Rhabdomyosarcoma of the External Ear Canal Treated with Brachytherapy

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Abbreviation Key

<table>
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<th>Abbreviation</th>
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<td>RMS</td>
<td>Rhabdomyosarcoma</td>
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<td>EBRT</td>
<td>External Beam Radiation Therapy</td>
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ABSTRACT

Rhabdomyosarcoma (RMS) is the most common pediatric soft tissue sarcoma. RMS of the ear usually involves the middle ear cavity. Treatment usually involves surgery, multi drug chemotherapy regimen, and high dose external beam radiotherapy (EBRT). We present a case of a 17-month-old Caucasian female with an unusual presentation of RMS localized to her external ear canal only, which was initially believed to be an otitis media. Treatment consisted of a regimen involving brachytherapy as the radiation modality rather than EBRT. The goal of brachytherapy use is to decrease morbidity associated with radiation of the head and vital organs in developing children.
INTRODUCTION

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children. It accounts for about 50% of all soft tissue sarcomas and 5% of pediatric cancers. While RMS of the ear is rare, it is the most common sarcoma of the ear and usually involves the middle ear cavity [1]. It is unusual for RMS to present at the external ear canal. Generally non-parameningeal RMS of the head and neck has a good prognosis. In patients with unresectable tumors, treatment consists of surgery, chemotherapy, and radiation. Radiation to the head and neck region in a young child is associated with complications due to proximity of the brain. The AMORE protocol for RMS of the head and neck was introduced in 1993 in an attempt to introduce brachytherapy as a treatment, which would reduce the long-term side effects that were noted with EBRT use in the head and neck region. Despite the introduction of this protocol, brachytherapy is still not a widely used treatment modality.

CASE DESCRIPTION

A 17-month-old Caucasian female presented to the office of her pediatrician in May 2015 due to the presence of dry blood on the outside of her right ear one day prior. There was no reported fever, ear pain, ear tugging, or other complaints. On examination of the patient’s right ear, a clustered mass was noted at the external auditory meatus (Figure 1). The patient was prescribed a course of antibiotics for possible otitis. Five days after antibiotic therapy was initiated, the patient’s mother noted that the mass was increasing in size. At this point the patient was taken to an otolaryngologist who cleaned the ear canal and revealed a polypoid mass. Biopsies were obtained and the pathology was consistent with embryonal rhabdomyosarcoma. Subsequently, an MRI, CT, bone scan, and bone aspiration and biopsy were done for staging purposes.

The patient was referred to the pediatric oncology department for further management. Patient was deemed to be Stage 1(non parameningeal head and neck tumor), Group III (Incomplete resection with gross residual disease ) according to the soft tissue sarcoma committee of the Children’s Oncology Group (COG) risk group and the Intergroup Rhabdomyosarcoma Study Group (IRSG) clinical grouping system. Surgery was deemed to be too invasive (with potential for unacceptable cosmetic deficits), hence the patient was started on
a chemotherapy regimen consisting of Vincristine, Dactomycin, and Cyclophosphamide. Follow up imaging at week 13 revealed a marked decrease in the size of the tumor and there was no visible mass on examination; however biopsy of the tumor bed revealed viable tumor cells. We decided on brachytherapy for local tumor control as the morbidity of conventional radiation this close to the brain would be significant. She received 7 treatments for a total dose of 21 Gy to the external auditory canal. Following brachytherapy she has continued chemotherapy per protocol. She is currently doing well with no evidence of disease. The patient’s hearing has not been affected by the tumor and she is achieving age appropriate developmental milestones.

**DISCUSSION**

Rhabdomyosarcoma is a malignant tumor of muscle tissue. Rhabdomyosarcoma has been classified into two main subtypes, of which embryonal is the most common, comprising 50-70% of cases. Alveolar occurs in 20-30% of cases, and 5% of occurrences are pleomorphic.[2] Embryonal rhabdomyosarcoma is generally associated with males, younger children, and is frequently diagnosed in the head and neck, bladder, or genital regions. Alveolar RMS has no gender or age bias and comprises approximately 1 case per 1 million uniformly from 0 to 19 years old.[3] Approximately 35% of RMS cases occur in the head and neck region.[4] Rhabdomyosarcoma of the head and neck region falls into three categories, 51% of cases occur in the parameningeal region (nose, paranasal sinuses, middle ear, mastoid), 24.5% of cases present in the non-parameningeal region (Parotid gland, oral cavity, larynx), and 24.5% are in the orbit. [5]

