Clinical Case Report

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Desmoplastic small round cell tumor of the middle ear

A case report

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Abstract

Rationale: Desmoplastic small round cell tumor (DSRCT) is a rare, aggressive and malignant tumor. This report describes a case involving DSRCT of the middle ear which no case has been reported in the literature till date.

Patient concern: A 59-year-old Chinese man with a 40-year history of repeated suppuration of his right ear and 1-year history of drooping of the angle of mouth. The CT of the middle ear and brain scan and enhanced MRI showed space occupying lesion in the right middle ear.

Diagnoses: Desmoplastic small round cell tumor of the middle ear.

Interventions: After relevant examinations, radical mastoidectomy and subtotal temporal bone resection were performed on the right ear under general anesthesia. The patient underwent postoperative adjuvant chemoradiation therapy.

Outcomes: The patient was counterchecked regularly,there was norecurrence of DSRCT of the middle ear. Four years after surgery, the CT and MRI of the middle ear mastoid showed right middle ear soft tissue shadow,but postoperative pathological results showed proliferative fibrous and vascular tissues with chronic inflammatory cell infiltration and necrosis.

Lessons: DSRCT is a relatively aggressive, malignant mesenchymal tumor, with a very poor prognosis.The diagnosis of DSRCT relies on immunohistological data. Early diagnosis, radical surgery, chemotherapy, and radiotherapy are considered a reasonable way to prolong survival.

Abbreviations: CT = computed tomography, DSRCT = desmoplastic small round cell tumor, MRI = magnetic resonance imaging.

Keywords: chemoradiation therapy, desmoplastic small round cell tumor, radical mastoidectomy, subtotal temporal bone resection

1. Introduction

Desmoplastic small round cell tumor (DSRCT) is a rare, aggressive, and malignant tumor primarily affecting young males. No standard therapy is currently available for patients with DSRCT, and DSRCT has a very poor prognosis, and its median survival range is from 17 to 25 months, with only a 29% actuarial 3-year survival rate and a 5-year survival rate of 18%.[1] It is characterized by a unique chromosomal translocation which leads to failure to suppress tumor growth. This tumor most commonly originates in the abdomen, and is also found in the pleural fluid, lungs, parotid gland, and other serosal surfaces. But no case of middle ear DSRCT has been reported in the literature till date. In this report, we describe a case involving DSRCT of the middle ear.

2. Case report

A 59-year-old Chinese man with a 40-year history of repeated suppuration of his right ear and 1-year history of drooping of the angle of mouth was admitted to the Department of Otolaryngology Head and Neck Surgery at our hospital in March, 2013. He also had a history of hearing loss, tinnitus in the right ear, and recurrent headache. He had no relevant personal or family history of malignancy. On physical examination, the right external auditory

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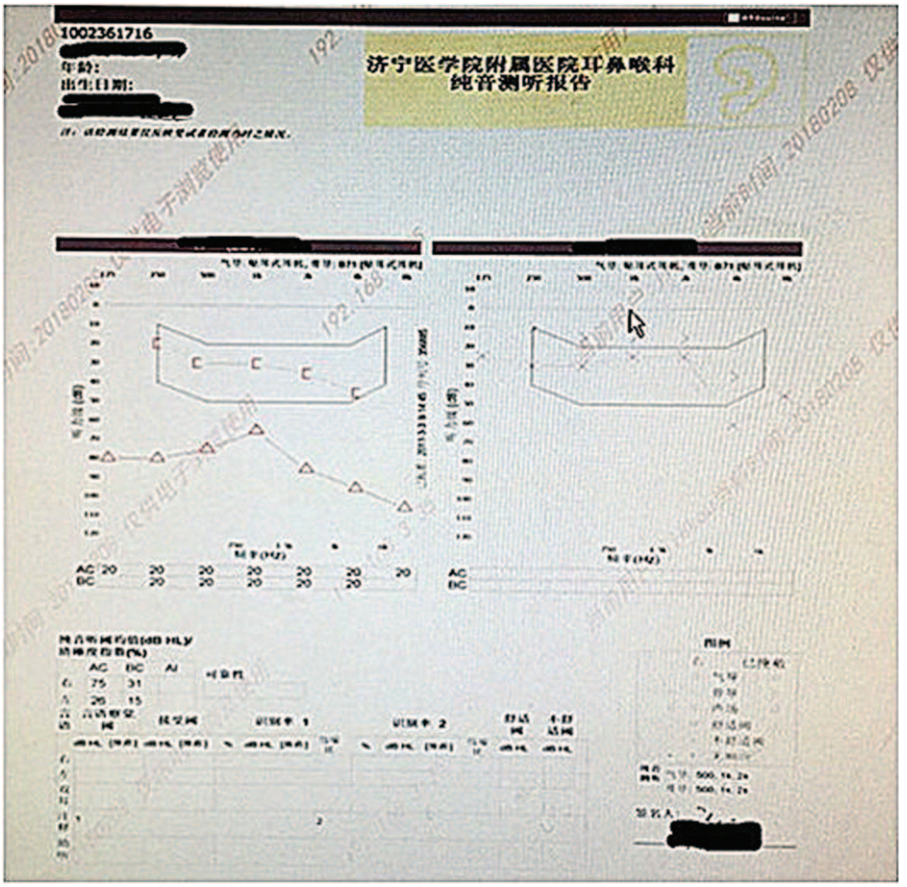

