

Difficulties in clinical staging of external auditory canal carcinoma – a case report

Trudności w ocenie zaawansowania raka przewodu słuchowego zewnętrznego – opis przypadku

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ABSTRACT:

Carcinoma of external auditory canal (EAC) is known to be very rare with a rate of 1–6 per million people. It affects mostly people over the age of 50 regardless of gender. The most common type of carcinoma of EAC is a squamous cell carcinoma (SCC). The aim of this paper is to present the case of 56-year-old woman with carcinoma of EAC. She was admitted to otolaryngology clinic due to ear buzzing lasting five months, periodic otorrhea and hearing deterioration in her right ear. In otoscopic examination there was identified a tumor in the right ear meatus. It filled almost the entire canal. Biopsy confirmed the presence of squamous-cell cancer. The tumor advancement was diagnosed using computer tomography of temporal bones, magnetic resonance imaging of head and ultrasonography of neck. Imaging examinations indicated T3 No stage. Doctors decided to perform lateral petrosectomy with elective neck dissection. During the operation, the results of histopathological examination of frozen specimens from marginal tissues revealed the invasion of anterior wall of ear canal, capsule of temporomandibular joint and parotid gland. The resection eventually included also parotid gland and temporomandibular joint with the caput of mandible. After recuperation, the patient had complementary radiotherapy. The presented case shows that precise imaging is necessary before surgical treatment, but unfortunately the invasion of cancer may not be estimated properly. Based on literature the best prognosis concerns wide resection with complementary radiotherapy, although the schedule of treatment is not determined.

KEYWORDS:

squamous cell carcinoma, external auditory canal, lateral petrosectomy, facial nerve

STRESZCZENIE:

Nowotwory przewodu słuchowego zewnętrznego (PSZ) są niezwykle rzadkie, występują z częstotliwością 1–6 przypadków na 1 000 000 pacjentów. Zazwyczaj dotyczą osoby w średnim wieku (50–60 lat), równie często kobiety, jak i mężczyźni. Najczęstszym typem histologicznym jest rak płaskonabłonkowy. Celem pracy jest omówienie przypadku raka PSZ u 56-letniej pacjentki leczonej w Klinice Otolaryngologii WUM. Pacjentka zgłosiła się z powodu trwającego od ponad 5 miesięcy szumu w prawym uchu, stopniowego pogorszenia słuchu i okresowego wycieku z prawego ucha. W badaniu otoskopowym uwidoczono guz w PSZ całkowicie wypełniający jego światło. Wykonano biopsję tkanek guza. Rozpoznano raka płaskonabłonkowego. Następnie wykonano tomografię komputerową kości skroniowych, rezonans magnetyczny głowy i badanie ultrasonograficzne szyi. Na ich podstawie ustalono stopień zaawansowania guza na T3 No. Zaplanowano zabieg petrosektomii bocznej z elektywną operacją węzłów chłonnych szyi. W czasie resekcji śródoperacyjne badania histopatologiczne potwierdziły naciekanie przez raka przedniej ściany PSZ, jamy bębenkowej, szczytu piramidy kości skroniowej, torebki stawu skroniowo-żuchwowego oraz ślinianki przyusznej. Poszerzono resekcję o staw żuchwowo-skroniowy wraz z głową żuchwy oraz śliniankę przyuszną. Przypadek pokazuje, że badania obrazowe są niezbędne przed planowanym leczeniem operacyjnym, ale rozległość nacieku raka często jest niedoszacowana. Dostępne w literaturze wyniki badań sugerują, że najlepsze rokowania mają chorzy poddani rozległej resekcji, uzupełnionej radioterapią, choć dokładny schemat leczenia nie został do tej pory w pełni ustalony.

SŁOWA KLUCZOWE: rak płaskonabłonkowy, przewód słuchowy zewnętrzny, petrosektomia boczna, nerw twarzowy

INTRODUCTION

Malignant neoplasms of the external auditory canal (EAC) are very rare. It is estimated that 1 to 6 people per million suffer from EAC cancer. It represents only 0.2% of all head and neck malignancies and only 4% of external ear cancers [1-3]. It is most often diagnosed in people aged between 50-60 years, equally often in women and men [7,9]. Histologically the most common malignancies of the EAC are squamous cell carcinoma or adenoid cystic carcinoma, less often basal cell carcinoma or adenocarcinoma [8].

