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Case report

The case of malignant peripheral nerve sheath tumor of the cheek

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ABSTRACT

Introduction: Malignant peripheral nerve sheath tumor (MPNST), formerly known as neurogenic sarcoma or malignant neuroblastoma, rarely occurs in the head and neck area and can mimic benign proliferative lesions. The tumor develops from the peripheral nerve cells and can occur on the surface of the skin neurofibromas in von Recklinghausen disease or de Novo.

Aim: The aim of this paper was to present the case of the 86-year-old female patient with MPNST in the soft tissues of the right cheek.

Case study: 86-year-old female patient was admitted due to the right cheek tumor growing for about a month of the size 5 × 5 cm that caused significant asymmetry of the face.

Results and discussion: The patient was diagnosed on the basis of the history, craniofacial visualization, histopathological examination of the tumor sample and referred for surgical treatment and possible adjuvant radiotherapy. Extensive resection of the tumor was followed by a reconstruction of buccal defect with regional lobes. The postoperative course was uneventful. Adjuvant radiotherapy was not performed due to a number of aggravating systemic illness and old age of the patient. She is the subject of constant surgical–oncological follow-up medical care. No local recurrence or distant metastases were observed during follow-up.

Conclusions: MPNST rarely occurs in the head and neck region. This kind of sarcoma can cause problems in the process of diagnosis and therapy. Resection is the primary treatment in case of MPNST. Due to high incidence of local recurrence and distant metastases, the patients treated for MPNST require continuous outpatient follow-up after the treatment.

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1. Introduction

A malignant peripheral nerve sheath tumor (MPNST) belongs to a group of sarcomas derived from Schwann cells, perineural or intraneural fibroblasts.^{1–3} This cancer can develop from skin neurofibromas common in Recklinghausen disease.^{4,5} MPNST is rare and represents about 5%–10% of all cases of soft tissue sarcoma, and the incidence in the head and neck is estimated at less than 15% of cases.⁶ Most cases occur in adults in their 40s or 50s. Patients with MPNST on NF-1 are usually 10 years younger than the subjects that do not show the characteristics of von Recklinghausen disease.^{4,5} The most common MPNST is described as non-characteristic slowly growing tumor usually located within the trunk and extremities. Other symptoms of the tumor include pain, paresthesia and motor function disorders, which may arise at a later stage in the area supplied by the nerve associated with tumor. The cases of craniofacial tumors have been reported in the bones, lips and oral mucosa.⁷ The diagnosis is based on history, laboratory tests and histopathological examination extended with immunohistochemical staining. The diagnosis can often be difficult.^{8,9} The treatment of choice is surgery involving radical removal of tissue with a wide margin more than 2 cm. Postoperative radiotherapy is recommended. This type of cancer has a tendency to recurrence.

Cases of multiple operations and surgical procedures after the removal of MPNST were described.

2. Aim

The aim of this paper was to present the case of 86-year-old woman with a rare location of the tumor in the head and neck as MPNST cases are not commonly described in the specialist literature as well as the difficulties in diagnostic and therapeutic process of this sarcoma.

3. Case study

Patient, 86-year-old woman, was admitted due to the right cheek tumor growing for about a month of the size 5 × 5 cm that caused significant asymmetry of the face. The ulceration was found in the central part of the lump caused by the expanding tumor. The tumor covered the full thickness of the cheek and infiltrated all the layers, causing coating of buccal mucosa in the right cheek which was identified in intraoral examination. The skin surrounding the tumor was flushed and inflamed. In addition, the patient complained of pain and bleeding from the tumor surface (Fig. 1). It was found in the medical history that the patient was treated for hypertension, heart failure, diabetes type II. Senile dementia was observed. Neurofibromatosis was excluded after the clinical examination and the history taking. The patient underwent a diagnostic ultrasound examination of the neck, the lung X-ray, fine needle aspiration biopsy of the lymph nodes. The ultrasound showed the presence of enlarged neck lymph nodes, which are described as 'reactive nodes.' Fine needle aspiration biopsy of these nodes was performed and



Fig. 1 – Tumor of the full-thickness of the right cheek in 86-year-old woman.

confirmed the absence of metastases. The biopsy of the tumor and histopathological verification was conducted to establish the diagnosis. The result of the histopathological examination at the Department of Pathology: malignant peripheral nerve sheath tumor type with local epithelioid differentiation. The immunohistochemical stainings showed positive results for protein S-100, Vimentin. Ki-67 index was 50%, SMA and CD31 were focally positive (Figs. 2–7). Craniofacial MRI was conducted due to the location of the tumor in the soft tissues of the cheek. MRI was discontinued due to the deterioration of the general state of the patient during the exam.

After oncological consultation and preoperative preparation, the patient was qualified to remove a tumor from her right cheek with a partial resection and simultaneous reconstruction of postoperative defect with a lobe of tissue from skin and muscle based on the sterno-cleido-mastoid muscle and chick lobe.

In course of the operation enlarged neck lymph nodes – group II were found. Lymphangionectomy was performed, the nodes were subjected to histopathological verification. The patient managed the operation well. Postoperative healing proceeded without complications (Fig. 8). Histopathological examination at the Department of Pathology, confirmed the diagnosis of MPNST, lymph node showed no malignant metastases. Due to the age and systemic comorbidities the patient was disqualified from adjuvant radiotherapy. She remains in constant regular surgical–oncological follow-up. No local recurrence or distant metastases was found during follow-ups. Local condition without lesions.

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