RMS of the ear is rare, with only about 3% of patients presenting with middle ear disease.[4] The symptoms are nonspecific such as pain and bloody discharge, which is commonly misdiagnosed and treated as otitis media, thus leading to a delayed diagnosis. The mean time of onset of symptoms to diagnosis of RMS is approximately 21 weeks, which also delays the start of treatment.[6] This delay can result in tumor growth causing cranial nerve deficits, as well as meningeal or brain parenchyma involvement.[7] It is not unusual for RMS of the ear to present concurrently with facial nerve palsies and loss of taste at the anterior 2/3 of the tongue. Our patient fortunately was diagnosed early in the course of the disease (within 10 days
of initial symptomatology), which may have prevented further involvement of surrounding structures.

The patient described in this case report had an unusual presentation of RMS at the external ear canal, to our knowledge this is a rare location for RMS to arise from. While there are multiple case reports regarding RMS of the middle ear canal there are very few case reports of RMS of the external ear canal. In 1966 Potter describes the case of a 3yo male who was originally diagnosed with bilateral otitis media which was later revealed to be a right sided RMS of the middle ear canal which had extended to the external ear canal.[8] Similar to our patient there is an initial misdiagnosis of otitis media due to the strong overlap of presenting symptoms. In 2014 a case was described of a 12-year-old girl who presented with a left ear canal mass, which histology revealed to be RMS. [9]

CT scan will evaluate any bone destruction and MRI can evaluate dural involvement and tumor proximity to the carotid artery and jugular vein.[4] Other destructive lesions of the temporal bones are radiographically similar to RMS, therefore a biopsy is crucial to the diagnosis. Tissue stains help differentiate RMS from other tumors with similar histopathology, such as lymphomas (CD20+ and CD3 +), and Ewing's Sarcoma (CD 99+).[10] RMS will be positive for multiple muscle markers, the most specific and sensitive marker is the muscle transcription factors MyoD and Myogenin.[11]

The standard of treatment for RMS includes excisional surgery, multidrug chemotherapy, and EBRT. EBRT has many side effects due to the collateral damage of surrounding anatomical structures. In the head and neck region ERBT can result in hearing loss, facial hypoplasia, developmental delay, xerostomia, dental problems, problems with the pituitary/hypothalamic hormones and second malignancies.[12, 13] Brachytherapy is comparatively less damaging because of the localized mechanism of action, which should spare the surrounding tissue.

The AMORE protocol consists of Ablative surgery, MOId technique with afterloading brachytherapy and immediate surgical Reconstruction after chemotherapy. Patients with head and neck RMS treated with EBRT were found to have higher incidences of hearing loss compared to survivors who had received AMORE based therapy. [12] While our patient’s treatment did not follow the AMORE protocol, her treatment did consist of the use of brachytherapy rather than EBRT. EBRT in head and neck RMS patients is also associated with an increased risk to develop any grade 3 event or more than 5 adverse events compared with
survivors who received brachytherapy based treatments. The 5-year overall survival after EBRT was 75%, which is comparable to 76.9% survival rate of AMORE based treatment.\[14\] The subjective impression is that the late side effects of brachytherapy were less significant and less frequent than with EBRT.\[15\] Overall survival rates are similar between EBRT and brachytherapy the quality of life is important and can be implied from the side effects noted in each treatment protocol.

In the case described, our patient received brachytherapy in the middle of her chemotherapy regimen. She has demonstrated a remarkable shrinkage in her tumor size such that surgery is not necessary. There has been no demonstrated involvement of surrounding structures and no further intervention for reconstructive purposes has been required.

**CONCLUSION**

This case demonstrates how the presentation of rhabdomyosarcoma at an unusual location can be misdiagnosed for a common benign pathology. Delay in diagnosis can result in further growth of the tumor, nerve damage, and worse. Maintaining a wide differential for recurrent non-remitting otitis media is important for this reason. We also demonstrate the use of brachytherapy, as treatment in attempt to minimize the long-term sequele patients with RMS of the head and neck region would normally experience with conventional radiation.
Figure 1: Rhabdomyosarcoma of the external ear canal
REFERENCES


