

Squamous Cell Carcinoma of the Middle Ear: Case Report and Literature Review

Mariam Harrak*, Khouloud Ziani, Madiha Chelakhi, Othmane Saqri, Nabila Sellal,
Mohamed El Hfid

Department of Radiotherapy, University Hospital Mohamed VI of Tangier, Tangier, Morocco
Email: *mariame.harrak@gmail.com

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Abstract

Malignant tumors originating from the middle ear are rare. The literature identifies chronic inflammation and Human Papillomavirus (HPV) infection as the most common risk factors. A CT scan to assess bony invasion and an MRI to evaluate soft tissue involvement, depth of invasion, and perineural invasion, followed by a biopsy, are indispensable for diagnosis and treatment. There is no standard treatment for squamous cell carcinoma of the middle ear, however, most reported cases are treated with surgical resection followed by postoperative radiotherapy. Given the challenges of achieving complete surgical excision, radiotherapy plays a crucial role in controlling middle ear cancers, as demonstrated in our case. We present a case of squamous cell carcinoma of the middle ear in a 63-year-old female with a history of chronic suppurative otitis media. The patient underwent a right subtotal petrosectomy without lymph node dissection followed by concurrent chemoradiotherapy. At the one-year follow visit, no recurrence or metastasis was detected.

Keywords

Chronic Suppurative Otitis, The Middle Ear, Squamous Cell Carcinoma, Surgery, Radiotherapy

1. Introduction

Malignant tumors originating from the middle ear are rare. The most commonly reported risk factors in the literature are chronic inflammation and Human Papillomavirus infection. A CT scan to assess bony invasion and an MRI to evaluate soft tissue involvement, depth of invasion, and perineural invasion, followed by a biopsy, are indispensable for diagnosis and treatment. There is no standard treatment for squamous cell carcinoma (SCC) of middle ear. In most reported cases,

the chosen treatment involves surgical resection followed by postoperative radiotherapy.

2. Case Report

A 63-year-old female patient was admitted to our hospital presenting with right ear pain persisting for the past 6 months, accompanied by facial paralysis for one month. Additionally, she had been experiencing purulent discharge and intermittent bleeding from the right ear for 2 years.

Upon initial otological examination, we observed purulent secretion from the right external auditory canal, along with granulomatous tissue in the tympanic cavity, which exhibited easy bleeding upon manipulation. Examination of the mastoid area revealed swelling, tenderness, and signs of inflammation. Further evaluation confirmed complete peripheral facial paralysis on the right side (**Figure 1**).

The appearance on the temporal bone CT scan favored a diagnosis of chronic right cholesteatomatous otitis media, without cervical lymph node involvement.

Microscopic examination showed the presence of large tumor cells with abundant eosinophilic cytoplasm and atypical, mitotic nuclei within the cytoplasm. These cells were observed surrounding keratin lamellae. The conclusive pathological diagnosis indicated a well-differentiated, keratinizing, and invasive squamous cell carcinoma of the right middle ear.

The patient underwent a right subtotal petrosectomy without lymph node dissection. The surgical intervention was not radical due to extension towards the carotid artery, the facial nerve and the internal jugular vein. No secondary localizations were detected in the cancer staging.

An MRI performed postoperatively showed a lesion in the right petrosectomy cavity measuring 4.1 cm by 3.8 cm, with infiltration into the right parotid gland, the right parapharyngeal fat, and partial invasion of the pterygoid muscle. Additionally, the process involves the petrous portion of the right internal carotid artery (**Figure 2**).

A thoraco-abdomino-pelvic CT scan was requested as part of a staging workup and did not reveal any secondary locations.

Postoperative radiotherapy with intensity-modulated radiation therapy (IMRT) technique was administered to the patient. She received in sequential a dose of 70 Gray in 35 fractions to the high risk target volume and 63 Gray in intermediate risk target volume. She received prophylactic neck irradiation 56 Gray in 35 fractions to the Levels II, III, IVa and V.

The head and neck were immobilized with a thermoplastic mask. In addition, a bolus was used and placed in contact with the external ear orifice of the mastoid region to be able to cover the externalized mass with the adequate dose.

During the course of radiotherapy, grade I radiodermatitis was noted, as well as grade I xerostomia with nearly complete disappearance of the intra and retroauricular mass.

The patient underwent 7 cycles of concomitant cisplatin with a dose of 40 mg/m² per week, which was well tolerated clinically.

No recurrence or metastasis was observed at the one year follow visit (**Figure 3**).

The patient reports a disappearance of pain and inflammatory signs, and an improvement in sleep quality, but she reports accentuation of the facial hemiparesis (**Figure 4**).



Figure 1. Clinical presentation before to the initiation of radiotherapy.

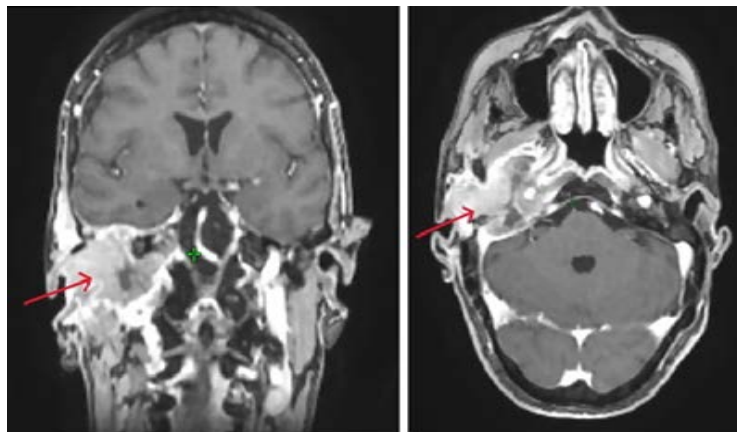


Figure 2. Postoperative MRI revealed a residual lesion in the right petrosectomy.

