Rhabdomyosarcoma of Middle Ear

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Case report

Abstract: A girl child aged 2 years presented with pain and mass Right ear since 15 days. CT scan and biopsy of mass gave a diagnosis of Parameningeal - Ear Embryonal Rhabdomyosarcoma. The patient improved well after chemo radiation. This case report defines the clinical features and optimal therapy for children with middle ear Rhabdomyosarcoma.

Keywords: IRSG, Middle ear, Prognosis, Rhabdomyosarcoma.

Introduction

Weber first described Rhabdomyosarcoma in 1854 and Stout described morphology in 1946 describing histology. [1] The name is derived from Greek words *rhabdo*, which means rod shape,and*myo*,which means muscle. Rhabdomyosarcoma is the most common soft tissue sarcoma in children and very rare above 45 years. [2] The head and neck is the principal location of Rhabdomyosarcoma in 30-50% of cases. [3] Rhabdomyosarcoma of the middle ear and mastoid is the most common malignant aural neoplasm in the pediatric population and advanced disease is common. Aggressive bone destruction with obliteration of normal landmarks occurs in majority.

Case report

A 2 ½ year girl child presented with complaints of pain right ear15 days, brown colored discharge from right ear 10 days,and gradually increasing swelling around the ear since 7 days. The child has dysphagia and vomitings along, with regurgitation of liquids through the nose. Ear examination showed blood clots in right ear with displacement of ear. Left ear was normal. CT scan was suggestive of lobulated heterogeneous enhancing mass lesion in the right temporal region extending into the nasopharynx with destruction of the entire right temporal bone, and also the vital structures around ear (Figure 1). All blood tests were normal. Biopsy was done from right ear and the mass in front of ear under general anesthesia and sent for Histo-pathological examination. Report was suggestive of embryonal Rhabdomyosarcoma (ERMS) shown figure 2. The child was started on chemotherapy and radiation. The child improved well.

Discussion

Rhabdomyosarcoma is a malignant tumour arising from sites with absent striated muscle i.e. nasal cavity, middle ear and vagina. [2] The tumour arises from primitive muscle cells and can occur anywhere. Ear Rhabdomyosarcoma may begin in the muscles of Eustachian tube, in the proper middle ear or from the pluripotential mesenchyme. But there is widespread local invasion throughout the petrous bone at diagnosis. [3] Common sites of occurrence were shown in table 1.

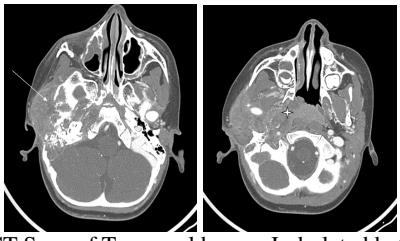

Figure 1: CT Scan of Temporal bone**s:** Lobulated heterogeneous enhancing mass lesion in the right temporal region, extending into the nasopharynx.

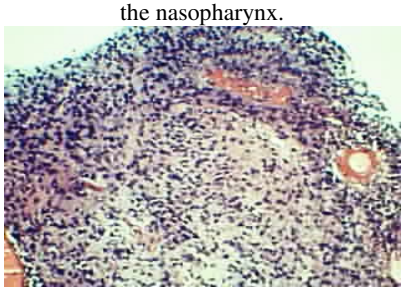

Figure 2: Histopathology of mass in the middle ear showing densely cellular tumor with alternating hypocellular areas consisting of round to spindle shaped primitive looking malignant tumor cells in a myxoid background suggestive of embryonal rhabdomyosarcoma.

Table 1: The most common sites of occurrence

Site of the tumor	Percentage
Head and Neck	28
Extremities	24
Genitourinary tract	18
Trunk	11
Orbit	7
Retro-peritoneum	6

Three subtypes have been described i.e., Embryonal (55%), Alveolar (20%), Pleomorphic or undifferentiated (20%) and botryoid variant of embryonal (5%). 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.

Aetiology

The cause is unknown. Embryonal subtype has different variations like loss of heterozygosity at band 11p15.5; tumor suppressor gene; elevated levels of insulin like growth factor; elevated N-myc level and point mutations in N-ras and K-rasoncogenes.[4] The environmental factors thought to be associated with increased incidence include parental use of marijuana and cocaine, intrauterine exposure toX-rays,[5] and previous exposure to alkylating agents.

Age

Below 15 years we see 87% and 13% cases between 15-21 years. In ages of 2-6 yrs head and neck or genitourinary tract have equal incidence.

History

Symptoms are because of mass in a location. Metastasis leads to blood dyscrasias and bone pains. Para-meningeal (ear,mastoid, nasalcavity, paranasalsinuses, infratemporalfossa, pterygopalatine fossa) Rhabdomyosarcoma presents with upper respiratory symptoms or pain.

Investigations

CBC count may be suggestive of pancytopenia. Liver function tests, renal function tests, urinalysis, blood electrolyte and chemistry are done before chemotherapy. Plain X-ray of primary and chest is mandatory in all cases. CT scan of primary and chest rules out bony erosion and secondaries. MRI delineates mass and invasion. Bone scans, Ultrasonography of abdomen and echocardiography are also done**.** Biopsy of the primary, Cytogenetics, Fluorescent in situ hybridization (FISH), Reverse transcriptase-polymerase chain reaction testing are done along with Bone marrow aspiration.

Classification

Classification is very important because management of Rhabdomyosarcoma involves combination of chemotherapy, surgery, radiotherapy. The treatment has to be done in a center with lot of experience in paediatric Oncology. Patients are categorized according to the risk, related to location, histology and surgical results. The studies performed by the Intergroup Rhabdomyosarcoma Study Group (IRSG) has defined components of multimodality therapy as a multidisciplinary effort and this has led to favorable outcome for paediatric Rhabdomyosarcoma after four clinical trials over 20 years. Definition of protocols and tailoring the components of multimodality therapy has improved survival rates and decreased side effects. [6]

Surgery

Surgery is done majority of times even when chemotherapy is the treatment of choice. Resection with wide margins is the option. Radiation takes care of lymph nodes.

Chemotherapy

A combination of Vincristine, Dactinomycin and Cyclophosphamide are used along with radiation. 5 year survival rate was observed in more than 90% of the cases.

Radiation

Radiation is given for 5-6 weeks after chemotherapy. In post surgical instances location and extent decide the radiation. In parameningeal cases radiation is started at diagnosis. Some cases need whole-brain irradiation in addition to primary therapy.

Ear

Middle ear RMS presents as an invasive, unresectable tumor that metastasizes to regional lymph nodes. Clinical presentation is like chronic otitis media. Parameningeal location has been an unfavorable location by IRSG. Middle ear RMS has a more favourable prognosis than other locations as they are histologically embryonal or botryoid. Improved supportive care and systemic chemotherapy has lead to improved prognosis.

Conclusion

A proper examination and established protocols are necessary in management of pain and discharge from the ear as all cases of pain and discharge from the ear are not the same, especially in children. Management of Rhabdomyosarcoma of ear has undergone lot of changes and the prognosis in Rhabdomyosarcoma of ear is very good. Even stem cell preservation therapy could not yield good results compared with chemotherapy. Future improvements in more intensive chemotherapy, standardization of radiation techniques and better supportive care for this young group of patients may further reduce the morbidity, side effects associated with treatment.

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