The most frequently reported initial symptoms of EAC malignancies include earache, discharge from the ear and feeling of a mass in the ear [4,5,18,19]. Less frequently: worsening of hearing, bleeding from the ear, dizziness and pruritus [1.18]. As tumor grows, swelling of the pinna area, paresis or facial nerve palsy may occur. [2,10,11,12,15].

Various classifications of EAC malignant tumors are available [6, 13]. The TNM scale developed at the University of Pittsburgh modified by Moody is the most widespread one (Table 1). It is considered the most reliable and predictive scale [1-4,8]. Determination of the T stage of tumor is performed on the basis of the assessment of the extent of infiltration in imaging studies, such as computed tomography of the temporal bones and magnetic resonance with assessment of the parotid gland, temporomandibular joint, infratemporal fossa and meninges.

The prognosis drastically decrease in advanced stages of the disease. The value of the 5-year survival rate is estimated at 85-100% in patients with stage I or II tumors, 50-68.8% in stage III and only 19.6-30% in stage IV [1, 3,4,11-14].

Surgical resection is the treatment of choice of malignant neoplasms of the external auditory canal [8, 10]. Surgical treatment is complemented with adjuvant radiotherapy and/or chemotherapy for higher stages. In case of inoperable tumors, palliative chemotherapy is used [5, 11]. Depending on the stage of cancer different surgical methods are used:

- wide resection of EAC - resection of the skin, cartilage and parts of EAC bones
- lateral temporal bone resection (LTBR) - resection en bloc of the EAC anterior to the facial nerve, tympanic membrane, malleus and incus;
- subtotal temporal bone resection (STBR) - resection as in LTBR with resection of the labyrinth and/or cochlea;
- total temporal bone resection (TTBR) - resection as in STBR with resection of the temporal bone pyramid;

In case of cancer invasion to adjacent structures, the resection of the parotid gland, the pinna and the subcutaneous tissue is per-



Fig. 1. Otoscopic examination of the right external auditory canal with tumor masses.

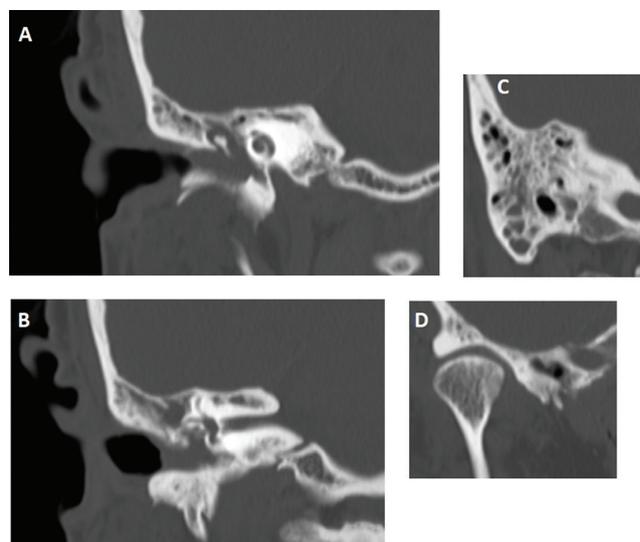


Fig. 2. Computed tomography of the right temporal bone. A. and B. Filled tympanic cavity C. Filled mastoid cells. D. Temporomandibular joint.

formed [1,3,4,15,16]. Resection of the neck lymph nodes should always be performed in the presence of clinically enlarged lymph nodes. In the clinical situation of N0, the decision about elective dissection of lymph nodes is made depending on the severity of the EAC cancer and its histopathological type.

CASE REPORT

A 56-year-old patient presented to the laryngological clinic with a history of ear buzzing for more than five months, hearing de-

Tab. I. University of Pittsburgh TNM staging system for carcinomas of the external auditory canal modified by Moody [1,2,8].

FEATURE	ADVANCEMENT	DESCRIPTION OF CLINICAL FEATURE ADVANCEMENT.
T	T1	Tumor limited to the skin of the external auditory canal without bony erosion or evidence of soft tissue extension
	T2	Tumor with limited external auditory canal bone erosion (not full thickness) or limited (< 0.5 cm) soft tissue involvement
	T3	Tumor eroding the osseous bone EAC (full thickness) with limited (< 0.5 cm) soft tissue involvement or tumor involving the middle ear and/or mastoid
	T4	Tumor eroding the cochlea, petrous apex, carotid canal, jugular foramen, or dura; or tumor with extensive soft tissue involvement (>5 mm); or with evidence of facial nerve paralysis
N	No	No regional lymph node metastasis
	N1	A single regional metastatic lymph node < 3 cm
	N2a	A single ipsilateral metastatic lymph node measuring 3–6 cm
	N2b	Several ipsilateral metastatic lymph nodes (< 6 cm)
	N2c	Contralateral metastatic lymph node
	N3	Metastatic lymph node > 6 cm
M	Mo	No distant metastasis
	M1	Distant metastasis
Stage	Stage I	T1NoMo
	Stage II	T2NoMo
	Stage III	T3NoMo
	Stage IV	T4NoMo lub N+ lub M+

