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Case Report

Underlying Rhabdomyosarcoma Mimicking as Otitis Media

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ABSTRACT

Rhabdomyosarcoma (RMS) is one of the most frequently encountered soft tissue sarcoma found in infants and children. The common sites of occurrence are the head and neck region, genitourinary tract, retroperitoneum and to a lesser extent, the extremities. It is rare in the temporal bone. RMS is a highly malignant tumour with extensive local invasions and early haemorrhagic and lymphatic dissemination. Despite aggressive approaches incorporating surgery, dose-intensive combination chemotherapy, and radiation therapy, the outcome for patients with metastatic disease remains poor.

A case of 4 year old female child presenting with facial palsy and ear discharge which was later diagnosed with rhabdomyosarcoma is presented to highlight the importance of taking this diagnosis into account while managing children with persistent ear discharge.

KEYWORDS: Chemo-radiation, Ear discharge, Facial palsy, Post auricular swelling, Rhabdomyosarcoma.

INTRODUCTION

Rhabdomyosarcoma (RMS), a tumour originating from the striated muscle, is the second most common soft tissue sarcoma encountered in childhood after osteosarcoma.[1] Rhabdomyosarcoma is thought to arise from immature mesenchymal cells that are committed to skeletal muscle lineage, but occasionally these tumours arise in tissues in which striated muscle is not normally found, such as urinary bladder. Almost two-thirds of cases of RMS are diagnosed in children <6 years of age although there is another mid-adolescence peak. It is slightly more common in males than in females (1.3-1.5:1).[2,3]

The incidence appears to be lower in Asian populations than among mainly white populations of Western countries.[2] Distinctive features correlate with the site of the primary tumour, the age at diagnosis, and the histologic subtype.[4] It is ubiquitous occurring almost everywhere but most commonly in the head and neck and the genitourinary (GU) areas.

Ear is comparatively a rare site for this neoplasm and accounts for less than 10% of all cases of head and neck.[5] Rhabdomyosarcoma in the middle ear mimics an aural polyp in appearance and the discharge is easily mistaken for otitis media. As a result, advanced disease with meningeal involvement is common at the time of diagnosis.[6]

CASE REPORT

A 4 year old female child presented with complaints of left sided facial asymmetry since 2 months and purulent left ear discharge since 2 months. There were repeated episodes of ear discharge in the past for which antibiotics were prescribed. There were associated complaints of left post auricular swelling and left ear ache. On pressing the swelling, discharge was seen. The child was refusing feeds. There was a grade V (House-Brackmann scale) LMN VIIth nerve palsy accompanied by ptosis and swelling of the left eye. (Fig: 1)

There was tenderness and oedema over the left mastoid. On oral examination, a posterior para-pharyngeal bulge was seen. As complicated otitis media was suspected based on this clinical picture, patient was immediately started on intravenous antibiotics and anti-inflammatory agents.

However, a high resolution computed tomography (HRCT) Temporal bone revealed a large abnormal soft tissue lesion involving the floor of middle cranial fossa causing bony erosion of the left petrous apex and greater wing of sphenoid. The lesion was extending from the middle ear laterally to the clivus medially. In the middle ear, the lesion was involving the internal auditory canal with destruction of the facial nerve canal. Medially, the lesion was extending into the left half of the clivus with bony erosion and destruction of carotid canal and the foramen lacerum. Anteriorly, the lesion was causing erosion of the posterior wall of the left sphenoid sinus and extending into the sphenoid sinus. Lesion was extending from the superior orbital fissure into the intraconal and extraconal portions of the left orbit. The lesion was encasing the optic nerve with loss of fat

planes with the superior rectus, inferior rectus, medial and lateral rectus muscles. There was complete destruction of the foramen ovale and foramen spinosum. Posteriorly, the lesion was causing erosion of sigmoid and dural plate, involving the jugular bulb and extending into the posterior fossa and causing mass effect on the left half of pons and the cerebellum. Laterally, the lesion was causing destruction of the facial nerve canal and extending to the internal auditory canal causing its complete obstruction.

