Metastasis of Liposarcoma in the Parotid Gland Two Years after the Initial Treatment

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ABSTRACT

Metastatic tumours of the parotid gland represent an extremely rare pathological entity. The majority of metastatic tumours of the region in question includes metastases of squamous cellular carcinoma and skin melanoma of the temporal region and the scalp, the described metastasis of liposarcoma represents a quite rare pathological entity. A 58-year-old female patient with a tumefaction in the parotid region was sent to the Clinic of Maxillofacial Surgery. Having examined the available medical records, it was found that the patient had been operated on three times. All surgeries were on the soft tissue of the right lower leg, with the last one being the amputation with a histopatohologically confirmed myxoid liposarcoma. The clinical case conference decided to conduct postoperative chemoradiation therapy due to the extensiveness of the tumour and endangered vital structures. Despite the conduction of appropriate postoperative procedures, radical surgical procedure, and postoperative oncological therapy, metastatic tumours of the parotid gland are characterized by a low five-year survival rate.

Keywords: Liposarcoma, metastasis, parotid gland

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INTRODUCTION

Metastatic tumours of the parotid gland represent an extremely rare pathological entity which comprises 8% of the total number of malignant tumours of this region (1).

Lymphogenous or hematogenous dissemination of malignant tumours of the facial skin and the scalp towards the parotid gland is much more frequent than the dissemination of infraclavicular malignity which has metastases in the parotid gland via the thoracic duct or Batson paravertebral venous plexus (2). The clinical manifestation of metastatic tumours is variable, from the occurrence of massive exulcerated tumours with foudroyant progression, the infiltration of the facial nerve and deep anatomic structures, to cases in which a tumour and its behaviour resemble benign tumours of the glandular tissue.

The aim of this paperwork is to present a case of a female patient with metastatic liposarcoma of the parotid gland two years after the initial treatment, along with the review of the conducted diagnostic and therapeutic procedures.

CASE REPORT

A 58-year-old female patient with a tumefaction in the parotid region was sent to the Clinic of Maxillofacial Surgery in Niš. According to the patient, the tumefaction had appeared 3 months earlier with a rapid increase in the last three weeks. Painful sensations were moderate, and the opening of the mouth limited due to which the patient consulted an oncologist who sent her to a maxillofacial surgeon after the examination.

Having examined the available medical records, it was found that the patient had been operated on three times. All surgeries were on the soft tissue of the right lower leg, with the last one being the amputation with a histopatohologically confirmed myxoid liposarcoma (liposarcoma myxoides).

Radiotherapy was conducted after the operation. The clinical examination confirmed an oval-shaped tumefaction in the right parotid region, 4x6 cm in size, painless to touch and immobile. The paresis of the marginal branch of the facial nerve, limited opening of the mouth of the first degree (trismus gradus I) and lymphadenopathy of I, II, III and IV level of the neck (Fig. 1) were also confirmed.

