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Rhabdomyosarcoma of the Middle and Inner Ear

Abstract: Rhabdomyosarcoma of the middle ear is a rare tumor. It may be locally invasive or spread by distant metastasis. It generally has a poor prognosis. We describe a case of rhabdomyosarcoma of the middle ear with extension to cavernous sinus, internal auditory and carotid canals.

Keywords: Rhabdomyosarcoma, Ear Neoplasm

Introduction

Rhabdomyosarcoma is the commonest soft tissue sarcoma in children under 15 years of age. It occurs predominantly in three regions; head and neck, genitourinary tract (retro-peritoneum) and extremities. Ear and mastoid bone are rare sites for rhabdomyosarcoma. Most cases of ear rhabdomyosarcoma show involvement of middle and external ear at the time of diagnosis.[1] Herein, we report on a patient with rhabdomyosarcoma affecting middle and inner ear, with meningeal extension and an intact tympanum.

Case Presentation

A five-year-old boy presented with sudden onset right eye deviation of one month duration. He also complained of inability to close his right eye and deviation of the right corner of his mouth for two weeks. He had no pain. In addition, he had a properioceptive defect and ataxia. On physical examination, there was right-sided sixth cranial nerve palsy and complete peripheral right facial nerve palsy. No nystagmus was detected. Some eardrum retraction was also observed. There was no detectable lymphadenopathy on palpation.

The right ear was reported to be deaf by means of audiometry. Tympanometry showed no movement of the right eardrum.

On axial CT scan, a lytic lesion was detected in the apex of the right petrous bone with involvement of mesotympanum and the eustachian tube (Figure 1-A). MRI revealed a space occupying lesion which was iso-signal on T_1 and hyper-signal on T_2 weighted images (Figure 1-B). Subsequent to Gadolinium administration, an abnormal enhancement of the cavernous sinus and internal auditory canal was observed (Figure 1-C).

The tumor was resected by a transmastoidal approach. Extensive involvement of cochlea, internal auditory canal, jugular foramen and cavernous sinus was observed at surgery. The surgical specimen was reported as embryonal rhabdomyosarcoma. Chemo-radiotherapy was begun after the pathology report.

Discussion

Rhabdomyosarcoma is a malignant tumor that usually arises from the sites where striated muscle tissue is normally absent (*e.g.*, in the common bile duct and urinary bladder) or where striated muscle is scanty (*e.g.,* in the nasal cavity, middle ear and vagina).

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Table 1: Rhabdomyosarcoma: Prognostic indicators

	Favorable prognostic factors	Unfavorable prognostic factors
Age	Infancy through childhood	Adulthood
Location	Orbit	Non-orbital, head and neck, paraspinal, abdomen, extremities
Size	< 5 cm	>5 cm
Subtype	Embryonal	Alveolar
Tumor spread	Localized; non-invasive without lymph node involvement or metastasis	Local invasion especially parameningeal or paraspinal, lymph node involvement; recurrence during therapy, incomplete surgical resection
Cytogenetic	-	Diploid DNA content

Rhabdomyosarcoma is the commonest soft tissue sarcoma in children under 15 years of age but is very rare in persons older than 45.[1] It is the commonest sarcoma of the ear.[2] Genetic factors may influence its development, as the disease rarely occurs in siblings[3] and is associated with other neoplasms such as congenital retinoblastoma,[4] familial polyposis,[5] and pleuro-pulmonary blastoma. [1]

Rhabdomyosarcoma has been classified in different ways; based on clinical and pathologic features (*e.g.,* Horn and Enterline classification, WHO classification and Intergroup Rhabdomyosarcoma Study); based on clinical behavior and cellular differentiation (*e.g.,* International Society for Pediatric Oncology), or based on prognosis (*e.g.,* International Classification of Rhabdomyosarcoma).

Embryonal, alveolar and pleomorphic types are the best known subtypes of rhabdomyosarcoma. The embryonal subtype accounts for approximately 49% of all rhabdomysarcomas.[6] It mostly affects children younger than 10 years of age and is rare in patients older than 40. It occurs mostly in head and neck, particularly in orbit and parameninges. Following head and neck, this tumor subtype is most likely occur in the genitourinary tract followed by deep soft tissues of the extremities, pelvis and retroperitoneum.[1] Histologically, embryonal rhabdomyosarcoma bears a close resemblance to various stages of the embryogenesis of normal skeletal muscle, however, with a much more variable pattern.[1] The spindle cell type of embroyonal rhabdomyosarcoma is more common in males, particularly in paratesticular region, and has a more favorable outcome.

The botryoid type of the embroyonal rhabdomyosarcoma usually has a grape-like macroscopic appearance and is usually found in the mucosal lining of the hollow viscera such as nasal cavity, nasopharynx, bile duct, urinary bladder and vagina.

Alveolar rhabdomyosarcoma, the second commonest subtype (31%), occurs more commonly in 10 to 25 years age group and has a predilection for the deep soft tissues of the extremities. It accounts for approximately 50% of all rhabdomyosarcomas of the extremities.