Postero-laterally, lesion was extending into the mastoid air cells with soft tissue opacification of the air cells. Inferiorly, the lesion was extending into the left para-pharyngeal and the left masticator space. The lesion was also causing medial displacement of the tongue.

Magnetic Resonance Imaging (MRI) (Fig: 2) showed features of either a primitive neuroectodermal tumour (PNET) or Rhabdomyosarcoma. It showed a large abnormal soft tissue of altered signal intensity as described above. It measured 6.3× 5.6×4.2 cm in size. The lesion appeared iso intense on T1W and hyperintense on T2W and FLAIR sequences. It showed a heterogeneous post contrast enhancement. Few non-enhancing/ necrotic areas were seen within.

On taking a biopsy, histopathological investigations confirmed the diagnosis of Rhadomyosarcoma. The patient was referred to oncology department and subsequently started on chemo-radiation.

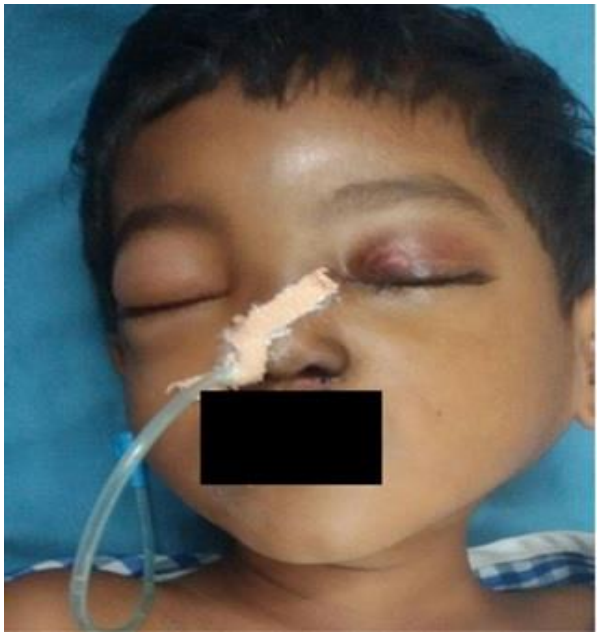

Fig.1- Left Orbital Swelling with peri-orbital oedema

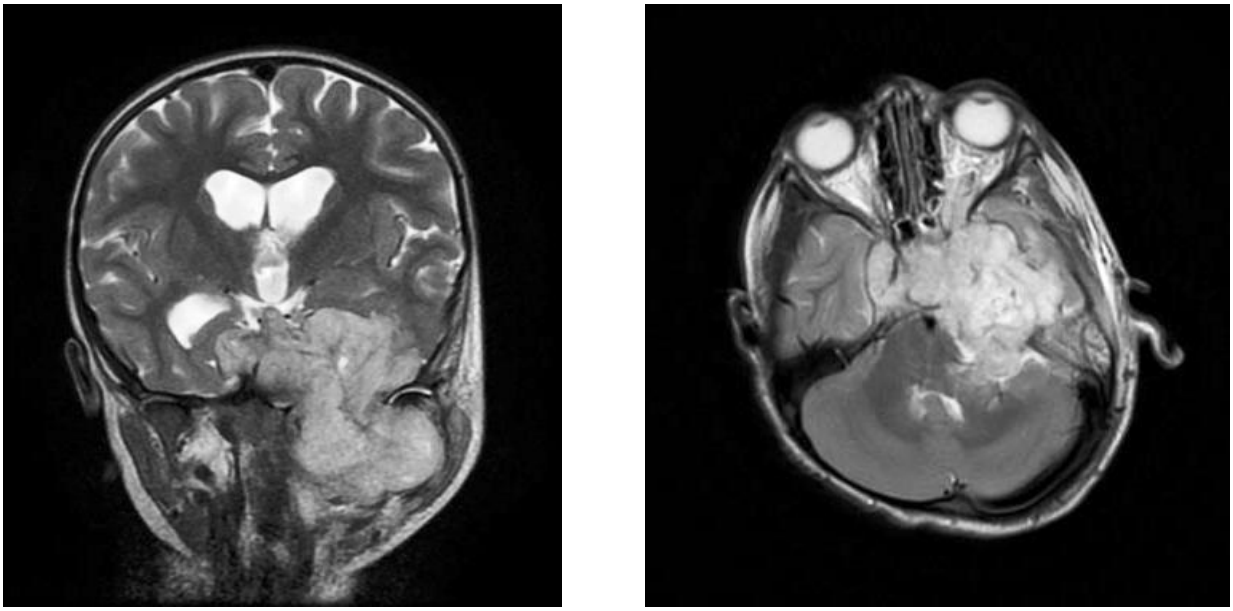

Fig. 2,3: MRI images showing intra- cranial extension of the Rhabdomyosarcoma

DISCUSSION

Rhabdomyosarcoma predominantly occurs in three regions; 1) head and neck, 2) genitourinary tract and retro-peritoneum and 3) extremities. The head and neck tumours are further divided into those that arise in the parameningeal region (50%), orbit (25%) and head and neck superficial, i.e. non para meningeal (25%).[7]

Para-meningeal tumours usually cause nasal, aural or sinus obstruction. These are often associated with cranial bone erosions that can manifest as cranial nerve palsies. Erosion of contiguous bone at the cranial base and intracranial extension may lead to headache, vomiting and systemic hypertension. Nasopharyngeal tumours can cause voice changes, airway obstruction, dysphagia and epistaxis while sinus tumour can be painful in addition to having persistent nasal discharge and occasionally epistaxis. Tumours of the middle ear or mastoid can present as a polypoidal growth from the ear, otitis media, or facial palsy. Laryngeal tumours can present with hoarseness. Regional lymph node metastases to cervical lymph nodes may be present in upto 20% casesdepending on the site. Distant metastasis is primarily to the lungs or the bones.

The further management of the tumour depends on its extent. The various treatment modalities include surgery, chemotherapy and radiotherapy.

