Embryonal Rhabdomyosarcoma with Intracranial Extension

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Abstract

A case of embryonal rhabdomyosarcoma of the middle ear presenting with facial paralysis and a polypoidal aural mass who subsequently had convulsions due to intracranial extension is presented for its rarity.

Embryonal Rhabdomyosarcoma of middle ear and mastoid is a disease of rare occurrence. It is an invasive mesenchymal tumour with high malignant potential, occurring predominantly in children. First case from India was reported by Mehra (1960). Deutsch and Felder (1974) could collect only 22 cases including 5 of their own from the world literature. Intracranial extensions were reported at post mortem studies. Karatay S. (1949) observed erosion of tegment and carotid canal. Myere's et. al. (1968) could find only one temporal bone report apart from their one case. A. S. Khurana et. al. (1987) have reported distant metastases to liver and spinal cord also.

Case Report

A 2 years old male child presented with the complaint of inability to close right eye (Fig. 1) and facial deformity on crying and some mass in the right ear since

45 days duration. There was history of removal of mass from right ear 30 days ago elsewhere but that regrown again filling the right ear. E.N.T. examination revealed a redish mass occupying right external auditory canal with some fullness in pre and postauricular areas. Baby had lower motor neuron facial palsy on the right side. There was no lymph node enlargement. Routine haemogram, Urine, test and X-ray chest were normal. X-ray mastoid revealed

Fig. 1 Patient with Right facial palsy.

bone erosion on right side. Excision of the mass was planned but a night prior to surgery baby had generalised convulsions so anticonvulsants were also administered and a C.T. scan of the head performed that revealed a space occupying lesion in the right temporal lobe. (Fig. 2). Surgical strategy was therefore changed. A temporal craniotomy on right side with simultaneous removal of as much of the mass from ear and mastoid as possible was planned, and removal of most of the visible mass was done, in association with the Neurosurgeon's. On temporal craniotomy the tumour was found to be totally extradural and was

extending to petrous and mastoid portion of the temporal bone. As far as aural part is concerned there was no evidence of any tympanic membrane nor ossicles. The entire area was occupied by the tumour mass. Postoperative period was uneventful.

The histopathology report (Fig. 3) confirms the diagnosis of rhabdomyosarcoma of botryoid variety. Patient is now undergoing chemo and radiotherapy combination.

Fig. 2 : CT Scan head showing space occupying lesion in the right temporal lobe.

Discussion

Rhabdomyosarcoma is a highly malignant tumour of mesenchymal origin. The essential characteristics being a rapid local spread beyond the confines of any attempted therapy. It is predominantly seen in childhood, 77% cases appear under the age of 12 years and 43.5% cases occur under the age of 5 years (Barne's and Maxwell 1972). Histopathologically four different varieties Viz. alveolar, pleomorphic, embryonal and botryoid type have been described by

Fig. 3 : Photomicrography showing small to spindle shaped tumour cells with perivascular arrangement. H and E x 16.

Horn and Enterline 1958. They also observed that embryonal and botryoid varieties are seen in paediatric age group. The commonest clinical presentation is

discharge from ear with aural polyp or granulation tissue in the ear. Facial paralysis is also one of the presenting features. II, III, IV, and VI cranial nerves may also be involved. Systemic and lymphatic spreads are well advanced at the time of presentation. Intracranial temporal bone spread has been explained by erosion of trautmann's triangle, with the fungation in the facial canal and involvement of internal auditory meatus have been reported in the literature. Prognosis of the rhabdomyosarcoma of mastoid is gloomy. On an average the survival being 7.2 months since the diagnosis (Jaffe et. al. 1971). Masson and Soule 1965 reported 88% mortality within 5 years and 55 % of this is seen in 1st year. Maxwell et. al. 1978 have however reported a survival even 12 years after the diagnosis. There is a lot of controversy about the treatment modality of choice. According to Hall and Enterline (1958) radical surgery in anatomically feasible situations should be adopted first. Masson and Sule (1965) employed radiotherapy alone or combined with surgery and chemotherapy or both. Raney et. al. (1983) reported complete response in 16 children and partial response in 5 with combined surgery, radiotherapy and chemotherapy. Even at present inspite of multifaculty approach the prognosis is still guarded one.

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