terioration and periodic leakage from the right ear. Previously, she was treated several times both locally and systemically due to the external ear inflammation. She negated earaches or the presence of dizziness. The patient reported a long-term history of asthma and hypertension treatment.

The otoscopic examination revealed a tumor of the EAC completely filling the lumen of the canal (Figure 1). There were no clinical features of paresis or facial nerve paralysis. Tumor tissue biopsy was performed. The result of histopathological examination confirmed the presence of squamous cell carcinoma in the right EAC. Computed tomography of the temporal bones, magnetic resonance of the head, and ultrasound examination of the neck were performed (Figure 2). In computed tomography, on the right side, a hyperdense area of soft-tissue inflammation and fluid filling the mastoid cells, surroundings of the ossicles, the tympanic cavity and pneumatized cells of the temporal bone pyramid were described. Bone destruction or changes in bone structure of the inner ear were not found, however, the infiltration of tissues exceeded 5 mm in thickness. MRI examination confirmed the extent of the infiltration described in CT and no evidence of infiltration of the meninges or right parotid gland was found. In the ultrasound examination of the neck, the cervical lymph nodes were not enlarged. Tumor stage T3 was determined.

The patient was qualified for surgical treatment. Lateral temporal bone resection (LTBR) was performed. The skin of the external ear

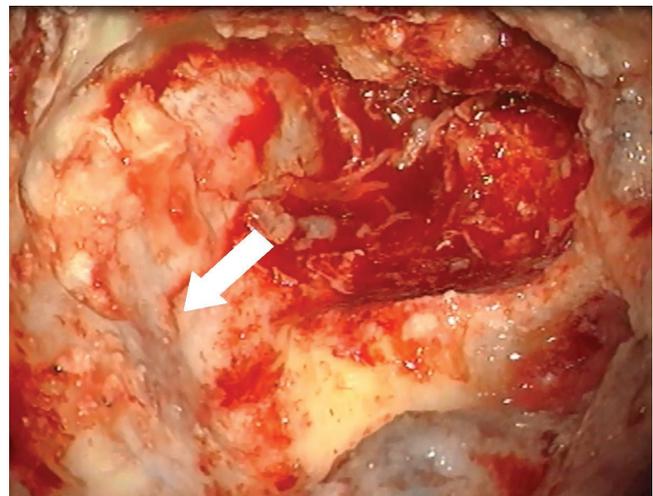


Fig.3. Intraoperative image of the middle cranial fossa. The arrow points to the meninges of the middle cranial fossa.

canal was cut 1.5 cm from the tumor infiltration and sutured back after reversing. The mastoid process was drilled to the level of the facial nerve, the tumor was removed from the antrum and from the cells directly adjacent to the base of the middle cranial fossa. During tumor resection, the meninges of the middle and posterior cranial fossa without neoplastic infiltration were exposed. The tumor mass was resected from the tympanic cavity. The tumor tissues penetrated to the apex of the pyramid. The completeness of

Tab. II. Five-year survival rates depending on the stage of cancer, histopathological type and treatment modality by different authors (CRT – chemoradiotherapy, RT – radiotherapy). [3,5,7,11,12]