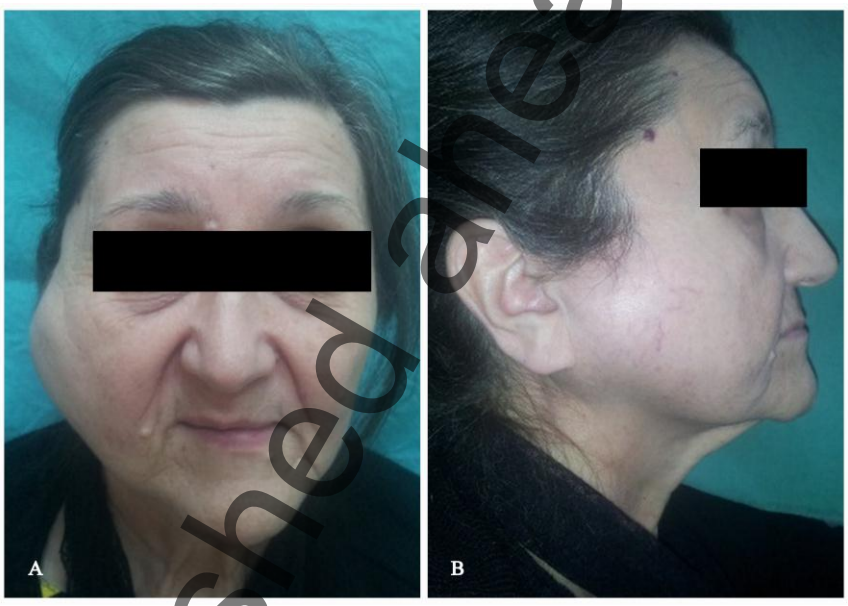

Fig. 1A: Tumefaction of the parotid region (frontal face appearance); Fig. 1B - Tumefaction of the parotid region (lateral face appearance).

As part of the diagnostic procedure, the X-ray of the lungs and ultrasound of the abdomen were done and both were within the physiological limits. Furthermore, the MSCT of the middle third of the face with noticeable existence of an expansive tumour formation of the parotid region, soft tissue density, with the extension into the parapharyngeal,

pterygomandibular and infratemporal space, was done. Also, the destruction of the posterior wall of the maxillary sinus with the propagation of the tumour into the distal segment was observed (Fig. 2).

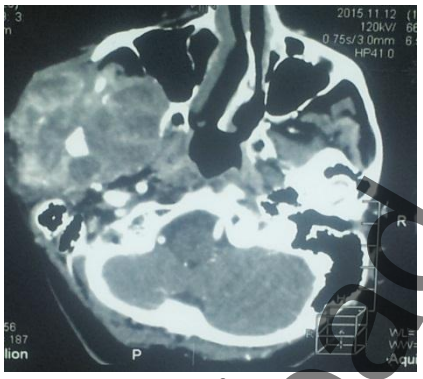

Fig. 2 – MSCT image of the expansive tumour formation

Moreover, the transantral excisional biopsy of the tumour of the distal segment of the maxillary sinus was conducted, as well as the aspiration biopsy (fine-needle aspiration biopsy – FNAC). Both biopsy specimens showed the existence of myxoid liposarcoma (Fig. 3).

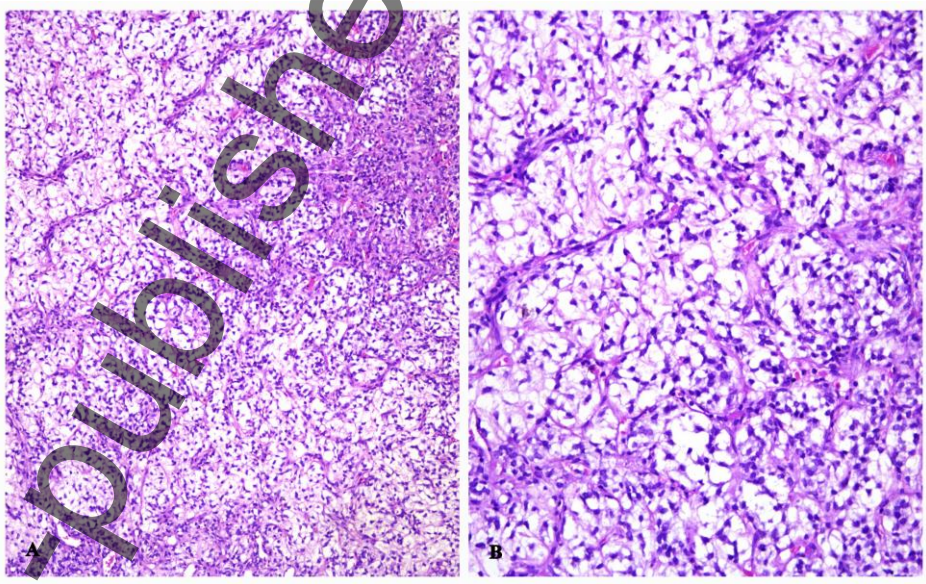

Fig. 3 - Metastasis liposarcoma in parotid gland, A-H&E, original magnification x10:
B- H&E, original magnification x20

The clinical case conference decided to conduct postoperative chemoradiation therapy due to the extensiveness of the tumour and endangered vital structures. The patient's general condition got worse during the fourth week of the oncological treatment due to the appearance of multiple intracranial and intrapulmonary metastases. The patient continued to live for two months after the pathohistological verification of the metastases of liposarcoma in the parotid region.

