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Malignant tumor of the peripheral nerve sheaths in the parotid gland – a case report

Złośliwy guz osłonek nerwów obwodowych w obrębie ślinianki przyusznej – opis przypadku

Authors' Contribution:
A – Study Design
B – Data Collection
C – Statistical Analysis
D – Manuscript Preparation
E – Literature Search
F – Funds Collection

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ABSTRACT:

Introduction: Neoplasms of the parotid glands constitute about 6% of head and neck tumours, the most common of which are: adenoma multiforme and Warthin's tumor. Schwannoma is benign, encapsulated tumor of the nerve cells (lemocytes, Schwann cells), most often it occurs in the trunk, head, extremely rarely observed in the parotid gland. 9% Schwannomas derives from the facial nerve sheath, constituting from 0.5–1.2% of all salivary gland tumors. The literature describes 80 cases of intraparotid Schwannoma. Malignant Schwannoma (MPNST) account for 5% to 10% of all soft tissue sarcomas. High-grade MPNST tumors are aggressive, with a tendency to relapse and metastasis.

Case report: We present a case report of a 84-year-old female patient presented to the Head and Neck Oncology Clinic of the Medical University of Lodz, due to the painful tumor of the left parotid region. The tumor appeared six months before hospitalization. During the physical examination, there was a polycyclic tumor with reduced mobility, the skin on the tumor was slightly red, lymph nodes uninvolved, facial nerve function preserved. BACC revealed tumour tissue composed of sheets and spindle-shaped cells. The patient was qualified for surgical treatment. Under the general anesthesia the superficial lobe of the parotid gland along with the tumor was removed. After the procedure, no facial nerve palsy was found. The patient did well post-operatively and was discharged home on the 8th day after surgery. Immunohistochemical staining showed the tumour cells to be diffusely and strongly immunoreactive for S-100 protein and Ki67 40–50%. Strong and diffuse staining for S-100 protein were consistent with the malignant peripheral nerve sheath tumour (MPNST). The patient was referred for radiotherapy, due to the postoperative outcome and advanced age, no adjuvant treatment was proposed. The patient has been in observation for 1 year. No relapse was found in the follow-up studies.

KEYWORDS: auriculotemporal nerve, malignant tumor, parotid gland, peripheral nerve sheaths tumor, Schwannoma

STRESZCZENIE:

Wprowadzenie: Guzy ślinianki przyusznej stanowią ok. 6% nowotworów głowy i szyi. Najczęstsze z nich to: gruczolak wielopostaciowy i guz Warthina. Schwannoma jest zmianą wywodzącą się z komórek osłonki nerwów obwodowych (lemocytów, komórek Schwanna); najczęściej dotyczy tułowia i głowy, wyjątkowo rzadko obserwowany jest zaś w śliniance przyusznej. 9% Schwannoma pochodzi z osłonki nerwu twarzowego, stanowiąc od 0,5–1,2% wszystkich guzów ślinianek. W literaturze opisano 80 przypadków wewnątrzśliniakowych Schwannoma. Transformacja złośliwa dotyczy 14%. Złośliwe Schwannoma (MPNST) stanowią od 5% do 10% wszystkich mięsaków tkanek miękkich. Nowotwory MPNST o wysokim stopniu złośliwości są agresywne, z tendencją do wznowy oraz przerzutowania.

Opis przypadku: Prezentujemy opis przypadku 84-letniej pacjentki, która zgłosiła się do Kliniki Nowotworów Głowy i Szyi UM w Łodzi z powodu bolesnego guza lewej okolicy przyusznej. Wspomniana zmiana pojawiła się 6 miesięcy przed hospitalizacją. W badaniu przedmiotowym stwierdzono policykliczny guz o ograniczonej ruchomości, skóra nad guzem była lekko zaczerwieniona, węzły chłonne niepowiększone, funkcja nerwu twarzowego zachowana. W BACC atypowe komórki wrzecionowate, budzące podejrzenie rozrostu złośliwego. Chora została zakwalifikowana do leczenia operacyjnego. W znieczuleniu ogólnym usunięto płat powierzchowny ślinianki przyusznej wraz z guzem i zmienioną skórą. Po zabiegu nie stwierdzono porażenia nerwu twarzowego. Przebieg pooperacyjny prawidłowy, pacjentka wypisana do domu w 8. dobie po zabiegu w stanie ogólnym i miejscowym dobrym. Wynik badania histopatologicznego materiału pooperacyjnego: dobrze odgraniczony wrzecionowaty guz z polami martwicy, profil immunohistochemiczny: S100+; Ki67 40–50%. Rozpoznanie – MPNST. Pacjentka została skierowana

