Rhabdomyosarcoma of the middle ear cavity presenting with Horner's syndrome

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ABSTRACT

Rhabdomyosarcoma is a rare malignant tumour of striated muscle accounting for 3% of all primary soft tissue tumours of the middle ear cavity. We describe a unique case of a rhabdomyosarcoma of the middle ear cavity in a four-year-old boy, presenting as an otherwise asymptomatic ipsilateral Horner's syndrome. This case involved tumour erosion through the tensor tympani semicanal, accessing the carotid canal and the sympathetic plexus running through it. This child was treated successfully with chemotherapy and proton beam therapy, and remains disease free at six years of follow-up. This case highlights the importance of investigating patients with non-resolving ear infections, polyps or granulation tissue.

KEYWORDS

Rhabdomyosarcoma – Horner's syndrome – Middle ear – Chronic suppurative otitis media

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Introduction

Rhabdomyosarcoma is a highly aggressive, locally invasive, malignant tumour of striated muscle, which often carries a very poor prognosis. It remains exceptionally rare, with an incidence estimated at 0.44 per 100,000 in caucasian patients and 0.13 per 100,000 in Afro-Caribbean patients.¹ Although rare, it currently accounts for approximately 60% of all soft tissue tumours in children with a mean age of presentation of 4.5 years old.^{2,3} Rhabdomyosarcomas vary widely in their anatomical presentations and it is estimated that 30–50% present in the head and neck region,^{4–5} with only 3% presenting within the middle ear or temporal bone. Within the literature, rhabdomyosarcoma of the middle ear cavity has been described to be associated with VII, VIII, IX, X and XI cranial nerve palsies.^{3,5} This is the first case within the literature describing a rhabdomyosarcoma of the middle ear cavity presenting with Horner's syndrome.

Case history

A four-year-old boy presented with a three-week history of persistent right-sided otalgia, right-sided partial ptosis and asymmetrical pupils. The child complained of no other symptoms, was systemically well and had no significant past medical history except episodes of recurrent self-limiting acute otitis media.

On examination, there was a right-sided partial ptosis with an associated right-sided constricted pupil (Horner's

syndrome). Colour vision, visual acuity and visual fields were normal and all cranial nerves were intact. Examination of the right ear revealed a dry, perforated tympanic membrane, with extruding granulation tissue. Neck examination revealed a tender, enlarged lymph node in the right cervical chain. He was afebrile with all clinical observations normal for his age.

Full blood count, white cell count, C-reactive protein and renal function tests were all within normal limits. Chest x-ray revealed no abnormalities and urgent computed tomography of the petrous temporal bones and brain was performed, followed by magnetic resonance imaging (MRI). The CT scan (Fig 1) showed opacification of the right middle ear cleft and mastoid process with adjacent dehiscence of the bony wall overlying the right carotid canal. The ossicles appeared intact with no breach of the mastoid air cells seen. The left middle ear cleft and mastoid process were normal, as too was the brain. MRI confirmed the CT findings, but also identified an occlusion of the right internal carotid artery at the level of the skull base closely related to the right middle ear and mastoid abnormalities.

An urgent elective examination under anaesthesia of his right ear was undertaken, revealing excessive middle ear granulation tissue. Biopsies were taken and sent for histopathological examination, which revealed scattered large cells containing irregular, hyperchromatic nuclei associated with a loose, myxoid stroma (Fig 2). Immunohistochemistry showed positive staining with desmin and myogenin, consistent with a diagnosis of rhabdomyosarcoma (Fig 3). The

Figure 1 Axial computed tomography showing complete opacification of the right middle ear cleft with bony destruction of the right carotid canal (asterisk).

Figure 2 Medium-powered photomicrograph showing sub-epithelial atypical cells including occasional large hyperchromatic nuclei adjacent to an area of necrosis. (haematoxylin and eosin $\times 20$).

patient underwent a twelve-month course of vinblastine chemotherapy with prednisolone under the direction of the paediatric oncology team. Following this, proton beam therapy (50.4 Gy) was undertaken in the United States. Follow-up at six years revealed no evidence of recurrence or metastasis (Fig 4).

Discussion

Four histological categories of rhabdomyosarcoma have been described within the literature and clinical practice: embryonal, pleomorphic, alveolar and botryoid.⁵ The vast majority of head and neck rhabdomyosarcomas, and indeed the case discussed here, are that of the embryonal

type, named as such because the cells resemble those present in skeletal muscle development.⁵ Rhabdomyosarcomas spread locally and metastasise to distant sites via haematological means, with the most common sites for secondary deposits being the lungs and bone.

Presenting symptoms of a rhabdomyosarcoma depend entirely on the anatomical area affected and region of invasion. By far the most common and earliest symptoms of a middle ear cavity rhabdomyosarcoma is that of chronic suppurative otitis media with or without granulation of the tympanic membrane and aural polyps.⁵ Spreading locally, the tumour quickly extends beyond the middle ear cavity, invading neighbouring structures encased within the temporal bone, developing new signs and symptoms depend-

Figure 3 Immunohistochemically stained photomicrograph with desmin (left) and myogenin (right) highlighting atypical cells beneath the epithelium, diagnosing rhabdomyosarcoma ($\times 20$).

Figure 4 Axial magnetic resonance image through the mastoid, revealing no evidence of disease recurrence at five years.

ing upon the path of destruction. Facial nerve involvement is commonly described, as well as the development of sensorineural hearing loss and vertigo.^{5,5} Other cranial nerve palsies described as a result of rhabdomyosarcoma invasion include cranial nerves IX, X and XI⁵.

This case describes a rhabdomyosarcoma, presenting with symptoms of right-sided chronic suppurative otitis media with a right-sided Horner's syndrome. Using a

combination of the CT and MRI images, intraoperative findings and the clinical history, the most likely striated muscle fibres responsible for the tumour would be those of the tensor tympani. Here, the tumour appears to have eroded through the tensor tympani semicanal and through a thin portion of petrous temporal bone, accessing the carotid canal and involving the sympathetic plexus running through it. These nerves provide sympathetic fibres to the head and neck and are involved in raising the eyelids, dilating the pupils, controlling facial sweat glands and contributing to vasoconstriction of blood vessels. Tumour involvement of the sympathetic plexus in this case, resulted in a unilateral Horner's syndrome.

This case not only represents a unique presentation of rhabdomyosarcoma of the temporal bone, but also highlights the difficulty in diagnosis of middle ear soft tissue tumours in children. Early diagnosis and treatment, using a multidisciplinary team, significantly improves clinical outcome. All children with a persistent suppurative otitis media, unresponsive to medical management, should therefore be considered for an early examination and biopsy of the middle ear cavity under anaesthesia. This report also highlights the importance of early imaging, especially in the presence of polyps or granulations.

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