Embryonal Rhabdomyosarcoma of the Mastoid in an Infant

R. K. Jain Reader, Dept. of Otolaryngology, **A. K. Asthana** Reader, Dept Radiotherapy & Radiation Medicine, **M Kumar** Reader, Department of Pathology. **Institute of Medical Sciences, Banaras Hindu University, Varanasi, India.**

Abstract

A case of embryonal rhabdomyosarcoma of the mastoid in a six weeks old infant is reported because of rarity and unusually early onset of the disease.

The embryonal rhabdomyosarcoma is a highly malignant tumour of rhabdoblats, the microscopic pattern of which simulates that of embryonal cells. Embryonal rhabdomyosarcoma of the temporal bone is extremely rare condition.

The following case is reported due to rarity of the condition possibility of its being the youngest reported patient of temporal bone rhabdomyosarcoma and its unusual presentation.

Case Report

S. B. 6 weeks old female infant was admitted (Hospital No. 18703) at University Hospital, Banaras Hindu University, Varanasi, with the complaints of a rapidly increasing swelling in the right temporal region for the past one month, inability to close the right eye, deviation of angle of the mouth to the left and inability to suck the breast for last 15 days.

Mother noticed a small swelling of 1 cm diameter at the right post-auricular region when the child was two weeks old, which was not preceded by trauma. The swelling kept on increasing in size rapidly, for the last 15 days the child had developed inability to close the right eye with deviation of angle of the mouth accompanied with inability to suck the breast.

On examination at admission the child appeared grossly cachectic with severe pallor and weighted

2.5 kg. Systemic examination of respiratory, cardiovascular and abdomen revealed no abnormality. Examination of liver, spleen and lymph nodes was within normal limits. Head circumference was 37 cms. Neurologically the child had right sided seventh (infranuclear type) and bilateral sixth cranial nerve palsies. Rest of the C.N. S. examination was within normal limits.

Local examination showed a large swelling (7 cm x 6 cm) present at the right periauricular region, extending from the petromastoid region posteriorly upto the angle of mandible anteriorly, superiorly to squamotemporal region down to the middle of anterior border of Sternomastoid muscle (Fig. 1). The pinna and posterior wall of the external auditory canal were displaced anteriorly occluding the external auditory meatus. The external auditory canal was filled with pinkish polypoidal mass. The swelling was smooth, lobulated, nontender and non-pulsatile. The consistency was firm at periphery and cystic in the centre. The margins were indistinct, local temperature was raised. The cervical lymph nodes were not enlarged.

Investigations

Haematological : Hb. 4.5 gm percent, Total leucocyte count 12,00/mm³, Differential leucocyte counts p-60, L-35, M-3, E-2. No abnormal cells were present in the peripheral blood smear.

Radiological : Skiagrams of mastoids showed a large soft tissue swelling with the evidence of extensive destruction of squamo mastoid region of the temporal bone and inferior aspect of occipital bone on right side. The mastoid air cells were also destroyed. In addition there was destruction of ramus of mandible on the same side. X-Ray chest was normal.

Histopathology : Histopathological examination revealed spindle shaped cells with tapering

Fig. 1 : The patient with a large swelling in right periauricular region.

bipolar cytoplasmic extensions and round to oval undifferentiated cells with little cytoplasm in a loose myxoid stroma. Occasional cells with eosinophilic cytoplasm and prominent nucleolus were also seen. P.T.A.H. staining showed cross-

striation in occasional cells. Diagnosis of embryonal rhabdomyosarcoma was made on these grounds (Fig. 2).

On the basis of these investigations the child was found unsuitable for any major surgical intervention or chemotherapy. Only palliative radiotherapy was given without any response and the child expired after three weeks.

Discussion

The first detailed description of rhabdomyosarcoma was given by Stout (1946). Rhabdomyosarcoma, although prone to wide and varied distribution in the head and neck, appears to show a predilection for orbit. Masson and Soule (1965) in their study of 88 cases of rhabdomyosarcoma reported 60% of cases either from orbit or from nose or nasopharynx. Temporal bone was involved only in two cases. Horn and Enterline (1958) reported 39 cases of rhabdomyosarcoma out of which only one case was of temporal bone.

Fig. 2 : Micro photograph showing cells in loose stroma. Occasional cell showing rhabdomyoblastic differentiation seen (Arrow) x 500 H & E.

Rhabdomyosarcoma have been classified in four pathological categories by Horn and Enterline (1958) : embryonal, alveolar, pleomorphic and mixed. The majority of the head and neck tumours are of embryonal variety. Rhabdomyosarcoma spreads locally and also

metastasize to distant sites ; both be haematogenous and to a lesser extent by lymphatic spread (Maurer et al. 1977).

Batsakis (1979) for the sake of clinical comparison divided rhabdomyosarcoma of the head and neck in five major groups. Group I constitutes the orbital neoplasm, group II consists of the tumours arising from the soft tissue of temple, face and neck. Intraoral, intranasal and pharyngeal rhabdomyosarcoma make up group III, group IV comprises of those rhabdomyosarcomas taking their origin from or intimately associated with air containing space of skull i.e. paranasal sinuses, middle ear and mastoid, remaining rhabdomyosarcoma of the head and neck are arbitrarily placed in group V.

The embryonal rhabdomyosarcoma is extremely lethal neoplasm. In Dito and Batsakis (1962) study the largest survival was only 23 months after the diagnosis. The average survival was 7.2 months. In Masson and Soule (1965) series 55% of cases died in the first year and nearly 88% died in less than 5 years. Equally poor prognosis was reported by other also. However, Maurer et al. (1977) and Jenkin & Sonley (1980) advocated that treatment of this highly malignant neoplasm needs to be aggressive with integrated approach of utilizing surgery, radiotherapy and intensive chemotherapy. Radical surgery is still advocated in certain sites but in head and neck the anatomical complexity and resultant functional and cosmetic deformity do not justify such a radical approach.

References

1. Batsakis, J.G. (1979) : In *Tumours of the Head and Neck*. Williams Wilkins Baltimore. p. 207.
2. Dito, N.R. and Batsakis, J.G. (1962) : Rhabdomyosarcoma of the head and neck; an appraisal of the biologic behavior in 170 cases. *Archives of Surgery*, 84 : 582.
3. Horn, R.C. Jr. and Enterline, H.T. (1985) : Rhabdomyosarcoma : a clinicopathological study and classification of 39 cases. *Cancer* 11 : 181.
4. Jenkin, R.D. and Sonley, M. (1980) : Soft tissue sarcomas in young, medical treatment advances in perspective. *Cancer* 46 : 621 - 629.
5. Masson, J.K. and Soule, E. H. (1965) : Embryonal rhabdomyosarcoma of the head and neck. Report of eighty-eight cases. *American Journal of Surgery*, 110-585.
6. Maurer, M., Moon, T., Donaldson, M., Farmandez, C., Gehan, E., Denman, H., Hays, S.D., Lawrence, W., Newton, W., Ragab, A., Raney, B., Soule, E. H., Sutow, W. W. and Tefft, M. (1977) : The Intergroup rhabdomyosarcoma study : A preliminary report. *Cancer* 40 : 2015-2026.
7. Stout, A.P. (1946) : rhabdomyosarcoma of skeletal muscles. *Annals of Surgery*, 123 : 447.