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na konsultację radioterapeutyczną. Z uwagi na wynik pooperacyjny oraz zaawansowany wiek, nie zaproponowano leczenia adiuwantowego. Chora pozostaje w obserwacji 1 rok. W badaniach kontrolnych nie stwierdzono wznowy.

SŁOWA KLUCZOWE: guz nerwów obwodowych, nerw uszno-skroniowy, Schwannoma, złośliwy guz ślinianki przyusznej

LIST OF ABBREVIATIONS

BACC – targeted fine needle aspiration biopsy
CRP – C-reactive protein
MPNST – malignant peripheral nerve sheath tumors
MR – magnetic resonance imaging
OB – Biernacki reaction
USG – ultrasound examination

INTRODUCTION

Peripheral nerve tumors derive from the neuroectoderm and are classified as soft tissue tumors [1]. In clinical practice, we most often observe benign peripheral nerve sheath tumors (Schwann cells, lemmocytes) such as: neurilemmoma and neurofibroma [2–7].

Malignant peripheral nerve sheath tumors, or MPNST constitute 5–10% of all soft tissue sarcomas [8]. The most common location of those tumors are the limbs and the torso (12–19% of cases), with the head and neck region of HN-MPNST covering only 4% of all sarcomas [9–11]. Nearly 50% of MPNST cases are associated with type 1 neurofibromatosis (NF1) and 10% are linked with ionizing radiation [12]. Other MPNSTs develop sporadically, with no prior history of radiation or genetic predisposition [8]. High-grade MPNSTs are characterized by aggressive clinical course, high local recurrence rate (28–75%) [9, 14–17] and distant metastases (28–34%) [9, 14].

Previous studies have demonstrated that the 5-year survival of patients with head and neck MPNST ranged from 20 to 50% [16, 18–20], with overall survival associated with age, tumor size, and stage at the time of diagnosis, insufficient tumor resection margin, early relapse and complementary radiotherapy [18, 20]. No relationship between the tumor and gender was found [13].

We present a case of MPNST arising from the parotid gland in a patient treated at the Head and Neck Cancer Clinic of the Medical University of Lodz.

CASE STUDY

An 84-year-old woman reported to the clinic due to a painful tumor in the left parotid gland. The lesion presented itself 6 months before hospitalization. The patient connected its occurrence with the trauma of this area, which had occurred six years earlier.

Physical examination demonstrated a polycyclic lesion with limited mobility. The skin above it was slightly red, not movable in relation to the tumor. Palpation showed no swollen lymph nodes and facial nerve function was preserved (Fig. 1.). Apart from this ENT examination, there were no deviations from the normal state.