Ear Rhabdomyosarcoma may begin in the muscles of Eustachian tube, the middle ear or from the pluripotent mesenchyme. But there is widespread local invasion throughout the petrous bone at diagnosis.[8]

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.[9] Physical examination usually reveals a grey-to purple, fleshy friable polypoidal mass in the ear canal penetrating the tympanum that infiltrates the surrounding soft tissue, hence, causing diffuse swelling in the peri-auricular region. Involvement of the apex of the petrous bone, internal auditory canal and the base of skull may lead to multiple cranial nerve palsies.[10]

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.[11]

Three subtypes have been described i.e., Embryonal (55%), Alveolar (20%), Pleomorphic or undifferentiated (20%) and botryoid variant of embryonal (5%).[12] Embryonal variant has different subtypes. The botryoid variant of ERMS arises in mucosal cavities, such as bladder, vagina, nasopharynx and middle ear. Lesions in extremities are more likely alveolar.

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 upto 67% of middle ear tumours.[8]

MRI is more efficient for evaluation of dural involvement with intracranial extension and to assess the proximity of the tumour to the carotid arteries and jugular veins. The signal intensity of the tumour is minimally hyper intense to muscle in T1-W images and markedly hyper intense to muscle on T2-W sequences. The tumour demonstrates intense homogeneous enhancement after Gadolinium administration.

Surgical excision of the tumour followed by irradiation and chemotherapy is the usual treatment. If there is para-meningeal involvement, as evident by skull base erosion on computerised tomography or pleocytosis with elevated protein and decreased glucose level in cerebro-spinal fluid, a combination of intrathecal chemotherapy and craniospinal irradiation is the treatment of choice.[13] Thus, a multi-modal therapy provides a best chance of survival in these tumours.

CONCLUSION

In children presenting with an ear mass and persistent ear discharge, rhabdomyosarcoma as a differential diagnosis should also be considered. Early diagnosis with referral to a multidisciplinary team will increase the chances of survival. Discharge, hearing loss and aural polyp, although commonly because of chronic otitis media, should prompt urgent investigation and biopsy, particularly if associated with facial palsy, lymphadenopathy or an obvious mass.

CONFLICTS OF INTEREST

No authors have any conflicts of interest or financial ties to disclose.

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