"Juvenile Alveolar Rhabdomyosarcoma of Mandible"

Amresh K. Saxena, Asst. Professor

Ajay Singhal, Junior Resident

Deptt. of ENT, L.L. R.M. Medical College, Meerut (U.P.)

Abstract :

Rhabdomyomatous tumours are the malignant neoplasms of complex composition and derivation. Three varieties of Rhabdomyosarcoma are described. These are -

1. Pleomorphic 2. Embryonal 3. Alveolar

Juvenile Alveolar Rhabdomyosarcoma usually affects extremities and less commonly trunk, orbit, soft tissues of the face (Cheek), Neck and oral cavity including the tongue and palate. A case of Juvenile Alveolar Rhabdomyosarcoma of Mandible is being reported. Relevant literature is reviewed.

INTRODUCTION

Malignant tumours of rhabdomyomatous composition and derivation constitute a complex group of neoplasms of uncertain histogenesis. Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma of children under the age of 15 years and more than one third of cases arise in head and neck region.

There are three major categories of rhabdomyosarcoma- Pleomorphic, Embryonal and Alveolar. Juvenile Alveolar Rhabdomyosarcoma is the 2nd most common type of rhabdomyosarcoma, first being the embryonal type. It accounts for approximately 20% of all rhabdomyosarcomas. It was first described by Riopelle and Theriault (1956), later by Enterline and Horn (1958) and comprehensively reviewed by Enzinger and Shivaki (1969). Rhabdomyosarcoma of maxillary antrum has been reported by Saxena and others (1989). Gupta et al (1997) reported a case of Rhabdomyosarcoma of parotid gland in a 10 years old female child.

Juvenile Alveolar Rhabdomyosarcoma arises mainly in the extremities and less commonly trunk, orbit, soft tissues of the face (Cheek),

Neck and oral cavity including the tongue and palate.

Because of its rarity, a case of Juvenile Alveolar Rhabdomyosarcoma of Mandible is being reported.

Case Report

A 7 years old male child was admitted to the department of ENT, L.L.R.M. Medical College, Meerut with chief complaints of swelling over right cheek for last 3 months and a mass filling whole of the oral cavity for last 2 months. The swelling was gradually increasing in size since last 3 months. There was no history of fever and trauma. General physical and systemic examinations were found to be within normal limits.

On local examination marked swelling of the right cheek, 8cm x 7cm in size was present, extending from lobule of the ear upto angle of mandible. On palpation, swelling was firm in consistency, nontender and fixed to deeper structures. A mass was found, filling whole of the oral cavity associated with foul smelling discharge. (Fig. 1) Haemogram and Urinalysis

Fig. 1 Photograph of the patient at the time of presentation showing extension of swelling over right cheek and presence of the mass in the oral cavity.

Fig. 2 X-Ray face Antero-Posterior View showing soft tissue shadow around right ramus and angle of mandible along with absorption of bone.

were within normal limits. X-Rays of the face - Antero-Posterior view and Lateral views were taken, which showed, soft tissue shadow around right ramus and angle of mandible along with absorption of bone (Fig. 2).

Debulking of the mass present in oral cavity was done after doing a preliminary tracheostomy as there was no space for endotracheal intubation. The mass was found arising from mandible and the mandible was found to be totally destroyed on right side.

The mass was sent for histopathological examination which revealed round to oval cells of moderate size and apparently unattached to each other, contained within small or large irregular spaces. The nuclei being situated at the opposite bulbous end of the cell causing the centre of each alveolous to appear as a cavity in the centre of which were found few or many free floating cells, consistent with Juvenile Alveolar Rhabdomyosarcoma (Fig. 3).

After confirmation of diagnosis, combination of Chemotherapy followed by Radiotherapy was planned. Chemotherapy comprised of three cycles of cyclophosphamide 500mg, vincristine

Fig. 3 Photomicrograph (H & E stain x 40) showing feature of Alveolar Rhabdomyosarcoma.

0.2mg and adriamycin 20mg given at an interval of 3 weeks. This was followed by external radiation by cobalt 60. Total dose of 6000 rads were given over a period of 6 weeks. This treatment produced marked reduction in the size of the tumour and swelling. The patient was kept in regular follow up.

Nearly five months after the completion of Radiotherapy, patient presented with a small swelling over skull which was gradually in-

creasing in size. X-Ray skull Antero-Posterior view and Lateral views showed radioopaque outline on right side of skull along with defined lytic areas in the right frontal region. FNAC of the swelling revealed it to be a metastatic lesion. Patient was again given Chemotherapy at 3 weekly interval. This time cisplatin 40mg. was also given in addition to previous combination of cyclophosphamide, vincristine and adriamycin. Total 5 cycles were given. Patient is under follow up for last 1½ years with a small swelling over right side skull.

Discussion

Rhabdomyosarcoma has a fairly uniform age incidence though it occurs predominantly in infants and children and somewhat less frequently in adolescents and young adult. It is rare in patients older than 40 years. The median age of patient varies somewhat according to histological type. Embryonal rhabdomyosarcoma including those of botryoid type are found mainly in the infants and young children. Alveolar rhabdomyosarcomas are observed mainly in children and adolescents and pleomorphic rhabdomyosarcomas occur mainly in older persons.

Males are more commonly affected than females in the ratio of approximately 1.3:1 but the male preponderance is less pronounced among cases occurring in adolescents and young

adults as well as in patients with rhabdomyosarcoma of the alveolar type.

The malignant nature of the lesion is shown both by histological appearance and by the presence of pulmonary metastases (Williams and Sloman 1956).

Factors affecting the prognosis of rhabdomyosarcoma in children include age, primary site, size, histological type and type of treatment. (Proops et. al. 1984). Children with tumours of embryonal histology had better prognosis than those whose tumours were of alveolar type. (Sutow et al. 1970, Grossfeld et. al 1977). Hays et. al (1983) reported that death rate of the alveolar type was 44% and of the embryonal type 13.5%.

Alveolar rhabdomyosarcoma is ordinarily a very malignant neoplasm. Growth is rapid and death with widespread blood born and lymphatic metastases within two or three years is usual (Enzinger and Shiraki 1969, Howorth et. al. 1980).

The usual accepted protocol of treatment for this aggressive neoplasm is surgery followed by Chemotherapy and Radiotherapy but complete excision was not possible because of head and neck anatomical complexity. Radiotherapy between 5000-6000 rads in 5-6 weeks is recommended to primary tumour by Intergroup rhabdomyosarcoma study report. (Maurer et. al. 1977).

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