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Metastatic Ceruminous Adenoid Cystic Carcinoma of the Lumbar Spine Causing Neurological Compromise: A Case Report

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Abstract

Background: Ceruminous glands are modified apocrine glands of the external auditory canal (EAC). Malignant tumours within the ceruminous glands are extremely rare, and the most common histological type is adenoid cystic carcinoma (ADCC), which has high recurrence and metastasis risks. Although a few cases of metastatic ADCC from other head and neck glands have been reported, metastatic ADCC originating from the ceruminous gland are extremely rare. **Case presentation:** We present an unusual case of spinal metastases of ADCC from ceruminous glands. A 61-year-old woman complaining of low back pain and both lower limbs pain was referred to our department. The primary ceruminous tumour was resected 26 years ago and recurred 6 years later, which was treated by radiotherapy. Three years ago, she presented with low back pain and was diagnosed as multiple lungs and bone metastases. The patient underwent tumour excision, decompression and fusion. The biopsy revealed metastatic ADCC. The symptoms were alleviated after surgery. **Conclusions:** ADCC of EAC is a pernicious malignant tumour that is characterized by slow-growing patterns and a high predisposition to recurrence and metastasis. Differential diagnoses of ADCC and benign tumours in the EAC are challenging, particularly at early stages. We report a rare case of ceruminous ADCC with a prolonged clinical history as well as spinal metastasis and highlight the significance of regular follow-ups for patients undergoing tumour excision in the EAC.

Keywords

ceruminous gland malignancy, adenoid cystic carcinoma, compressive syndrome, spinal metastasis, case report

Background

Incidences of malignant tumours of the external auditory canal (EAC) are extremely low, affecting about 1-6 cases per million people, annually. Among them, squamous cell carcinoma (SCC) is the most common histological subtype.[1] Ceruminous glands are modified apocrine glands of EAC. Neoplasms that originate from ceruminous glands comprise a minority of EAC malignancies. Among them, adenoid cystic carcinoma (ADCC) is the most prevalent malignant ceruminous neoplasm, accounting for about 5% of primary malignancies of EAC,[2-4] followed by ceruminous adenocarcinoma not otherwise specified and ceruminous mucoepidermoid carcinoma.[5]

Compared to their benign counterparts, ceruminous carcinomas usually occur at earlier ages, with slight

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predilections for females. They are characterized by otalgia, a mass in the EAC, hearing changes as well as otorrhea, and are difficult to be distinguished from benign neoplasms.[6] Due to the invasive nature of disease and procrastination of treatment, ceruminous carcinomas are associated with high incidences of lung, brain, and bone metastases.[3,4] ADCC has been proved to be a highly malignant but slow-growing tumour through clinical follow-up with a marked propensity for invading the perineural, lymphatic and nerve sheaths.[4] Among the 3 types of ceruminous carcinoma, ADCC metastases are relatively frequent that happened in about 27% patients, with lungs being the most frequent sites, followed by the brain and bone.[2,6,7] We report a woman presenting with low back pain and lower extremities symptoms diagnosed with spinal metastasis of ceruminous ADCC. She had a long, complicated clinical course after initial resection of tumour in EAC 26 years ago.

Case Presentation

A 61-year-old female patient complaining of low back pain and lower extremities pain with paraesthesia was referred to our institution. She had a paroxysmal sharp low back pain, with pain and numbness in both lower extremities. Physical examination revealed tenderness and percussion pain of the L1-L3 spinous process, accompanied by deep percussion pain. Bilateral superficial sensation was reduced in the cutaneous area corresponding to nerves below L1. Perineal sensation was abnormal. Lumbar mobility was limited. Power in knee extension and ankle dorsiflexion were 4/5 bilaterally while tone was decreased in lower limbs. Bilateral knee and Achilles tendon reflexes were reduced while both ankle clonus test and Babinski sign were negative and straight leg raising was normal.

Magnetic resonance imaging (MRI) outside our institution suggested multiple abnormal signals of the cervical vertebrae, thoracic vertebrae, lumbar vertebrae, pelvic bone, as well as bilateral femur, and enhanced mass at L2 spinal canal level whose distinction with adjacent bone marrow and meninges was unclear. Subsequent abdominal contrast-enhanced computed tomography (CT) scans showed nodular changes in bilateral adrenal glands and multiple irregular hyperdense shadows in the thoracic vertebrae, lumbar vertebrae, iliac bone, and femur (Figure 1).