Figure 1. Pure tone audiometry: severe conduction deafness in the right ear.

canal was swollen and narrow, showing purulent discharge and the eardrum of the right ear was incompletely attended, the wrinkles on the right forehead disappeared, hypophasis or lagophthalmos, the right nasolabial groove became shallow, drum gills leak, and the corner of the mouth was skewed to the left. The results of pure tone audiometry show severe conduction deafness in the right ear (Fig. 1). Computed tomography (CT) of the middle ear of Shanghai Seventh People's Hospital showed space occupying lesion in the right middle ear (Fig. 2A). In our hospital, the CT of middle ear mastoid showed disappearance of right mastoid gas chamber, appearance of soft tissue shadow, and extensive destruction of the local bone. The soft tissue shadow is communicated with the

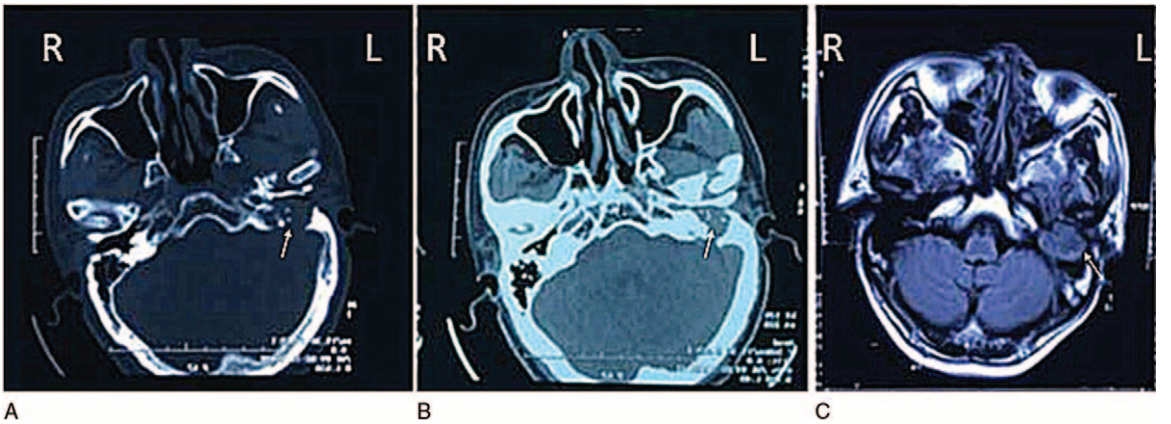

Figure 2. (A) Computed tomography (CT) of middle ear of Shanghai Seventh People's Hospital showing space occupying lesion in the right middle ear. (B) CT of the middle ear mastoid showed disappearance of right mastoid gas chamber, appearance of soft tissue shadow, and extensive destruction of the local bone. The soft tissue shadow was communicated with the middle cranial fossa and the cerebellar hemisphere. (C) The brain scan and enhanced MRI showed right middle ear mastoid mass, neoplastic lesion (cumulative right cranial fossa) and local mastoiditis. MRI = magnetic resonance imaging.

