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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

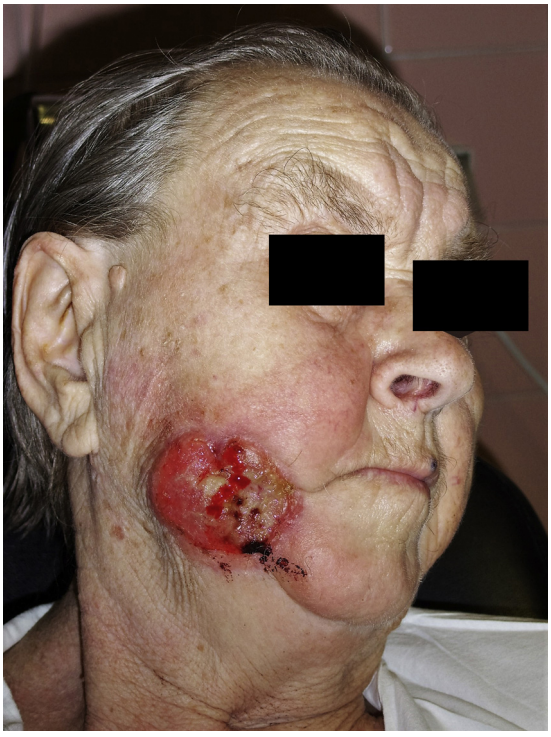

Fig. 1 – Tumor of the full-thickness of the right cheek in 86-year-old woman.

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.

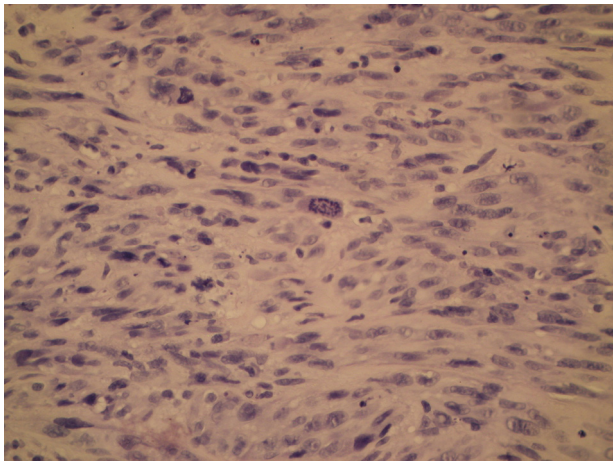

Fig. 2 – Poorly differentiated neoplasms with multiple mitotic figures (H&E staining, magnification 40×).

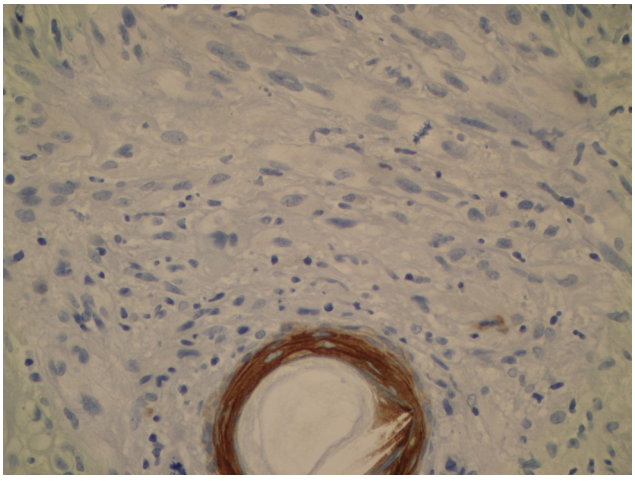

Fig. 3 – CK PAN negative reaction. Positive internal control – skin appendage (magnification 40×).

4. Results and discussion

MPNST is a sarcoma derived from the sheath of nerve cells.[1–3,10] It is rare in the region of the head and neck. It constitutes 5%–12% of malignant cancer cases in the soft tissue.[1,5] The incidence of MPNST in the general population is estimated at 0.001%.[5] A significant number of cases are recorded in the population of people with von Recklinghausen disease, which amounts even 40%–50% of the reported MPNST cases.[4,5] This type of neoplasm does not show gender predilection.[7,11] It was found, however, that people with von Recklinghausen disease are subject of MPNST about 10 years earlier (30–35 years of age), compared with those in whom the disease progresses de novo (40–46 years of age).[4,5] Although the incidence in the head and the neck was estimated at 8%–16%, the tumor occurs very rarely in the area of the face and the mouth. Yet, in specialized literature only isolated cases located within the jaw, lips, mucous membranes of the cheek and tongue were described.[4,6,7,12] In the presented case of a patient with MPNST, it was located in the soft tissues of the cheek, the neoplasm infiltrated full thickness of the cheek, it was very extensive and characterized by the absence of sharp borders.