The patient had a prolonged and complicated clinical history. Twenty-six years ago, she underwent cerumen adenoma resection in the right EAC. Biopsy indicated low-grade malignancy, and more details were not available. Six years later, the tumour recurred and radiotherapy was performed in other institution. Three years ago, the patient developed low back pain without pain and numbness in the lower limbs. MRI showed multiple metastases of the lumbar spine. Further examination revealed multiple bone metastases throughout the body while biopsy of sacral and iliac bones revealed metastatic adenocarcinoma. She was subjected to chemoradiation in this institution and symptoms were relieved. One year ago, she suffered from an exacerbation of low back pain, radiating to the left lower extremity with numbness. Radiotherapy was performed in combination with Zoledronic acid administration to prevent bone adverse events and oral analgesic, however, the efficacy was poor. Three months ago, the patient was admitted into the oncology department of our institution for further treatment. Chest CT scans revealed multiple nodules in both lungs. Given osteogenic changes of bone metastases, bilateral internal iliac arteries chemoembolization under guidance of Digital Subtraction Angiography (DSA) was performed. The symptoms in both lower extremities of the patient were significantly alleviated after operation.

The patient underwent posterior L2 vertebrae tumour resection and biopsy with spinal canal decompression, autogenous iliac bone graft fusion and internal fixation (Figure 2). During the procedure, we found intradural and extradural tumours at the L2 vertebral plane. After surgery, there was an immediate improvement in symptoms in both lumbar and lower extremities. Haematoxylin eosin staining of the sample indicated a histological type of adenoid cystic carcinoma (Figure 3A) while immunostaining revealed P63 (+), S-100 (−), CD117 (−), SMMHA (+), Calponin (+), EMA (+), and CK7(+) (Figure 3B). Based on these findings and her clinical history, metastatic adenoid cystic carcinoma of ceruminous gland was diagnosed. Then, the patient was discharged and followed up as an outpatient. She died 2 years after operation because of pulmonary metastases.

Discussion

Ceruminous carcinoma of the EAC is extremely rare.[8] According to the latest World Health Organization Classification of head and neck tumours, ceruminous carcinomas are classified as ceruminous adenocarcinoma, adenoid cystic carcinoma, and mucoepidermoid carcinoma.[9] ADCC, the most common ceruminous malignant tumour, has a relatively favourable outcome compared to the other 2 types.[5,6] Similar to general ADCC, ADCC of EAC is characterized by slow-growing but aggressive patterns with a high propensity to recur and metastasize.[2,8] Pulmonary, intracranial and bone metastases have been previously reported.[3,10,11] A limited number of spine metastasis cases of ADCC from other gland origins of head and neck, such as parotid gland, submandibular gland, tongue base, and lacrimal gland have been reported.[12-14] However, spinal metastatic ADCC arising from

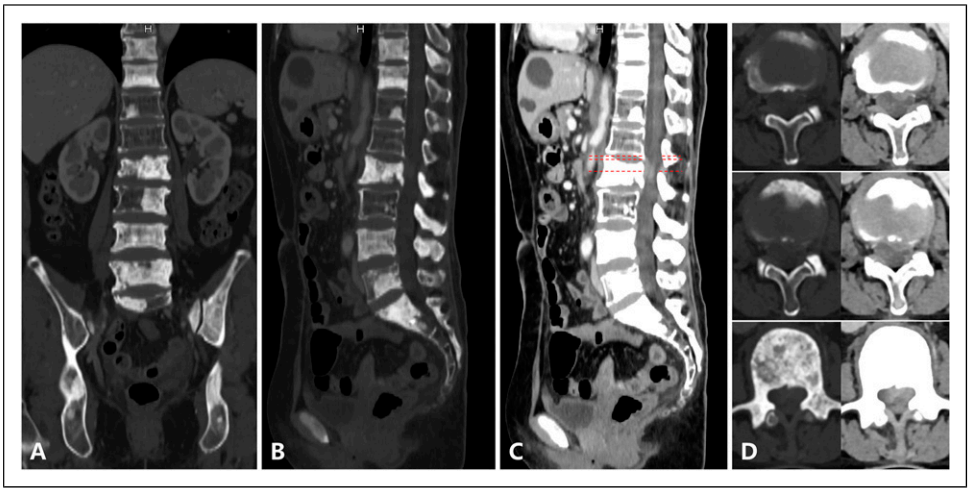

Figure 1. Preoperative coronal reconstructed (A) and sagittal bone window CT (B) show multiple irregular hyperdense lesions in the vertebrae of thoracic, lumbar and sacrococcyx, iliac bone. The sagittal soft-tissue window CT shows the soft tissue density mass in the spinal canal at L2 level (C) and the corresponding axial CT suggests Grade 2-3 epidural spinal cord compression (ESCC) (D).

