11). Approximately 60% of the leiomyosarcomas will recur after local excision (4). Simple enucleation without resection of the surrounding soft tissues is the main cause for local recurrences (11). Our patient had three operations on the primary tumour side all of which resulted with recurrence. The fourth attempt was solely aimed to reduce the bulk of the tumour in order to provide comfortable eating. According to Mindell and Calcaterra (3), tumour can even recur up to six years following initial therapy.

As lymph node metastasis is occasionally encountered in leiomyosarcomas prophylactic neck dissection is not routinely indicated (3, 7). However, the presence of any palpable nodes in the neck is an indication for neck dissection. In our case, despite the huge tumour size and three recurrences, no adenopathy could be palpated in the neck. This is an example for its rare spreading into the lymphatic system.

Thirty three percent of leiomyosarcomas spread via blood stream, mostly to the lungs and the vertebrae. Our patient had lung metastasis twice without any evidence of vertebra metastasis. Although the presence of distant metastasis is generally thought to reduce the survival rate considerably, we advocate the removal of solitary metastasis surgically. This approach provided our patient two years of survival.

Leiomyosarcomas of other parts of the body are known to be minimally radiosensitive and this appears to hold true for the head and neck region (1,7). Although chemotherapy can be added to radiotherapy, experience with this mode of treatment is still insufficient(5). In our patient, radiotherapy and chemotherapy had no effect in controlling the primary tumour and metastasis.

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