It can be concluded from the descriptions presented in the literature that MPNST is most often characterized by a slow, asymptomatic growth, which is confirmed in the observed case. The time of tumor growth (one month) declared by the patient was recognized as false due to dementia. Only when the tumor reaches a larger size and ulceration and bleeding occur. Moreover, pain, paresthesia, or loss of sensation in the area supplied by the nerve occurred.

The most commonly reported tumor location is related to the trunk and proximal parts of the limbs.[10,13] Somewhat less frequently in the literature were presented tumors located in the subcutaneous tissue, mediastinum or the retroperitoneal space.[10,12] MPNST tumor occasionally develops in the parenchymal organs.[10] The rapid growth of neurofibromas in patients with neurofibromatosis type I (NF I), pressure or invasion of adjacent structures and tissues, pain and bleeding are the symptoms of malignancy and MPNST development should be suspected. In case of the presented patient, typical symptoms of advanced tumor were observed i.e. large size of the tumor bleeding from ulcerous surface and itching and burning. Imaging examinations are used for the diagnostic–therapeutic purposes, which allow to determine the extent of and location of the tumor, its relation with the surrounding tissue and anatomical structures. The MPNST diagnosis is determined by histopathology sample taken from the tumor. Since the histopatological picture is often not very distinctive, immunohistochemistry for the presence of protein S-100 (positive result in 50%–90% of cases), vimentin, glial fibrillary acidic protein (GFAP), neuron specific enolase (NSE), myelin basic protein (MBP) are advisable. In the presented case the diagnosis was established in the biopsy. There were difficulties in establishing the extent of the cancerous tissues especially in the cheek because the MRI had to be discontinued due to cardiopulmonary disorder, which occurred in a patient during the test. MPNST treatment of choice is surgery and consists of the removal of the tumor with a wide margin of healthy tissue. Often, it is necessary to extend the surgery and excise muscles adjacent to the tumor, nerves and blood, including bone resection. In addition, supplementary radiation therapy is recommended.[13,14]

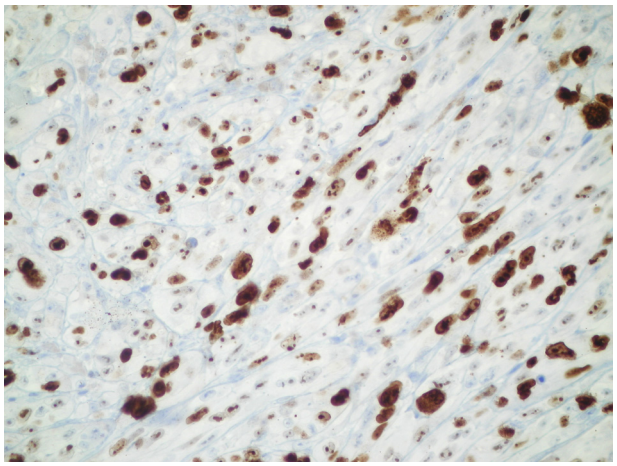

Fig. 4 – Ki-67 high index (magnification 40×).

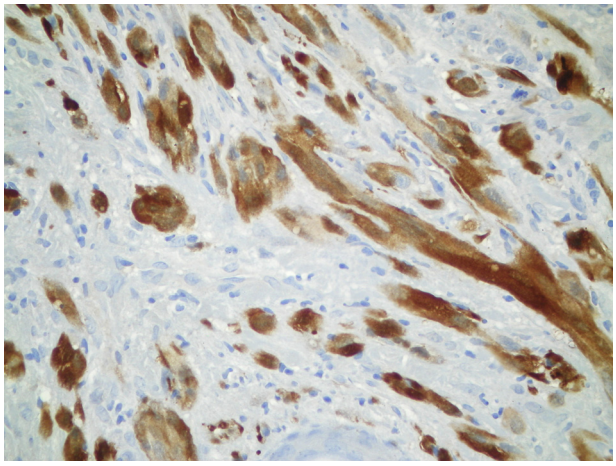

Fig. 5 – S100 strong nuclear and cytoplasmic positive reaction in tumor cells (magnification 40×).