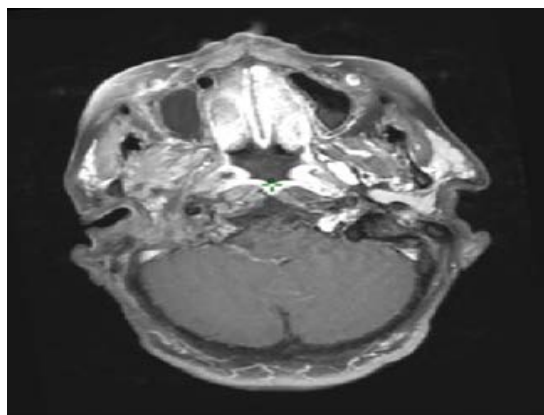


Figure 3. MRI performed 3 months after radiotherapy showing significant response.



Figure 4. Clinical presentation after to the initiation of radiotherapy.

3. Discussion

Malignant tumors originating from the middle ear are rare, accounting for only 0.25% of malignant tumors occurring in this region. The most common malignancy of middle ear is conventional keratinising squamous cell carcinoma. Non keratinising squamous cell carcinoma primary in middle ear is extremely rare [1]-[3].

The etiological factors of primary middle ear carcinomas are unknown, but a possible cause is metaplasia of the middle ear mucosa caused by chronic inflammation [2] [4]. Between 75 to 85% of primary middle ear carcinomas are secondary to chronic suppurative otitis media [1] [5]. In our case, the patient had a history of chronic suppurative otitis media for over 2 years.

The other risk factors include sporadic reports of primary ear malignancies developing in areas treated with radiation for other head and neck tumors and HPV infection which has been found in a significant number of cases [6] [7].

In the majority of series, otorrhea typically presents as the initial symptom, while pain, facial nerve palsy, and the presence of a mass in the external auditory canal occur later as the tumor progresses.

Diagnosis is often delayed as these symptoms resemble more common otologic conditions.

Literature indicates that stage III or IV disease is marked by the presence of a mass obstructing the external auditory canal [8]-[10]. Our case had a similar presentation.

While there is no universally accepted staging system for squamous cell carcinoma of the ear and temporal bone, the modified University of Pittsburgh staging system is commonly used. Generally, tumors limited to the external auditory canal (T1 and T2) have a better prognosis compared to those involving the middle ear, mastoid, or facial nerve (T3 and T4) [1] [9] [11].

CT scan are essential for assessing bony invasion, while MRI is crucial for evaluating soft tissue involvement, depth of invasion, and perineural invasion. These imaging techniques should be performed prior to proceeding with a biopsy to obtain a definitive diagnosis. Regional or full-body imaging is often performed to evaluate for metastases [12].

Due to its rarity and aggressive oncological behaviour, there is no established standard treatment for squamous cell carcinoma of the external auditory canal and middle ear. In the majority of reported cases, the chosen treatment involved surgical resection followed by postoperative radiotherapy [3] [13].

Radiotherapy is administered often in combination with chemotherapy when surgery is not feasible [14] [15]. The definitive Chemoradiotherapy may be also an effective alternative to surgical resection [16].

A multi-institutional review of 87 records, which focused primarily on the roles of surgery and radiotherapy (RT) in patients with confirmed squamous cell carcinoma of the external auditory canal and middle ear who were treated between 1984 and 2005, found that the 5-year disease-free survival rates for patients with T1, T2, and T3 stages were 83%, 45%, and 0% in the RT group, and 75%, 75%, and 46% in the surgery plus RT group, respectively. The study concluded that RT is the treatment of choice for stage T1, while surgery combined with RT is recommended for stage T2 or T3 cancer [17].

Surgery is the primary treatment approach, subtotal temporal bone resection, or total temporal bone dissection depending on the extent of the disease. Complete excision with adequate margins is preferred. In general, tumors localized within the external auditory canal (T1) are typically managed with limited resection procedures such as local excision or radical mastoidectomy. Conversely, more advanced lesions (T2 - T4) usually require more extensive treatment approaches such as en bloc resection, partial temporal bone resection, subtotal petrosectomy, or total petrosectomy [10].

Radiotherapy is often recommended as an adjunct to surgery or for palliative purposes rather than as a primary curative treatment. Adjuvant radiation is recommended for positive margins, perineural invasion, bone invasion, and lymph node involvement.

This irradiation carries the risk of osteoradionecrosis of the petrous bone with bone exposure and persistent suppuration. Postoperative irradiation with intensity modulation aims to limit the sequelae [18].

The complex anatomy of the middle ear and temporal bone includes crucial structures such as, temporal lobe, facial nerve, internal carotid artery, and internal jugular vein, all closely positioned. The middle ear is situated within the petrous part of the temporal bone and contains the auditory ossicles. Consequently, addressing tumors in this area often requires extensive surgical interventions that could impact functionality and lead to esthetic complications and render surgeries with oncologic limits unfeasible [19]-[21].

In literature, the five-year survival rate for patients who undergo surgery and radiotherapy is about 25% - 55%. Patients with petrous bone invasion and positive margins like our case, have a poorer prognosis [22]-[24].

4. Conclusion

Tumors affecting the middle ear are infrequent, leading to limited resources in the

literature. Large series are rare, and there are no controlled clinical trials, resulting in non-standardized approaches to staging and management. The difficulty of achieving a complete surgical excision makes the role of radiotherapy important in controlling middle ear cancers, as in our case.

Authors' Contributions

All authors have been involved in the process of writing and have approved the final manuscript.

Conflicts of Interest

The authors declare that they have no competing interests.

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