Embryonal Rhabdomyosarcoma of Nasopharynx

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Abstract

A rare case of Embryonal Rhabdomyosarcoma in a 5 year male child involving Nasopharynx with intracranial secondaries is reported. The child presented with difficulty in breathing and swallowing with change of the voice and bleeding from nose and mouth off and on. Histopathology of the excised mass proved the diagnosis of RMS & CT scan showed secondaries in the left frontal & temporal to be of brain. In spite of the surgery and chemotherapy, the disease proved to be fatal within 3 months of diagnosis.

A Case of Embryonal Rhabdomyosarcoma of nasopharynx is reported because of its being very rare at this site. Embryonal Rhabdomyosarcoma is a relatively uncommon but highly malignant tumour found in small children. It constitutes 4.8% of all malignant diseases in children under 15 years (Young & Miller 1975). Although it was first described by Soderberg 1933 and since then many cases of RMS involving mastoid or middle ear had been reported but only few cases involving the region of nasopharynx have been described in literature. Pandhi et al (1980) reported a series of 12 cases out of which 2 cases were one each of alveolar and embryonal type of RMS involving the nasopharynx.

Case Report

A 5 year male child was admitted to ENT ward of Rajindra Hospital, Patiala with the complaint of difficulty in breathing and swallowing with change in the voice since 3 months. There was history of bleeding from nose and mouth off and on for the last 2 months. General physical examination showed a poorly nourished and emaciated child. Local examination showed a granular mass in the left nasal cavity. On probing it was friable non-tender and did not bleed on touch. Posterior limit of the mass could not be assessed. Examination of throat showed smooth downward bulge of soft palate on the left side extending into the lateral wall of pharynx behind the posterior pillar of left tonsil. Examination under general anaesthesia confirmed the presence and attachment of granular mass to the postero-superior wall of nasopharynx and filling the left choana as well.

The mass was removed per orally after retracting the palate. It measured 7.5 cm x 4 cm.

Investigations

X-ray nasopharynx revealed complete opacification of left side of nasopharynx and nasal cavity.

Biopsy of the mass showed cells with pleomorphic hyperchromatic nuclei lying in loose myxoid background and Focal areas of eosinophilic intercellular hyaline material suggestive of embryonal rhabdomyosarcoma (Fig. 1).

C.T. Scan showed an area of increased attenuation in left frontal lobe extending across the midline.

Fig 1 Photomicrograph of R.M.S. showing pleomorphic hyperchromatic nuclei in loose myxoid background with focal areas of eosinophilic intercellular hyaline material H & E - 400 x

Fig. 2. CT— Scan Head showing secondaries in frontal and temporal lobe of brain.

A rim of increased density was seen with central area of necrosis with surrounding area of perifocal oedema. Another similar lesion was also seen in left temporal lobe with obliteration of left temporal horn, confirming secondaries brain (Fig. II).

Post-operatively the child was put on chemotherapy combination of cisplatin, vincristine and endoxan in appropriate doses. Follow up of the case was done but ultimately the child died within a period of 3 months.

Discussion

R.M.S. is the most common soft tissue tumour

occurring particularly in children under 15 years of age. There are four variants of Rhabdomyosarcoma such as Embryonal, Alveolar, Pleomorphic and Botryoid variety (Horn & Enterline 1958). In India the first case of R.M.S. of middle ear was reported by Mehra (1960). Embryonal variety of R.M.S. arises from unsegmented and undifferentiated mesoderm. Its frequency in order of predilection of various sites in Head & Neck is orbit, nasopharynx, cheek, external ear, maxillary antrum and retrotonsillar area (Donaldson et al 1973). Tumour is most commonly seen in infants and children (Shenawy et al 1980). The disease has also been reported to occur in larynx and hypopharynx (Frugoni & Ferlita 1976). The tissue of origin of R.M.S. is derived from totipotent mesenchymal cells or from muscles and in the present case may be palatal muscles around the Eustachian tube. Although characteristic feature is embryonal myoblasts, the diagnosis of R.M.S. depends upon showing myxoid area in different mesenchymal blastoma of elongated cells just under the squamous epithelium. Spread of R.M.S. to regional lymphnodes occur in 3% cases involving H & N region (Maurer et al 1977), although spread to bones, other organs and intracranial spread rarely has been reported in literature. In our case beside local spread, secondaries were seen in left frontal and left temporal lobe of brain. Treatment of R.M.S. is still an enigma and even many combinations have offered no hope of cure.

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