DISCUSSION

Liposarcomas belong to the group of soft tissue sarcomas (STS) with extremely variable behavior, ranging from non-invasive to extremely expansive tumour changes of foudroyant genesis (3).

They can be diagnosed in all parts of the body, but they are most frequent on the limbs and retroperitoneum, comprising 1% of the total number of all human tumours (4).

On a pathohistological level, liposarcomas are divided into three groups. The first group includes atypical lipomatous tumour, well-differentiated (WDLPS) and dedifferentiated liposarcoma (DDLPS), the second group includes myxoid and round-cell LPS, whereas the third includes the pleomorphic type of LPS (5).

The systemic dissemination of LPS ranges from 1 to 18% with dominant occurrence in the lungs [75%] (6). The initial absence of the tumour process in the lungs of the presented patient can be interpreted by the dissemination of the process via Baton paravertebral plexus (7).

Frequent local recurrence (40–80%) after the initial treatment is in direct correlation with the increased rate of metastases, which represents another clinical characteristic of LPS

(8). Compared to secondary liposarcoma, de novo provides a higher percentage of remote metastases, which can be justified by a considerably more aggressive tumour biology (9). The occurrence of remote metastases of LPS is associated with a bad prognosis (9). Ghadimi et al. state that the one-year or five-year survival rate is considerably higher in patients who underwent surgery as part of the treatment of secondary deposits (10). The same authors also suggest that the localization of metastases does not affect the survival rate, but that they noticed a trend of a lower survival rate in patients with secondary deposits in the lungs. The therapy of LPS implies an extremely radical initial surgical procedure to reduce the possibility of local recurrence and secondary deposits, along with the conduction of appropriate oncological therapy (11). Subtotal or radical parotidectomy accompanied by lymphadenectomy of the neck is standard surgical procedures in treating operable lesions. Response and sensitivity to systemic therapy depends of liposarcoma subtype and also anatomic site. Traditional regimen used in metastatic setting containing doublets of doxorubicin/ifosfamide or gemcitabine/docetaxel. This regimens are also used in our country. Response rate is between 25% and 35% and survival is 12 to 18 months (12–14).

Today we have a few newer agents in metastatic disease like trabectedin and eribulin which have received recent FDA approval for application in the second-line setting for liposarcoma (15–17). In 90% of well-differentiated liposarcoma (WDLS) and dedifferentiated liposarcoma CDK4 oncogene is amplified and Pablociclib, a potent CDK4/CDK6 inhibitor also used in breast cancer, has shown activity by increasing progression free survival (18, 19). We have positive results of early phase clinical trial of RG7112, MDM2 antagonist, but further investigation is needed (20). Other agents investigated in clinical studies like agonists of PPAR-gamma (regulator of adipocytic

differentiation) and nelfinavir (protease inhibitor used in HIV treatment) have shown no proven benefit (21, 22).

Given that the majority of metastatic tumours of the region in question includes metastases of squamous cellular carcinoma and skin melanoma of the temporal region and the scalp, the described metastasis of LPS represents a quite rare pathological entity reported only twice in the available literature, by Lopez and Trabelsi (23, 24). Given a wide range of pathological entities which comprise the differential diagnosis of the mentioned lesions, the treatment of primary and secondary tumours of the parotid gland represents a true challenge in everyday work of maxillofacial surgeons and pathologists. The initial biopsy and appropriate radiological diagnostics are the bases for the creation of a future therapy plan. Despite the conduction of appropriate postoperative procedures, radical surgical procedure, and postoperative oncological therapy, metastatic tumours of the parotid gland are characterized by a low five-year survival rate.

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