

Rare ceruminous gland-originated tumor of the external auditory canal: a case report

Rzadki nowotwór przewodu słuchowego wywodzący się z gruczołów woskowinowych – opis przypadku

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ABSTRACT: External auditory canal neoplasms comprise less than 1 percent of all head and neck tumors. They are ceruminous gland-originated tumors in 2.4 percent of cases. In the past, all these tumors were called ceruminomas. In 1972 Wetli et al. classified these neoplasms into four groups: ceruminous adenomas, ceruminous carcinomas, adenoid cystic carcinomas and pleomorphic adenomas. Ceruminous adenocarcinoma is very rare. It has no specific symptoms; therefore, an appropriate, quick diagnosis is difficult; it is very aggressive. Treatment should be based on tumor resection and followed by radiotherapy. We presented a case of a 56-year-old female with a mass in the external ear canal, who underwent diagnostic surgical excision of tumor in the external auditor canal. Postoperative histopathological diagnosis was ceruminous adenocarcinoma (complete excision) and the patient received radiation therapy. Follow-up CT and MR – performed one year after surgery - showed possible local recurrence of pathology. The patient underwent maximal surgical resection with subtotal petrosectomy but postoperative histological examination showed no malignant cells. Treatment of external auditory canal neoplasms should be based on radical surgical resection. Diagnostic imaging is very important in follow up examination but it can have limited value in terms of its specificity.

KEYWORDS: ceruminous adenocarcinoma, external auditory canal

STRESZCZENIE: Nowotwory przewodu słuchowego zewnętrznego stanowią <1% nowotworów złośliwych regionu głowy i szyi. 2,4% wszystkich nowotworów występujących w tej lokalizacji to nowotwory wywodzące się z gruczołów woskowinowych. W przeszłości określano je wspólną nazwą „ceruminoma”. W 1972 roku Wetli i wsp. na podstawie różnic histologicznych opracowali szczegółową klasyfikację, w której wyróżnili 4 kategorie: ceruminous adenomas, ceruminous carcinoma, adenoid cystic carcinomas i pleomorphic adenomas. Autorzy przedstawiają przypadek 58-letniej pacjentki z guzem przewodu słuchowego zewnętrznego, u której, w celach diagnostycznych, usunięto guzek przewodu słuchowego zewnętrznego ucha lewego. W pooperacyjnym badaniu histopatologicznym stwierdzono nowotwór ceruminous adenocarcinoma (zmiana usunięta z marginesem zdrowych tkanek). Chorą skierowano na uzupełniającą radioterapię. W kontrolnych badaniach MR i TK wykonanych po roku od zabiegu wysunięto podejrzenie wznowy procesu nowotworowego. Pacjentkę ponownie operowano, wykonano petrosektomię boczną, jednak w pooperacyjnym badaniu histopatologicznym wykluczono wznowę procesu nowotworowego. Leczenie nowotworów przewodu słuchowego wywodzących się z gruczołów woskowinowych powinno opierać się na radykalnym leczeniu chirurgicznym. Badania obrazowe mają znaczenie w kontroli pooperacyjnej, jednak w niektórych przypadkach ich specyficzność jest ograniczona.

SŁOWA KLUCZOWE: ceruminous adenocarcinoma, przewód słuchowy wewnętrzny

INTRODUCTION

External auditory canal neoplasms account for <1% of all malignant neoplasms located in the head and neck [1, 2]. The incidence rate is 1/1 000 000/year [3]. It most often affects adults between 50 and 60 years of age [4, 5]. Among the cancers present in the discussed location, those derived from the ceruminous glands deserve special attention. Ceruminous glands together with sebaceous glands are located in the dermis of the cartilaginous portion of the external auditory canal and are responsible for the production of ear-protecting ear wax [6, 7]. Cancers from this area are extremely rare. It is worth mentioning that they constitute only 2.4% of all external auditory canal neoplasms [8]. Due to a wide variety, both in clinical and histological terms, tumors often constitute a serious diagnostic problem, which results in late diagnosis. They are very aggressive; therefore, their treatment should be based upon surgical removal of the lesion with a healthy tissue margin, possible excision of neck lymph nodes and on complementary treatment (radio- and chemotherapy) [9, 10, 11].

CASE REPORT

A 58-year-old woman was admitted to the Clinic with increasing pain of the left ear and pus and blood purulent leakage from the ear. The patient also reported profound hearing loss on the left side which she had experienced for many years. For several months, the patient gradually developed a feeling of ear occlusion, tinnitus. Due to the aforementioned complaints, the patient was admitted to a district hospital, where treatment typical of inflammation of external auditory canal was implemented. In the absence of improvement, the woman was referred to the PUM Otolaryngology Clinic.

