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Ceruminous Adenocarcinoma of External Auditory Canal: A Very Rare Case of Malignant Tumor of the Temporal Bone

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I. Introduction

Ceruminous glands are specialized apocrine glands located in the cartilaginous part of the external auditory canal (EAC). Primary malignant tumors arising from these glands are infrequent, and they may present a diagnostic dilemma because of their varied clinical and histological manifestations. Here we report a patient with an extended ceruminous adenocarcinoma. This article highlights the importance of early diagnosis for optimal management of this pathology.

II. Case Report

A 37 years old man, without a history of diabetes mellitus or immunesupression, presented to our head and neck department with one (01) year history of left purulent otorrhea with the decreased hearing of the left ear and facial paralysis without vertigo. He denied experiencing dizziness, tinnitus, vertigo or earache.

On physical examination, there was a retro-auricular fistula with local inflammatory signs. Otoscopy showed a large polyp on the cartilaginous part of the left EAC, with mucopurulent discharge in the EAC. The tympanic membrane was not visible. Audiometry revealed conductive hearing loss on the left. The 4-frequency pure tone average was 50 dB for air conduction and 10 dB for bone conduction. We didn’t note any palpable cervical lymph nodes.

A Computed Tomography scan of the temporal bone showed the presence of a mass in the left EAC and middle ear with lytic lesions of the facial canal nerve, carotid canal and EAC (figure 1).

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Figure 1: CT scan showing the collection in the left EAC, with osteolysis and osteitis of the temporal bone (Arrows).

Using retro-auricular approach, the mass of the EAC was removed (figure3), and histological analysis showed a poorly differentiated carcinomatous proliferation primarily suggestive of a neuroendocrine tumor. We completed with immunohistochemistry that showed an intense and diffuse positivity of cytokeratin 7 and CD117. The final diagnosis was ceruminous adenocarcinomas.
Because of the extension of the mass, the decision was to propose for intensity-modulated radiotherapy; he received a total of 60 Gy.

III. Discussion

Ceruminous adenocarcinomas are a rare malignant tumor that arises from the ceruminous gland found in the cartilaginous part of EAC. It is the 2nd most common type of carcinoma arising from ceruminous-glands [1]. Most affected patients are in their fifth or sixth decades of life and average age at diagnosis of 48 years. Its incidence is equal between males and females [1, 2]. There are no known risk factors for the development of these tumors, although the otitis externawas implicated in animal models [2]. Ceruminous adenocarcinoma exhibits a prolonged subclinical phase, often lasting years, before presentation [3].
The most common symptoms reported in the literature are: mass of the EAC, hearing changes, otorrhea, bleeding, headaches, equilibrium changes, pain and facial paralysis, and less frequently lymphadenopathy or tinnitus. Although cervical lymph node metastasis is uncommon, distant metastasis to the bones, lungs, and brain have been described [4].

Imaging plays important role in predicting and delineating the EAC tumor extent. Axial and coronal computed tomographic scans help in detecting bony erosion and disease extension as well as in excluding a primary parotid or middle ear tumor [3].

Histologically, ceruminous adenocarcinoma is classified as high-grade and low-grade tumors based on the extent of glandular differentiation and the proportion of solid areas [6, 8]. Low-grade tumors may mimic the histopathologic features of the benign ceruminous adenomas but are distinguished from the latter by the presence of stromal invasion and a desmoplastic stromal response. High-grade adenocarcinomas extensively infiltrate the surrounding tissues as irregular glands, cords, and sheets of overtly malignant cells. You marked nuclear pleomorphism and abundant mitotic figures. High-grade tumors may exhibit minimal or no evidence of apocrine derivation and needs to be distinguished from metastatic adenocarcinoma originating from other sites [6].

The mainstay of management of this tumor is combined surgery and radiotherapy. Radiotherapy has a limited role in treatment as a conjunctive therapy with surgery and in cases of distant metastasis. As a surgical procedure, Hicks has proposed initial en bloc surgical resection. And then, if tumor have an extension to the middle ear, resection of the temporal bone and adjacent structures is necessary [4, 7]. In our case, there is a tumor extension to the carotid canal, complete resection was impossible. Surgical margin positivity at initial surgery, bone extension, perineural invasion, and local recurrence seem to be prognostic factors for cases in the literature. Chemotherapy did not prevent the development of distant metastasis. Prognosis may be difficult to predict, 25% of patients with ceruminous adenocarcinoma died of disease a mean of 2 years after presentation. The disease-specific mortality is 35% [8].

IV. Conclusion

Ceruminous adenocarcinoma is a rare malignant subtype of ceruminous gland neoplasm; its diagnosis is most often challenging, comprehensive evaluation including physical examination, multimodal imaging studies and thorough histologic analysis of an adequate biopsy that includes surrounding tissue is necessary to arrive at the correct diagnosis. Prognosis may be difficult to predict but complete surgical excision coupled with radiation yields the best long-term prognosis.

References Références Referencias