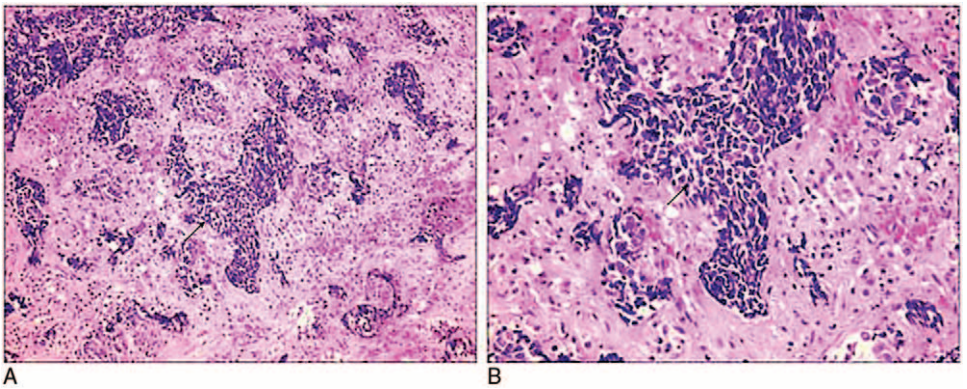

Figure 3. Routine morphology. (A) Tumor nests with a fibromyxoid stroma (H&E, original magnification ×20). (B) Tumor nests with a fibromyxoid stroma (H&E, original magnification ×40).

middle cranial fossa and the cerebellar hemisphere (Fig. 2B). The brain scan and enhanced magnetic resonance imaging (MRI) showed right middle ear mastoid mass, neoplastic lesion (cumulative right cranial fossa), and local mastoiditis (Fig. 2C). Surgical treatment of the tumor was performed in our department. Intraoperatively, the mastoid, tympanic membrane, and tympanum are filled with fish-like changes, bone destruction, facial nerve has been eroded and partly disappeared, rapid pathology of the tumor demonstrated small cell malignancy. We carried out radical mastoidectomy and subtotal temporal bone resection, with an extended resection of the mastoid. The mass of the tumor was extending forward into the petrous apex, upward adhesion with the dura mater, backward adhesion with the sigmoid sinus, inward deterioration of the facial nerve and semicircular canal, and downward contact with the glomus jugulare. Subtotal resection of the temporal bone was carried out, which showed electrocoagulation of dural and sigmoid sinuses, and other suspicious regions. Muscle flap surgery was performed to fill the surgical cavity filled with gelatin sponge and gauze iodoform. Then the incision was sutured, and then the local pressure dressing was applied. Postoperative pathology examination showed small cell malignant tumor, and morphology confirms stimulation of fibrous hyperplastic small round cell tumors (Fig. 3A and B). The patient received the vincristine, actinomycin D, and cyclophosphamide multiagent systemic chemotherapy regimen consisting of vincristine (2 mg, day 1), adriamycin (75 mg/m^2, day 1), and cyclophosphamide (1.2 g/m^2, day 1) in the Department of Oncology after the operation. The patient received radiotherapy after chemotherapy (180 Gy/session; total dose 4500 Gy). There was no recurrence of DSRCT of the middle ear and required no mastoid scan, enhanced CT (Fig. 4A), and MRI (Fig. 4B). In May, 2017, we discovered a reddish mass with smooth surface in the right ear external auditory canal on endoscopic examination in our hospital (Fig. 5A). CT of the middle ear mastoid showed right middle ear soft tissue shadow and bone destruction (Fig. 5B). The middle ear scan and enhanced MRI showed right middle ear tumor after surgery, right middle ear-inner ear abnormal signals, and right external auditory canal nodules, confirming chronic inflammation of the surgical region and nodular lesion of the external auditory canal (Fig. 5C). No tumor metastasis was detected by whole body bone scan and chest CT. External auditory canal lesion excision was performed in our department in May, 2017. The intraoperative frozen tissue showed fiber hyperplasia and vascular tissue with chronic inflammation and necrosis. Postoperative pathological results showed proliferative fibrous and vascular tissues with chronic inflammatory cell infiltration and necrosis. Immunohistochemical evaluation exhibited the following results of CD99 (−), CK (−), SMA fraction (+), ki-67 (+, 1%–3%).