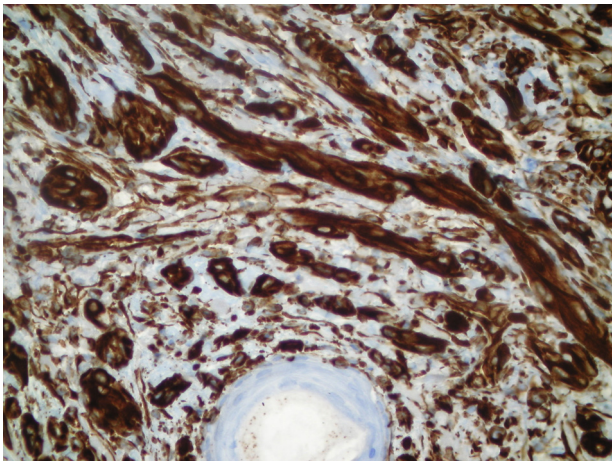

Fig. 7 – Vimentin strong and diffused positive reaction (magnification 40×).

Radiation therapy and chemotherapy may be used in the palliative treatment of advanced non-operative MPNST cases to reduce pain and prevent distant metastasis. Adjuvant radiotherapy was not performed in the presented patient due to a general condition, cardiopulmonary disorders, diabetes, hypertension and senior age of the patient.

The prognosis in cases of MPNST depends on many factors: tumor size, its location, preserving surgical margins after excision and the fact whether the tumor was primary or recurrent lesion and coexistence of NF I. Generally, five-year survival prognosis is estimated at 16%–52%.[4,5] Local recurrences occur in 40%–42% of cases and distant metastases are found in 65% of patients. Metastasis usually occurs through blood vessels to the lungs and bones. Adverse prognosis established in people with NF I and five year survival is estimated in this group of patients at around 20%. Due to the incidence of local recurrence and distant metastases, patients treated for MPNST require continuous outpatient follow-up after treatment. The patient presented in this paper in two-year period after surgery had no local recurrence or distant metastasis.

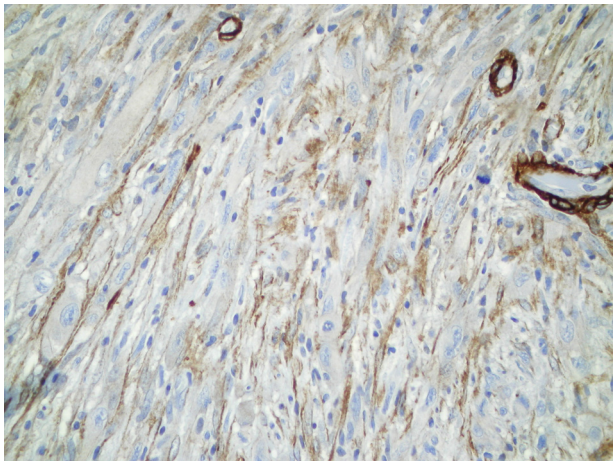

Fig. 6 – SMA stain rather week but positive reaction (magnification 40×).

