Melanotic Neuroectodermal Tumour of Maxilla In Infancy

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Abstract

Melanotic neuroectodermal tumour of infancy is an uncommon pigmented neural crest derivative. It is usually found affecting children aged one year or less. We present a case report of a four month old male infant with this tumour arising from the right maxilla showing extensive involvement of the surrounding bony and soft tissue structures.

Melanotic neuroectodermal tumour of infancy (M.N.T.I), is a rare dysembryogenetic neoplasm which affects children less than one year of age. Majority of these tumours present in the first three months of life (R.M. Irving et al., 1993). Though considered to be benign, 3.2% of these show a malignant potential (Joseph C. George et al., 1995). Described first by Krompecher in the year 1918, the tumour has 23 different synonyms. Melanotic epithelial odontoma, Retinal analage tumour, Melanotic prognoma, Pigmented admantinoma, Melanotic ameloblastoma, Melanocytoma to name a few. Earlier thought to be odontogenic in origin, the tumour is now found to be a neural crest derivative on the basis of

histopathological, biochemical and immunocytochemical analysis. Raised levels of vanillylmandilic acid (V.M.A.) in a 24 hour urine sample and an increased serum α -fetoprotein have been reported with these tumours.

Case Report

A four month old male child was brought to the E.N.T. outpatient department of Smt. Sucheta Kriplani Hospital, Lady Hardinge Medical College, New Delhi. He had presented with one month history of a progressively increasing mass arising from the right half of the hard palate and alveolus. It was smooth, globular, non-cystic and was seen protruding into the oral cavity (Fig. 1). The child had stridor occasionally and was tracheostomized for the same. Pale pink in colour the tumour showed a leash of blood vessels adding a bluish hue locally at few spots. It was hard,

Fig. 1 Showing the tumour in the oral cavity.

non-tender, non-fluctuant, non-pulsatile and did not bleed on touch. The covering mucosa was normal. The other E.N.T. examination revealed a narrowed left sided nasal cavity due to the compressive effect of the mass. The right side malar process was more prominent. The eye of the same side showed mild proptosis. Ocular movements were normal. All cranial nerves were intact.

The routine hematological profile of the child was within the normal range. X-ray PNS (Water's view) showed obliteration of the maxillary sinus on the right side with bony expansion of the alveolus and the

maxilla. The C.E.C.T. of the paranasal sinuses and the brain revealed a soft tissue density mass (35-50HU) on the right side of the hard palate and the alveolar margin of the same side. It showed tumour extensions into the soft tissues of the cheek with bony erosion of the maxilla, ethmoid sinuses, nasal cavity, infratemporal fossa and pterygopalatine fossa of the right side. There was anterior displacement of the right orbit with erosion of the lamina papyracea (Fig. 2). The punch biopsy report finally clinched the diagnosis of M.N.T.I. The 24 hour urinary V.M.A. level was also found to be raised which further supported the diagnosis.

Child was taken up for surgery. Total maxillectomy with clearance of the disease from ethmoids, infratemporal fossa and pterygopalatine fossa was carried out. Post-op. recovery was uneventful (Fig. 3). The tumour mass excised showed areas of slate grey

Fig. 2. C.T. Scan showing tumour in the Maxilla

discoloration. The cut section had an impacted deciduous tooth. The microscopic sections stained with H and E showed fibrous connective tissue stroma with large number of cuboidal epithelial cells laden with melanin. The tubules at places were found to be filled with neuroblast like cells. Malignant cells were not seen (Fig. 4).

Discussion

93% of the M.N.T.I's originate in the head and neck area. The common sites are anterior alveolar ridge of the maxilla (68.8%), skull (10.8%), mandible (5.8%), and brain (4.3%) (Ahmed E Saggam et al., 1998). The

Fig. 3. Post operative photograph of the patient.

other sites of involvement are the epididymis, thigh, mediastinum, ovary and shoulder (Cutler et al., 1981). The neoplasm affects both the sexes in equal proportions.

M.N.T.I's are usually non-capsulated. In a few cases they may possess a fibrous capsule. Shrouded with controversies pertaining to its origin, the most widely accepted theory is that the neoplasm is a neural crest derivative. It is supported by the fact that it secretes

Fig. 4 Histopathology of lesion showing cuboidal epithelial cells laden with melanin. Tubules at places are found to be filled with Neuroblast like cells (H&E stain 40x).

large amounts of V.M.A. Tumour has to differentiated from pyogenic granuloma, hemangioma, lymphangioma, histiocytosis X, fibrosarcoma, chondrosarcoma, Rhabdomyosarcoma etc.

The treatment modalities of these tumours range from local excision with a long term follow-up, excision of the tumour mass with no margins or with a 5mm clinically normal surrounding margin. 10-15% show a local recurrence, while 3-4% may show a malignant potential. Therefore in cases of rapid repeated recurrences, possibility of malignant potential should be kept in mind.

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