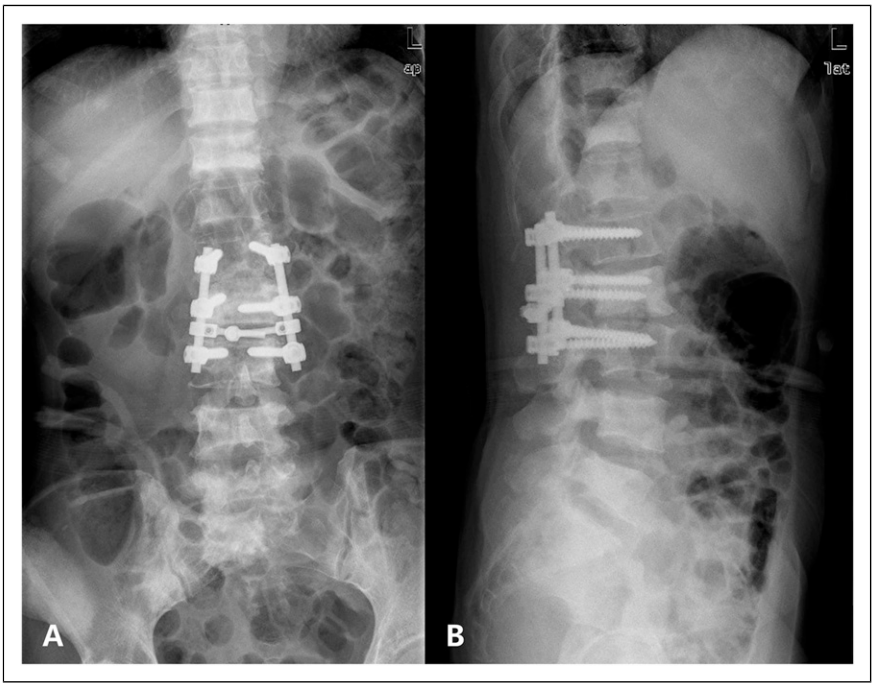

Figure 2. Postoperative anteroposterior radiograph following instrumented stabilization (A). Postoperative lateral radiograph following instrumented stabilization (B).

ceruminous gland in the EAC is exceedingly rare. Marchesini et al.[15] reported a male patient complaining of back pain and worsening myelopathy and was diagnosed with intramedullary spinal cord metastasis from ADCC of EAC. In the present case, the patient was diagnosed with metastatic ADCC of ceruminous gland in EAC and mainly presented with neurological compression symptoms. In addition, this patient experienced recurrence and multiple distant metastases with a prolonged and complicated course of nearly 3 decades, emphasizing the necessity of regular follow-ups for a long period of time as well as the significance of informing the patient of possible relapse after resection of the primary ADCC in the EAC.