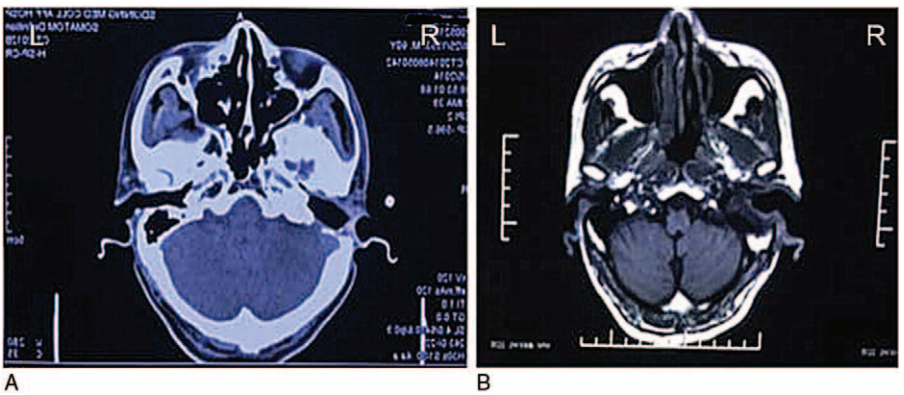

Figure 4. (A) Computed tomography (CT) of the middle ear and mastoid scan showing postoperative changes in the right middle ear carcinoma. (B) Magnetic resonance imaging (MRI) of middle ear and mastoid showing postoperative changes of the right middle ear carcinoma.

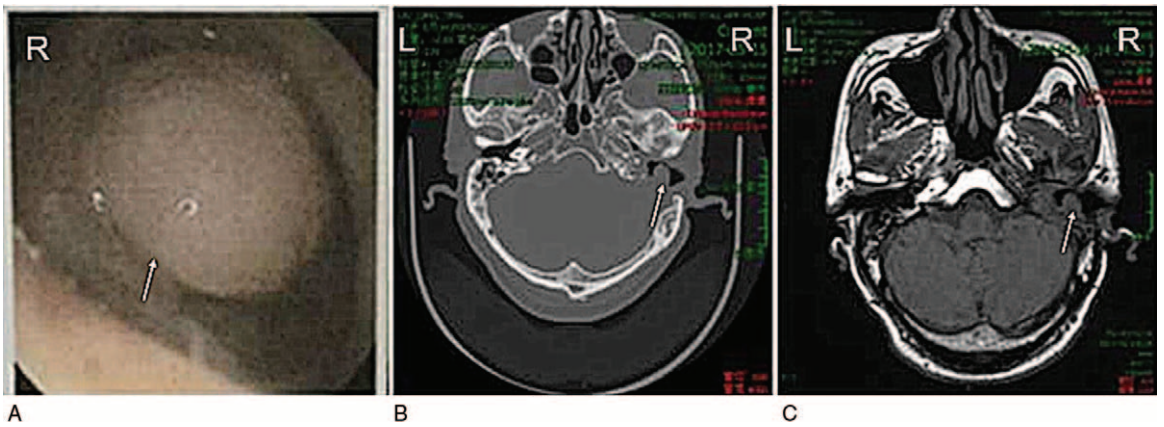

Figure 5. (A) Endoscopic examination showing smooth surface reddish neoplasm in the right ear external auditory canal. (B) CT of the middle ear mastoid showing right middle ear soft tissue shadow and bone destruction. (C) Middle ear scan and enhanced MRI showing right middle ear tumor after surgery, right middle ear-inner ear abnormal signals, and right external auditory canal nodules, confirming chronic inflammation of the surgical region and nodular lesion of the external auditory canal. CT = computed tomography, MRI = magnetic resonance imaging.