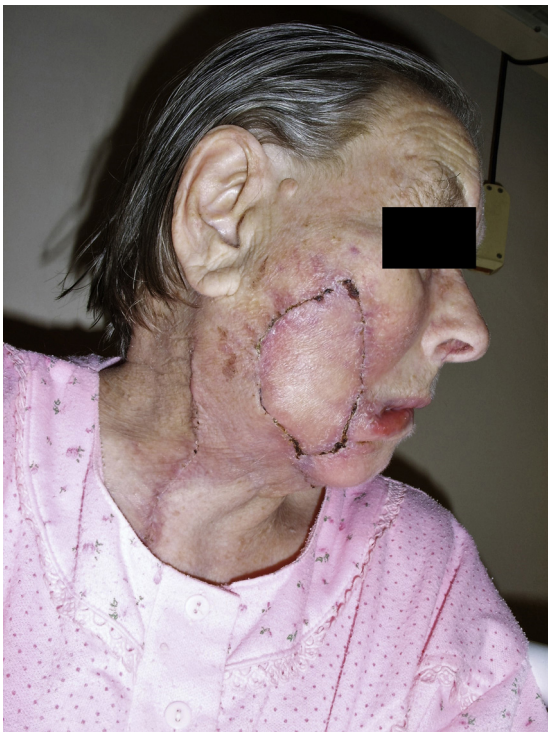

Fig. 8 – Status after resection of MPNST in the right cheek, regional lymphangionectomy and reconstruction of postoperative defect with a lobe of tissue from skin and muscle.

5. Conclusions

(1) MPNST rarely occurs in the head and neck region.
(2) This kind of sarcoma can cause problems in the process of diagnosis and therapy.

(3) Resection is the primary treatment in case of MPNST.
(4) Due to high incidence of local recurrence and distant metastases, the patients treated for MPNST require continuous outpatient follow-up after the treatment.

Conflict of interest

None declared.

REFERENCES

1. Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: diagnostic overview and update on selected diagnostic problems. *Acta Neuropathol.* 2012;123:295–319.
2. Rekhi B, Ingle A, Kumar R, DeSouza MA, Dikshit R, Jambhekar NA. Malignant peripheral nerve sheath tumors: clinicopathological profile of 63 cases diagnosed at a tertiary cancer referral center in Mumbai. *Indian J Pathol Microbiol.* 2010;53:611–618.
3. Kar M, Deo SV, Shukla NK, et al. Malignant peripheral nerve sheath tumors (MPNST) – clinicopathological study and treatment outcome of twenty-four cases. *World J Surg Oncol.* 2006;4:55.
4. Öztürk Ö, Tutkun A. A case report of a malignant peripheral nerve sheath tumor of the oral cavity in neurofibromatosis type 1. *Case Rep Otolaryngol.* 2012. http://dx.doi.org/10.1155/2012/936735.
5. Doorn PF, Molenaar WM, Buter J, Hoekstra HJ. Malignant peripheral sheath tumors in patients with and without neurofibromatosis. *Eur J Surg Oncol.* 1995;21:78–82.
6. Fernandes AM, Rodrigues Johann ACB, da Silveira-Júnior JB, Ferreira de Aguiar MC, Vieira do Carmo MA, Mesquita RA. Malignant peripheral nerve sheath tumor of the tongue. *Oral Oncology Extra.* 2006;42:210–212.
7. Salehinejad J, Nasseh A, Jafarian AH, Bashardoust N. Malignant peripheral nerve sheath tumor in the maxilla: report of a rare case. *J Dent Mater Technol.* 2013;2:34–37.
8. Ducatman BS, Scheithauer BW, Piepgras DG, Reiman HM, Ilstrup DM. Malignant peripheral nerve sheath tumor. A clinicopathologic study of 120 cases. *Cancer.* 1986;57:2006–2021.
9. Evans DGR, Baser ME, McGaughran J, Sharif S, Howard E, Moran A. Malignant peripheral nerve sheath tumors in neurofibromatosis 1. *J Med Genet.* 2002;39:311–314.
10. Gryglicka B, Węgrzyn-Szkutnik I, Milanowski J, et al. Malignant peripheral nerve sheath tumor associated with clear cell renal cell carcinoma – case report. *Pneumonol Alergol Pol.* 2012;80:269–274 [in Polish].
11. Cashen DV, Parisien RC, Raskin K, Hornicek FJ, Gebhardt MC, Mankin HJ. Survival data for patients with malignant schwannoma. *Clin Orthop Relat Res.* 2004;426:69–73.
12. Yamazaki H, Tsukinoki K, Shimamura K, Kaneko A. Malignant peripheral nerve sheath tumor with perineural cell differentiation arising from the tongue. *Oral Oncol Extra.* 2005;41:77–80.
13. Wroński K. Malignant peripheral nerve sheath tumor (MPNST) – case report and review of literature. *New Med.* 2015;1:3–5.
14. Ferner RG, Gutmann DH. International consensus statement on malignant peripheral nerve sheath tumors in neurofibromatosis. *Cancer Res.* 2002;62:1573–1577.