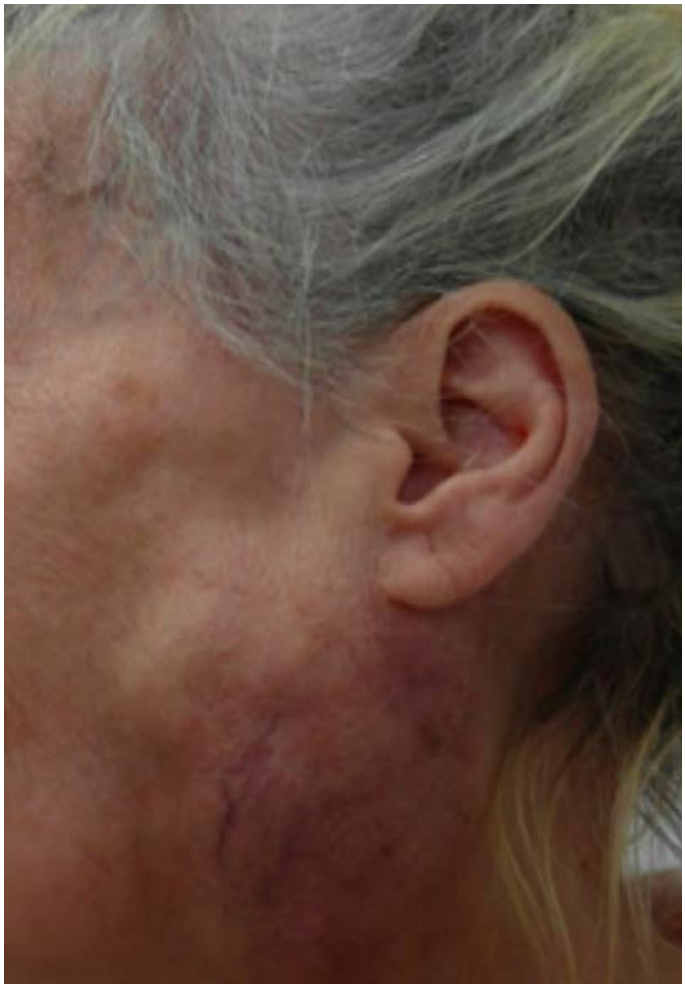

Fig. 1. Tumor of the left parotid gland. Reddened and inflamed skin visible under the tumor.

Laboratory tests, i.e. blood cell count, CRP, OB, were within normal limits. Chest x-ray also revealed no pathological changes.

Targeted fine needle aspiration biopsy (BACC) revealed few, diffused, mainly atypical spindle cells, suspected of malignant growth. Magnetic resonance imaging (MR) of the craniofacial structures and neck showed a well-defined polycyclic tumor of the superficial lobe of the left parotid measuring 27 x 26 x 37 (Fig. 2a.–b.). No swollen lymph nodes were found in the neck.

The patient was qualified for superficial parotidectomy on the left side. The superficial lobe of the left parotid was excised along with the tumor and the changed skin under endotracheal anesthesia. There was no infiltration or connectivity of the tumor with the facial nerve. Intraoperative facial nerve monitoring was used during the procedure. Facial nerve function was preserved after the procedure. Histopathological examination of postoperative specimen revealed a well-defined fusiform tumor with necrotic fields

