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

Incidence 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. 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, followed by ceruminous adenocarcinoma not otherwise specified and ceruminous mucoepidermoid carcinoma.

Compared to their benign counterparts, ceruminous carcinomas usually occur at earlier ages, with slight...
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Physical examination revealed tenderness and percussion
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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
perception pain. Bilateral superficial sensation was re-
duced in the cutaneous area corresponding to nerves below
L1. Perineal sensation was abnormal. Lumbar mobility
was limited. Power in knee extension and ankle dorsi-
flexion 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 insti-
tution suggested multiple abnormal signals of the cervical
vertebrae, thoracic vertebrae, lumbar vertebrae, pelvicones, 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 pre-
vent 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 chemo-
embolization under guidance of Digital Subtraction An-
giography (DSA) was performed. The symptoms in both
lower extremities of the patient were significantly allevi-
avated after operation.

The patient underwent posterior L2 vertebrae tu-
mour resection and biopsy with spinal canal decom-
pression, 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 carci-
oma (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
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Discussion
Ceruminous carcinoma of the EAC is extremely rare.8
According to the latest World Health Organization Class-
ification of head and neck tumours, ceruminous carci-
nomas are classified as ceruminous adenocarcinoma,
adenoïd cystic carcinoma, and mucoepidermoid carci-
noma.9 ADCC, the most common ceruminous malignant
tumour, has a relatively favourable outcome compared to
the other 2 types.6,8 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
metastases 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
ceruminous gland in the EAC is exceedingly rare. Marchesini et al.\textsuperscript{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 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 malignancies have less specificity, manifesting with otorrhea, otalgia, and hearing loss, leading to neglect from both patients and attending doctor at the early stage. Similarities in histomorphology magnifies the difficulty of accurate diagnosis. 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.

Despite its pernicious and malignant nature, ceruminous ADCC are slow-growing and may be associated with a prolonged clinical course. 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. Patients with a longer duration of symptoms before diagnosis also have high recurrence risks. 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. Perzin et al. 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.

Since first resection of mass in EAC 26 years ago, our patient experienced a long clinical course. Dong et al. 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. Bonaparte et al. 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

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).
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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