3. Discussion

Desmoplastic small round cell tumor is an extremely rare, highly aggressive, and malignant mesenchymal neoplasm of undetermined histogenesis. It was first described by Gerald and Rosai in 1989 as a newly featured clinicopathologic entity.[2] It is prevalent in adolescents and young adults, and the incidence in males was 3 times more than that in females.[3] Majority of DSRCTs originate from the abdominal and pelvic cavities,[4] but beyond that, few stem from the retroperitoneum, omentum, and mesentery, in addition to multiple peritoneal tumors. The mechanism of histogenesis of this tumor remains unclear, but might be derived from multipotential differentiated primitive mesenchymal cells or neuroectodermal and primitive mesenchymal tissue.

The diagnosis of middle ear DSRCT remains challenging. Because most DSRCTs are found in the abdominal cavity, some nonspecific gastrointestinal clinical signs and symptoms including nausea, vomiting, abdominal distension and pain, a palpable abdominal mass, are the main problems leading to patients visiting a doctor. In our case, the patient was presented with otorrhea and bleeding, and no significant difference was observed with other common diseases of the middle ear. Hence, early detection is not easy, and can be easily misdiagnosed and missed. DSRCT was diagnosed based on the clinical signs and symptoms, and also radiological findings, but mainly based on the pathological and immunohistochemical examination. DSRCT exhibits a characteristic t(11; 22) (p13; q12) chromosomal translocation that brings the EWS gene on chromosome 22 to the WT1 gene on chromosome 11. The EWS-WT1 protein can attenuate tumor suppressor function of EWS gene and promote the expression of growth factor which is inhibited by normal WT1 gene.[5] The EWS-WT1 gene fusion protein serves as a disease-specific marker and yields a definitive diagnosis of DSRCT.[6]

Currently, there are no consensus management and therapeutic protocols for DSRCT.[7] This is attributed to its extreme rare incidence, high aggressive behavior, and lack of validated staging system for DSRCT. Apart from this, DSRCT is a rapidly progressing and prone to extensive lymphogenous and hematological metastases in the early stages. Liver and lungs are the 2 most common regions involved in distant hematogenous metastases.[7,8] At present, the treatment of DSRCT is still under research. Radical surgical resection, radiotherapy, and chemotherapy are possible therapeutic strategies, although none of them appeared to have better efficacy compared to others. Aggressive radical surgical resection is still the mainstay of treatment for DSRCT. Radical surgical resection is defined as definitive removal of at least 90% of the tumor burden.[9] Radical surgical resection was highly significant in prolonging the overall survival. It has been reported that the 3-year survival was 58% in patients treated with gross tumor resection compared with no survivors past 3 years in the nonresection cohort.[10] But recent studies showed that surgical resection does not improve survival in patients with distant metastasis.[11] DSRCT has a high degree of malignancy. Patients often have distant metastasis before visiting the doctor. It is difficult to completely remove, and easy to relapse after surgery. So, radical surgery, combined with full height-dose chemotherapy or local radiotherapy, is still the best treatment for survival.[12] Chemotherapy can prolong survival to some extent. At present, the most commonly administered chemotherapeutic agents for DSRCT include doxorubicin, cyclophosphamide, vincristine, ifosfamide and etoposide. Despite the combined therapeutic approach, DSRCT still have a very poor prognosis, with only a 29% actuarial 3-year survival rate and an 18% five-year survival rate.[1]

In our case, the patient underwent radical surgical resection at an early stage. Also, he received active systemic chemotherapy and radiotherapy after the operation. After a 4-year follow-up, he presented with no recurrences. So, surgery combined with chemotherapy and radiotherapy can prolong the survival to some extent.

Desmoplastic small round cell tumor is a relatively aggressive, malignant mesenchymal tumor, with a very poor prognosis. The diagnosis of DSRCT relies on immunohistological data. Early diagnosis, radical surgery, chemotherapy, and radiotherapy are considered a reasonable way to prolong survival.

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Author contributions

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