Pleomorphic rhabdomyosarcoma is a rare variant that almost always arises in adults older than 45 years of age. This subtype is more common in males and most usually arises in skeletal muscles of the extremities, particularly the thigh.[1]

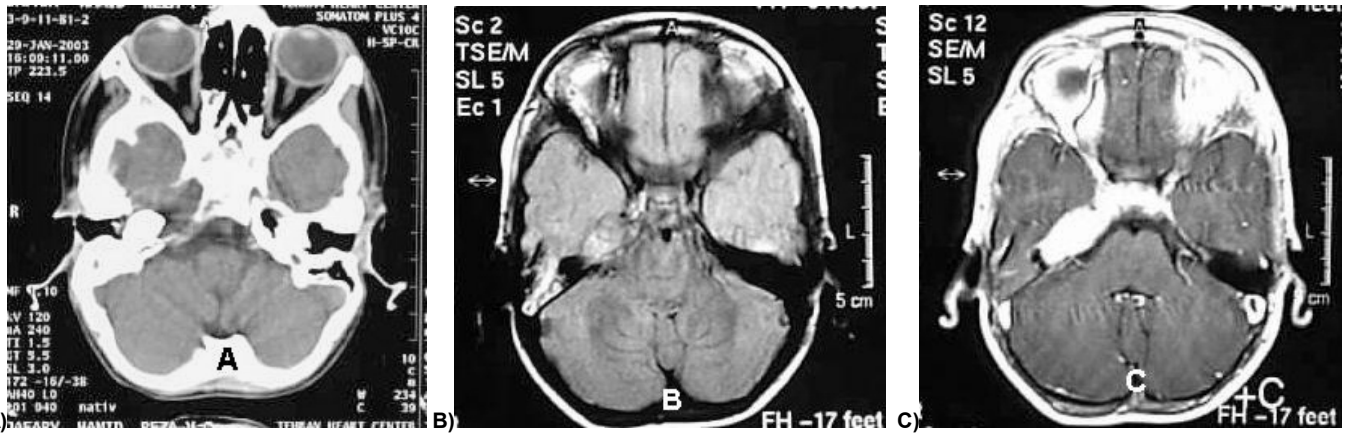

Figure 1: **A)** Plain CT scan: Tumoral lesion with erosion of the apex of the temporal bone and extension to the eustachian tube. **B)** Axial T_1-WI MRI: The tumor reachs the cavernous sinus and internal auditory canal. **C)** Axial enhanced MRI: Intense enhancement is noted in the tumor and adjacent meninge.

Rhabdomyosarcoma predominantly occurs in three regions; 1) head and neck, 2) genitourinary tract and retro-peritoneum, and 3) extremities.

The head and neck is the principal location of rhabdomyosarcoma (30%–50%).[7]

The most common site involved in the head and neck are nasopharynx and orbit. In 558 cases of rhabdomyosarcoma reviewed by the Armed Forces Institute of Pathology, only 17 (3%) cases were in the ear or mastoid.[1] Ear rhabdomyosarcoma may begin either in the muscles of the eustachian tube, in the proper middle ear, or from the primitive pluripotential mesenchymal rests.[8] However, usually at diagnosis there is a widespread local invasion throughout the petrous bone.[7]

Clinically, rhabdomyosarcoma of the middle ear is manifested initially as a chronic otitis media. The clinical progression is quite fast. The facial nerve palsy is usually present at the diagnosis.[7]

Physical examination usually reveals a gray-to-purple, fleshy friable polypoid mass in the ear canal penetrating the tympanum that infiltrates the surrounding soft tissue, hence, causing diffuse swelling in the peri-auricular region.[1]

Involvement of the apex of the petrous bone, internal auditory canal and the base of skull may lead to multiple cranial nerve palsies.[9] Approximately 30% of these patients will already have some neurologic deficits at the diagnosis. Major metastatic sites are the pulmonary lymph nodes and bone marrow.

Among the imaging modalities, CT scan is the technique of choice for evaluation of bone destruction. Aggressive bone destruction with obliteration of the normal landmarks of the skull base occurs in up to 67% of middle ear tumors.[7]

MRI is more efficient for evaluation of dural involvement with intracranial extension, and to assess the proximity of the tumor to the carotid arteries and jugular veins.

The signal intensity of the tumor is nonspecific, that is, the signal is minimally hyperintense to muscle in T_1-W images and markedly hyperintense to muscle on T_2-W sequences.

The tumor demonstrates intense homogeneous enhancement after Gadolinium administration.[10]

Surgical resection of the tumor followed by irradiation and chemotherapy is the usual treatment. Upon parameningeal involvement, as confirmed by skull base erosion on CT or pleocytosis with elevated protein and decreased glucose level in cerebro-spinal fluid, intrathecal chemotherapy and craniospinal irradiation are recommended.[11] Prognosis depends on several factors as depicted in Table 1.[1,2,11,12]

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