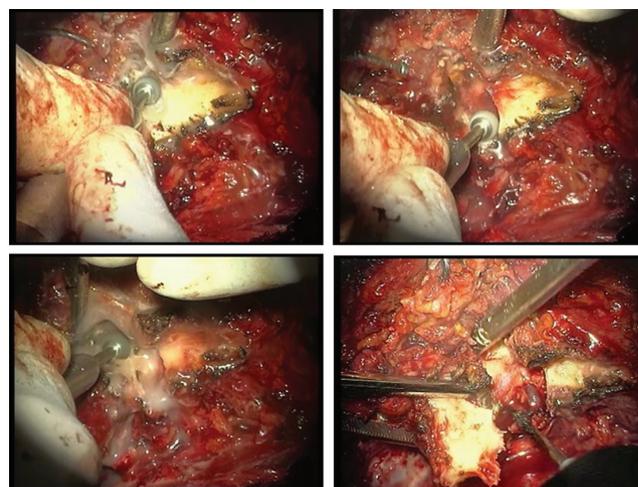
AUTHOR	SAMPLE SIZE	STAGE [FIVE-YEAR SURVIVAL]	HISTOPATHOLOGICAL TYPE [FIVE-YEAR SURVIVAL]	TYPY OF THERAPY (5-YEAR SURVIVAL)
Duro et al., 2007 [3]	34	Stage I and II (87%) Stage III and IV (21%)	Adenocarcinoma (100%) Squamous cell carcinoma (43%) Basal cell carcinoma (33%)	
Hosokawa et al., 2014 [5]	23	Stage I (100%) Stage II (75%) Stage III (100%) Stage IV (33%)	Squamous cell carcinoma (61,5%) Other carcinomas (100%)	Surgery (100%) Suregry + CRT/RT (100%) CRT (0%)
Gurgel et al., 2009 [7]	215	All stages (36,4%)	Squamous cell carcinoma (23,9%) Other carcinomas (62,9%)	Surgery (69,2%) Surgery + RT (26,4%) RT (14,6%)
Yin et al., 2006 [11]	96	Stage I (100%) Stage II (100%) Stage III (67,2%) Stage IV (29,5%)	Squamous cell carcinoma (66,8%)	
Zhang et al., 1999 [12]	33	Stage I (100%) Stage II (100%) Stage III (68,8%) Stage IV (19,6%)		Surgery + RT (59,6%) RT (28,7%)

the resection was confirmed by the histopathological examination of frozen specimens intraoperatively. The tumor infiltrated the anterior wall of EAC. The result of intraoperative histopathological examinations of the temporomandibular joint capsule confirmed the presence of cancer cells. The caput of the mandible was resected (Figure 4). The facial nerve was dissected and denudation was performed. Complete parotidectomy was carried out. During the surgical debridement of the temporal bone from the tumor and removal of the salivary gland, disruption of the facial nerve occurred. The sutures 10-0 were placed on the facial nerve at the level of the 2nd geniculum (Figure 5). Next, the lymph nodes from the I and II neck region were removed together with the right submandibular gland. Fat tissue collected from the abdominal wall was inserted into the post-operative cavity in the temporal bone.

Histopathological examination confirmed the diagnosis of squamous cell carcinoma.

Postoperatively, the patient had paralysis of the right facial nerve. The patient was started on moisturizing treatment of the anterior segment of the eyeball to protect the cornea. The postoperative wound was healing properly. After being discharged home, the patient was referred to the radiotherapy center for adjuvant treatment.

The presented case study of EAC cancer shows that despite a series of imaging tests, accurate assessment of the extent of infiltration was possible only intraoperatively. In case of stage III and IV of malignant neoplasms of the EAC, due to the progression of the tumor infiltration at the base of the skull or the temporomandibular joint, it is not possible to perform the classic onco-

**Fig. 4.** Intraoperative image from the resection of the mandible head.

logical “en bloc” surgery. The extent of surgical resection is based on the results of intraoperative histopathological examination of samples taken from the margins of the surrounding tissues.

DISCUSSION

Neoplasms of EAC are very rare diseases. There are reports of cases or series of patients available in the literature, but there is still a shortage of multicenter, randomized trials [1,8]. In most of the described cases, the correct diagnosis is made late because the symptoms are highly unspecific. The most common symptom, i.e. leakage from the ear, suggests inflammatory changes of the