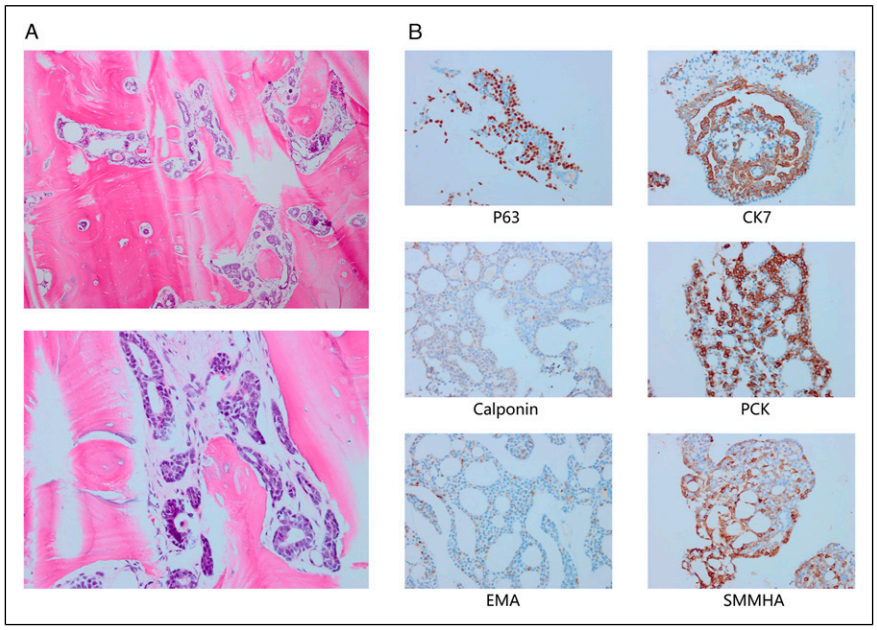

Figure 3. Hematoxylin eosin (HE) staining resulted in a diagnosis of the tumor as adenoid cystic carcinoma (A). P63 (+), CK7(+), Calponin (+), PCK(+), EMA (+), and SMMHA (+) were suggested (B).

Ceruminous malignancies have less specificity, manifesting with otorrhea, otalgia, and hearing loss, leading to neglect from both patients and attending doctor at the early stage.[4,16] Similarities in histomorphology magnifies the difficulty of accurate diagnosis.[17] Our patient was diagnosed with ceruminous adenoma at the first place with biopsy revealing low-grade malignancy. However, histopathological results after lumbar surgery indicated ADCC. This result suggests a confusion of ADCC and benign tumour of EAC at initial diagnosis, which may lead to inappropriate treatment and lack of strict follow-up. Although low-grade cancers such as adenoid cystic carcinoma have similar features with benign lesions, otalgia may occur in the early course of most patients and last for several years before established diagnosis.[3,4,6]

Despite its pernicious and malignant nature, ceruminous ADCC are slow-growing and may be associated with a prolonged clinical course.[4] Most patients have symptoms for several years before established diagnosis. Delayed treatment at the early stage results in local invasion or distant spread of tumours. Even after adequate treatment, recurrence is common for these malignancies. Recurrences are more likely to happen when the tumour is histologically identified on or close to excision lines.[3] Patients with a longer duration of symptoms before diagnosis also have high recurrence risks.[2] A long-term study involving 27 patients with EAC carcinoma found that recurrences were prone to happen in patients aged < 60 years and with lymphovascular invasion.[18] Perzin et al.[3] reported 16 patients with ADCC involving the EAC. Nine patients had a total of 26 local recurrences, most of which occurred within 2 years of initial resection. Two of them were living with unresectable tumours while 5 ended up with fatal outcomes caused by intracranial extension or pulmonary metastases of diseases. Given slow growth patterns of most ADCC, a patient may live for many years, even with systemic metastases.[4,12] Since first resection of mass in EAC 26 years ago, our patient experienced a long clinical course. Dong et al.[2] identified 22 patients with ADCC of EAC. In this series, 9 patients developed recurrences with a median follow-up time of 8 years, only one of them had an isolated regional recurrence, 6 died of their disease at a mean of 10 years after first intervention, while 3 were living with distant metastases at their final follow-ups.[2] Bonaparte et al.[8] identified 66 cases previously reported in a quantitative assessment of ADCC within EAC. Distant metastases were confirmed in 38% of the patients, with lungs being the most common sites. Of them, 3 patients were diagnosed with multiple metastases. Given the high recurrence rates and metastasis risks, long-term follow-up of ADCC patients is necessary. Due to scarce cases and high heterogeneity, evidence-based treatment recommendations are

lacking. While the efficacy of adjuvant radiation therapy is controversial, this conservative therapy can be used for symptomatic palliation.[3]

Conclusion

Adenoid cystic carcinoma arising from the ceruminous gland is difficult to differentiate from benign neoplasms in the external ear canal at early stage and delayed diagnosis is common, which lead to high risks of recurrence and systemic metastasis. We report a case of ceruminous adenoid cystic carcinoma with a prolonged clinical history and multiple metastasis. This case highlights the significance of regular follow-ups for patients undergoing excision of tumours in external auditory canal, and comprehensive examination is required in patients with musculoskeletal symptoms and a history of a mass in external auditory canal.