On otoscopic examination performed in auditory canal lumen, a hard tumor was revealed on the left side; it was covered with inflamed skin and a blood-purulent exudate.

In tonal audiometry: in the right ear (RE) - average hearing at the level of 15 dB, left ear (LE) - average hearing at the level of 90 dB.

After being admitted, swab was collected from the left ear - negative culture for bacteria and fungi.

Histopathological examinations of tumor specimens led to the suspicion of mucoepidermoid carcinoma and it was recommended to remove the lesion for histopathological verification.

Computed tomography (CT) of temporal bones was perfor-

med, which revealed a lesion in the form of shading to the auditory canal and mastoid process, without accompanying bone destruction (Fig. 1.). Diagnostics was extended with magnetic resonance imaging (MR). Lesion limited to the cartilaginous portion of the auditory canal, in the mastoid process there were inflammatory lesions.

The patient was operated on; a tumor of the cartilaginous portion of the canal was found blocking its lumen during surgery. The lesion was removed entirely, from double access (intra-auricular and post-auricular) with a margin of healthy tissues. No evidence of bone infiltration was found. Uncomplicated postoperative course.

Post-operative histopathological examination revealed ceruminous adenocarcinoma (tumor removed in the limits of normal skin tissue). The patient was referred for supplementary radiotherapy. On irradiation, there was ear discharge were, which resolved after conservative treatment. The patient remained under supervision of the Outpatient Clinic. After 8 months from surgery, otoscopic examination showed that the external auditory helix was properly healed without recurrence.

The end of radiotherapy was followed by positron emission tomography, which revealed no pathological enhancements.

An outpatient MR performed a year within surgery revealed an area of 17 × 23 × 18 mm located at the base of the left temporal lobe and in the mastoid process of the pyramid of the temporal bone, demonstrating pathological enhancement within the external auditory canal and the skin of the entrance to the canal, raising the suspicion of a recurrent neoplasm. Diagnosis was supplemented with CT (Fig. 2.) of the temporal bone, which revealed small bone defects corresponding to the lesion described in MR on the posterior and upper bone wall of the auditory canal. These lesions were not visible in the CT scan performed prior to the first treatment. The whole clinical image suggested suspicion of cancer recurrence. The patient was qualified for surgery once again.

Physical examination showed a healed scar in the left ear. In otoscopy - external auditory canal slightly narrowed, skin unchanged, matt tympanic membrane, without pathological changes. In tonal audiometry - deafness of the left ear.

The patient was re-operated. Lateral petrosectomy was performed; lesions that could correspond to suspected foci from imaging examinations were marked with ink. Postoperative histopathological examination showed no evidence of neoplastic infiltration. Currently, after 6 months of treatment, the patient remains under the care of the Outpatient Clinic.

DISCUSSION

In the past, all tumors derived from ceruminous glands, whether benign or malignant, were referred to collectively as “ceruminoma”. It was not until 1972 that Wetli et al. developed a detailed classification based on histological differences, in which they distinguished 4 categories: ceruminous adenomas, ceruminous adenocarcinomas, adenoid cystic carcinomas and pleomorphic adenomas [12]. In 1977 Pulec added mucoepidermoid carcinoma to the aforementioned classification [1].

Ceruminous adenocarcinoma is a rare malignant tumor that develops from the cervical glands present in the external auditory canal [13]. It does not give characteristic symptoms, its most frequent ailments are: feelings of ear occlusion, hearing loss, otalgia and tinnitus [8, 14]. The variety of symptoms reported by patients is confirmed in the literature. Bilici et al. and Kim et al. analyze cases of patients which only experienced hearing loss, while Shim presents a case of a patient who, apart from hearing loss, complained of pain and pathological secretion from the ear, dizziness and sensory loss on half of face [10, 13, 15].

The described patient reported due to severe ear pain and blood-purulent secretion. In addition, she also reported hearing loss with tinnitus.

In the case in question, otoscopic examination in the lumen of the external auditory canal revealed a hard, elastic tumor covered with red skin and a blood-purulent secretion. According to Shim, conducted physical examination showed exophytic tumor that completely fills the anterior part of the canal, and in the patients described by Bilici et al. and Kim et al., otoscopic examination showed polypoid masses emerging from the walls of the canal and covering the tympanic membrane. [10, 13, 15].