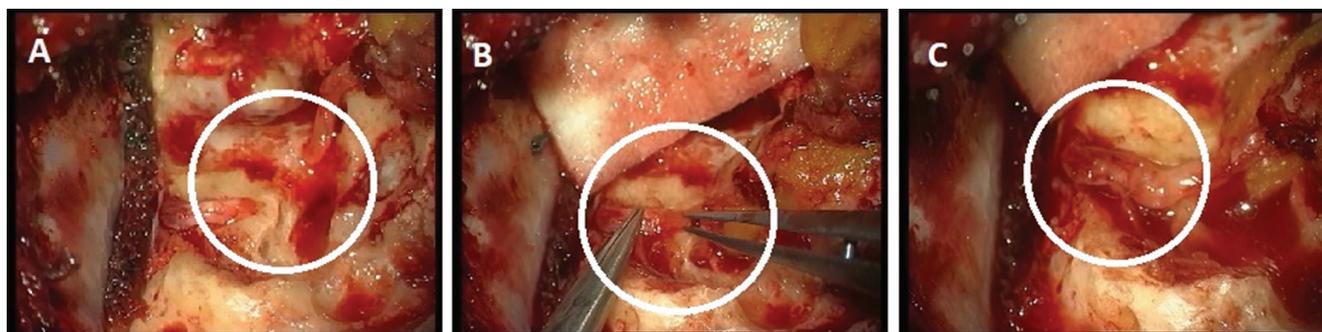


Fig. 5. Intraoperative images from the facial nerve reconstruction. A. Disruption of the facial nerve at the level of the 2nd geniculum of the facial nerve. B. Stitching. C. Facial nerve with stitches.

external or middle ear. Other symptoms are also more common in other diseases. Zhen et al., basing on the analysis of 16 case reports, concluded that these nonspecific symptoms contributed to an incorrect initial diagnosis in 68% of patients with cancer of the external auditory canal. According to the authors, the most frequent misdiagnosis is external otitis, chronic otitis media with effusion, cholesteatoma, papilloma, granulation tissue, neuralgia. This results in the diagnosis at an advanced stage of the disease, which is associated with a worse prognosis [1,3,11,19]. Also in the presented case study, the diagnosis was made after several months of inappropriate therapy. The important principle is that in case of standard otitis externa or middle ear inflammation treatment failure after 2-3 weeks, it is necessary to consider the biopsy [2]. It should also be remembered that the location of the lesion, the depth of collected samples and the size of the sample affect reliable biopsy results. [2]. It is worth noting that some of the misdiagnoses with long-term history are considered to be diseases associated with an increased risk of developing cancer. Such diseases include chronic otitis media or external otitis [1,3,5,10-12]. Previous radiation of the head and neck region is also a risk factor for cancer development [3, 19].

According to the literature, the most common type of EAC cancer is squamous cell carcinoma, which constitutes 60-70% of cancers [1,5,7,18,20]. The second most common type is adenoid cystic cancer, although in some publications it occurred with a higher frequency than SCC [2]. Less common malignancies in this location are basal cell carcinomas, adenocarcinomas or melanomas. The histological type of cancer correlates with prognosis. The highest rate of the 5-year survival is associated with adenoid cystic cancer. Squamous cell carcinoma has worse prognosis [3,5,7]. The stage of cancer of the auditory canal is also a prognostic factor. Selected results of 5-year survival rates depending on the type of cancer and its stage reported in the literature are presented in Table 2.

Diagnostic imaging is necessary to determine the extent of infiltration of EAC cancer. Computed tomography of the temporal

bones and base of the skull will be better at visualizing the destruction of bones, whereas the magnetic resonance imaging will show the infiltration of soft tissues: parotid gland, infratemporal fossa, and meninges [2].

No clear treatment regimen has been established due to the lack of randomized studies. Most authors agree that the treatment of choice is surgical tumor resection [1,2,8,10]. Early, radical surgical treatment is a beneficial prognostic factor. The extent of resection within the temporal bone remains a problem. The authors state that complete petrosectomy, especially in cases T3 and T4, improves the prognosis. Unfortunately, it is associated with more frequent postoperative complications and deterioration of the quality of life [1]. Goolding et al. recommend removal of the parotid gland and the mandible caput in all patients with stage IV, as infiltration of the infratemporal fossa is often undiagnosed in radiological exams [21]. Radiotherapy is recommended as an adjuvant treatment in any case of positive surgical margins [2,4,11,13]. Most authors recommend adjuvant radiotherapy also after surgical treatment of cancer in stages III and IV [11,15,19]. Combined treatment correlates with a statistically better prognosis than single-modality therapies, i.e. surgery or chemotherapy or radiotherapy [7,9]. Chemotherapy is used in palliative treatment.

CONCLUSION

Although the cancers of the external auditory canal are extremely rare, biopsy should be considered for cases of non-responsive inflammation of the external and middle ear. Various classifications of EAC carcinomas and the lack of standards of therapeutic treatment make it difficult to make unambiguous decisions. Imaging examinations are necessary before surgical treatment. Unfortunately the extent of cancer infiltration is often underestimated. Reports of results available in the literature suggest that patients undergoing extensive resection complemented with adjuvant radiotherapy have the best prognosis.

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