Declaration of Conflicting Interests

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References

1. Lobo D, Llorente JL, Suárez C. Squamous cell carcinoma of the external auditory canal. *Skull Base*. 2008;18(3):167-172. doi:10.1055/s-2007-994290.
2. Dong F, Gidley PW, Ho T, Luna MA, Ginsberg LE, Sturgis EM. Adenoid cystic carcinoma of the external auditory canal. *Laryngoscope*. 2008;118(9):1591-1596. doi:10.1097/MLG.0b013e31817c42a8.
3. Perzin KH, Gullane P, Conley J. Adenoid cystic carcinoma involving the external auditory canal. A clinicopathologic study of 16 cases. *Cancer*. 1982;50(12):2873-2883. doi:10.1002/1097-0142(19821215)50:12<2873::aid-cncr2820501230>3.0.co;2-r.
4. Hicks GW. Tumors arising from the glandular structures of the external auditory canal. *Laryngoscope*. 1983;93(3):326-340. doi:10.1288/00005537-198303000-00016.
5. Crain N, Nelson BL, Barnes EL, Thompson LDR. Ceruminous gland carcinomas: a clinicopathologic and immunophenotypic study of 17 cases. *Head Neck Pathol*. 2009;3(1):1-17. doi:10.1007/s12105-008-0095-9.
6. Nagarajan P. Ceruminous neoplasms of the ear. *Head Neck Pathol*. 2018;12(3):350-361. doi:10.1007/s12105-018-0909-3.
7. Arato J, Michel RP, BaxterCeruminoma JD. Ceruminoma. *J Otolaryngol*. 1980;9(6):482-486.
8. Bonaparte JP, Trites J, Hart R, Taylor SM. Survival when treating adenoid cystic carcinoma of the external auditory canal: quantitative assessment of case reports. *J Otolaryngol Head Neck Surg*. 2009;38(4):468-476. doi:10.2310/7070.2009.080195.
9. Thompson LDR. Update from the 4th edition of the world health organization classification of head and neck tumours: tumours of the ear. *Head Neck Pathol*. 2017;11(1):78-87. doi:10.1007/s12105-017-0790-5.
10. Koopot R, Reyes C, Pifarré R. Multiple pulmonary metastases from adenoid cystic carcinoma of ceruminous glands of external auditory canal. A case report and review of the literature. *J Thorac Cardiovasc Surg*. 1973;65(6):909-913.
11. Conlin PA, Mira JL, Graham SC, Kaye KS, Cordero J. Ceruminous gland adenoid cystic carcinoma with contralateral metastasis to the brain. *Arch Pathol Lab Med*. 2002;126(1):87-89. doi:10.1043/0003-9985(2002)126<0087:Cgaccw>2.0.Co;2.
12. Riche M, Mathon B, Mokhtari K, Carpentier A, Amelot A. Cylindroma spine metastasis: long-term follow-up is required. *Neurochirurgie*. 2020;66(1):45-49. doi:10.1016/j.neuchi.2019.09.024.
13. Kumar V, Nair R, Upadhyaya S, Nayal B, Shetty A. Extradural spinal metastasis of adenoid cystic carcinoma (ACC): a case report. *J Clin Diagn Res*. 2015;9(1):XD09-XD10. doi:10.7860/JCDR/2015/10644.5498.
14. Ahmed A, Rajankulam Ganesan SK, Haleem S, Nicoll J. A rare case of late solitary vertebral metastasis from an adenoid cystic carcinoma of the lacrimal gland. *BMJ Case Rep*. 2017;2017:bcr2016218371. doi:10.1136/bcr-2016-218371.
15. Marchesini N, Feletti A, Bernasconi R, Ghimenton C, Sala F. Intramedullary spinal cord metastasis from an adenoid cystic carcinoma of the external auditory canal: case report. *Br J Neurosurg* 2021;1-5. doi:10.1080/02688697.2021.1923653.
16. Park JM, Kong JS, Chang KH, et al. The clinical characteristics and surgical outcomes of carcinoma of the external auditory canal: a multicenter study. *J Int Adv Otol*. 2018;14(2):278-284. doi:10.5152/iao.2018.4952.
17. Ebelhar AE, West DS, Aouad RK. Ceruminous adenoid cystic carcinoma of external auditory canal. *J Int Adv Otol*. 2017;13(2):292-294. doi:10.5152/iao.2017.3929.
18. Correia-Rodrigues P, Ramalho S, Montalvão P, Magalhães M. External auditory canal carcinoma: clinical characteristics and long-term treatment outcomes. *Eur Arch Oto-Rhino-Laryngol*. 2020;277(10):2709-2720. doi:10.1007/s00405-020-06019-2.