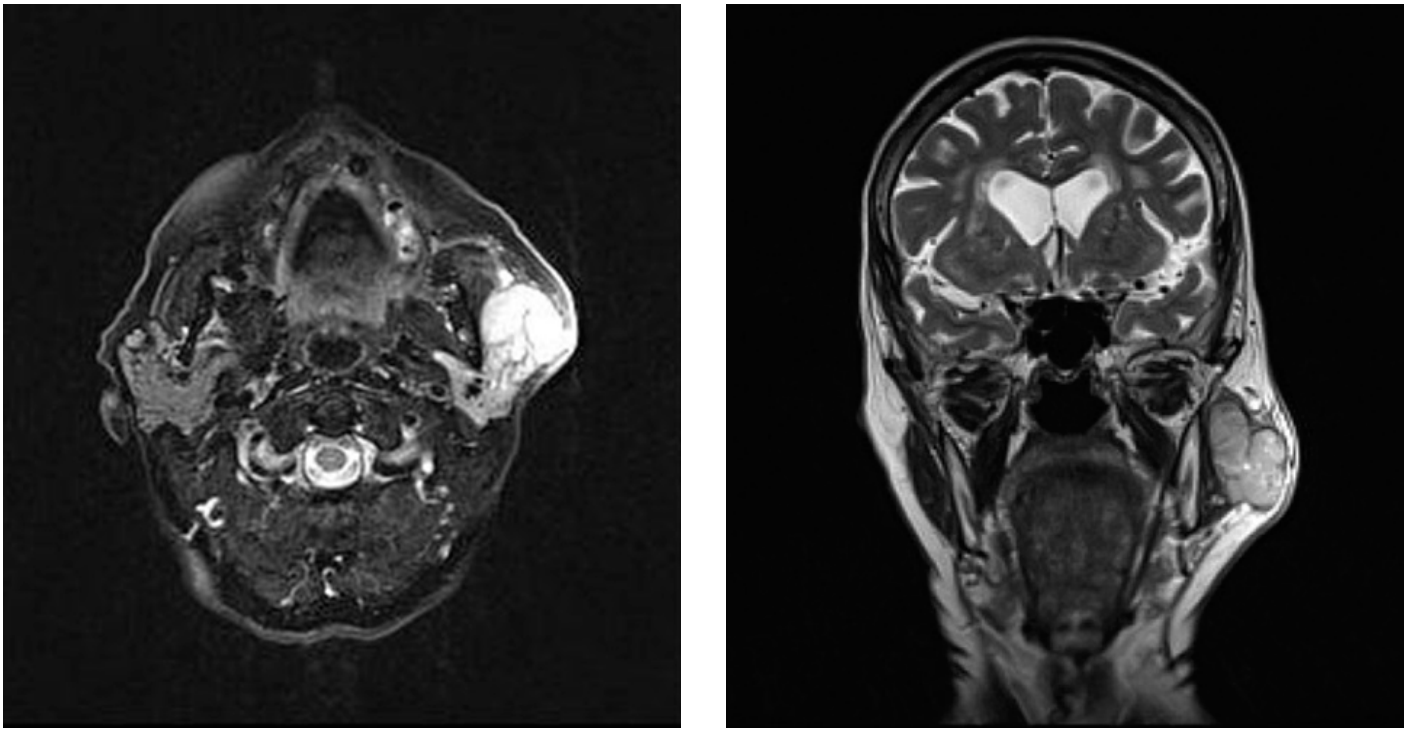

Fig. 2. A polycyclic tumor in the anterior part of the superficial lobe of the left parotid, with a smooth contour, without infiltration of the masseter muscle and the cortex of the mandible, without penetration into the deep lobe.