Imaging examinations: computed tomography and magnetic resonance imaging, prove extremely useful in accurate assessment of the extent of lesions. [6, 10]. The hospitalized patient had CT and MR performed in the Clinic, on the basis of which it was found that the fundamental lesion is limited to the cartilaginous portion of the canal. It is worth noting that CT and MR are complementary studies, and MR allowed to precisely determine tumor size and differentiate lesions in the mastoid process between the infiltrate and inflammation.

Ceruminous adenocarcinoma rarely metastasizes to cervical lymph nodes. There have been reports of patients with brain, lung and bone metastases in the literature [14, 16].

The treatment of choice is surgical removal of the lesion, follo-

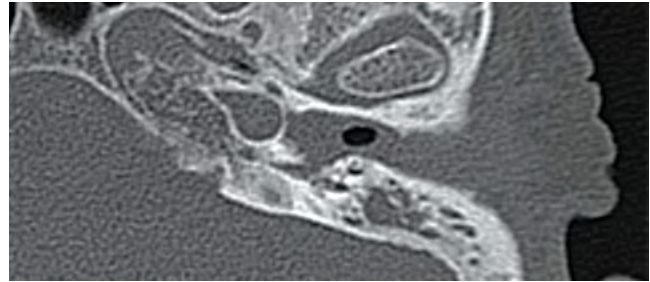


Fig. 1. CT examination - “opacity lesion” of the left external auditory canal.

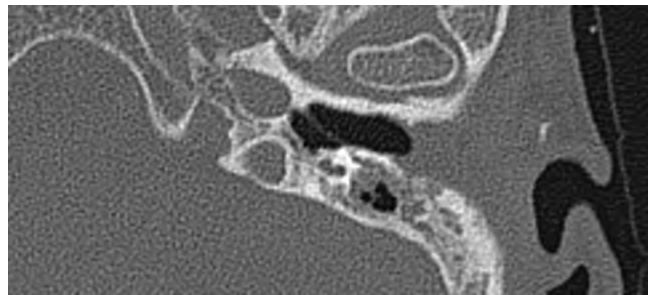


Fig. 2. CT scan - minor bone defects on the posterior and upper bone wall of the external left ear canal.

wed by radiotherapy [14, 17]. The essence of the treatment is excision of the lesion with a margin of healthy tissues [15]. Hicks is the only to believe that parotidectomy and cervical lymphadenectomy should be performed during surgery as well. [14].

The patient admitted to the Clinic was operated on. The tumor was excised together with surrounding fragments of the external auditory canal and reconstruction of the canal was performed. Bone infiltration was not found during the procedure. Post-operative histopathological examination revealed ceruminous adenocarcinoma (it was also confirmed that the tumor was removed in the limits of normal skin tissue). After receiving the result with the final diagnosis, the patient was referred for supplementary radiotherapy.

According to Hicks, the most recurrences occur within a few months after the procedure, however, there are patients who have experienced recurrences even after 7 years of surgery [14]. No cancer recurrence was found within 8 months after the procedure in the described case.

Signs suggestive of recurrence of the neoplastic process were found in the outpatient clinic after about a year of MR examination. This was also suggested by CT examination of the temporal lobe performed during the patient’s stay at the Clinic. It is worth noting that the only complaints reported by the patient were headaches, and no significant deviations from the norm

were found in otoscopy. The patient was re-operated (left-sided lateral petrosectomy was performed). Qualification for petrosectomy was facilitated by the fact that the patient had severe hearing loss on this side. Postoperative histopathological examination excluded recurrence. Lesions found in imaging studies were associated with inflammation of the auditory canal and chronic leakage that occurred in the course of radiotherapy, which the patient underwent after the first surgery.

Only two cases of recurrent adenocarcinoma tumors have been found in the literature. Both patients had a history of symptoms suggestive of pathology (ear pain with leakage of purulent content and hearing impairment), which was confirmed in otoscopic examination (in the first case a tumor was found, in the second polypoid mass filling the canal lumen) and performed imaging examinations. Both patients were operated

on and in both cases a positive result was obtained in postoperative examinations [18, 19].

CONCLUSIONS

Tumors of the ceruminous glands (such as ceruminous adenocarcinomas) are extremely rare. They do not produce any characteristic symptoms; therefore, they are often diagnosed late. In surgical treatment, it is important to remove the lesion with a margin of healthy tissue. If confirmed in post-operative study, supplementary treatment (radiotherapy) is required. Due to the possibility of recurrence (even after 7 years of surgery), patients should remain under constant ENT care. Imaging studies are important in post-operative follow-up, but in some cases their specificity is limited.

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