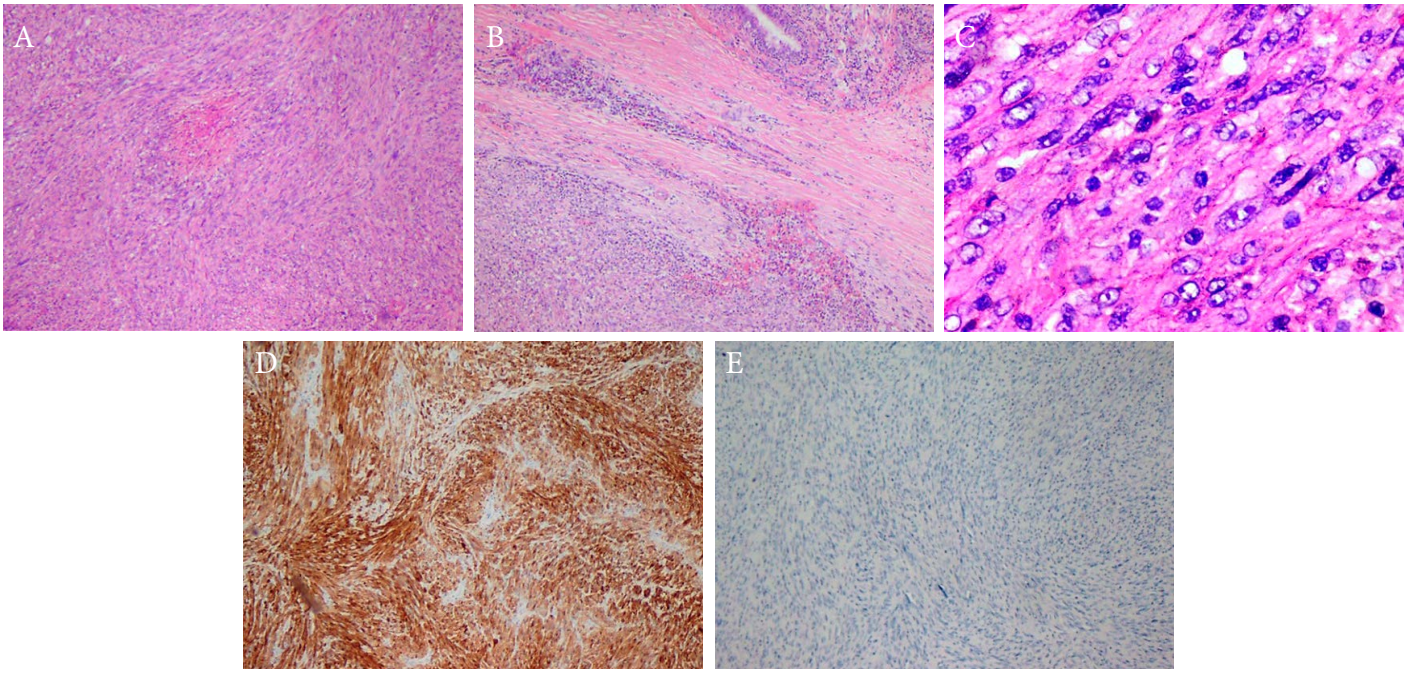

Fig. 3. Histopathological preparations of the removed tumor: (A) visible hypo- and hypercellular areas, H + E staining, 10x magnification; (B) visible scattered pleomorphic cells H + E staining, 40x magnification; (C) visible hypercellular area with numerous spindle cells H + E staining, 100x magnification; (D) immunohistochemistry – positive reaction to S100; (E) immunohistochemistry – positive response to Ki67.

(Fig. 3a.–c.). The immunohistochemical profile of neoplastic cells: S100 and Ki67-positive (Fig. 3d.–e.). In view of the radical nature of the procedure and the advanced age of the patient from complementary radiotherapy, she has been under the constant care of an outpatient clinic for a year. Control imaging (USG, MR) revealed no tumor recurrence.

DISCUSSION

MPNST is a rare, aggressive cancer associated with a high rate of relapse, distant metastasis and mortality. Diagnosis can be quite difficult, especially in the head and neck region, and is a challenge for both the ENT specialist and cooperating diagnosticians,

i.e. pathologist and radiologist. The rare prevalence of MPNST in the head and neck, especially in the parotid gland, may sometimes lead to dormant alertness during diagnosis.

MPNST usually presents as a poorly-encapsulated tumor, with blurred margins of its borders. Histologically, two types of tumor cell system are distinguished: monomorphic spindle cells arranged in cellular "herringbone" (storiform pattern) or displaying Schwann cell-like qualities, with a characteristic palisade cell nuclei formation [21].

The final diagnosis of MPNST is based on immunohistochemistry. A panel of tests for the presence of S100 protein, smooth muscle actin (ACTA2), CD68, pancytokeratin and Ki67 should be performed [22]. MPNST demonstrates a strongly positive result on the S100 protein. The proliferation marker Ki-67 is usually low and fluctuates around 1%. Staining of ACTA2 aims to exclude myoma from smooth cells, and CD68 – xanthomatous histiocytic lesion in schwannoma.

Pancytokeratin staining is performed to exclude spindle cell carcinoma or other epithelial tumor. Other types of staining, such as calretinin, may be useful in differentiating MPNST from neurofibroma [9].

Differential diagnosis should consider multiform adenoma, myoepithelial cell carcinoma, spindle cell carcinoma and spindle cell melanoma.

The treatment of choice is radical resection of tumor with adjuvant chemoradiotherapy.

In surgical trials, special attention should be paid to connectivity of the tumor with the facial nerve, where the use of intraoperative monitoring of the facial nerve is helpful. In cases where there is no direct communication with the facial nerve, no infiltration is found, and the tumor resection provides a sufficient oncological margin, the behavior of the facial nerve should be considered.

In the case described herein, no connection with the facial nerve was found during the surgery, while connectivity with the skin with the involvement of the superficial lobe and severe pain may indicate that the starting point was the sensory nerve, most probably the auriculotemporal.

Despite comprehensive treatment with comprehensive surgery, resection and radiation therapy, the tumor shows an aggressive clinical course, with local recurrence in 54% of cases, and distant lung and bone metastases in 65% of cases [23, 24]. Our patient did not receive adjuvant treatment due to the radicality of the excision and the advanced age of the woman. She remains under strict oncological control, and no recurrence was seen in follow-up examinations.

CONCLUSIONS

MPNST is a rare malignancy arising from cells of peripheral nerve sheath. MPNST is associated with neurofibromatosis type I (NF1). Diagnosis can be made on the basis of histopathological examination with a panel of immunohistochemical tests. The treatment of choice is radical tumor excision with adjuvant therapy. Due to the high rate of relapses, it is necessary to monitor patients after treatment.

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