CASE REPORT

Melanotic Neuro-ecto-dermal tumor of infancy- 2 cases of a rare tumor

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Abstract

Two cases of melanotic neuro-ecto-dermal tumor of infancy (MNTI), involving maxilla presented at 4 and 6 month of age with increasing intra-oral mass interfering with feeding and bleeding from that mass. Patients were pale and had firm to hard non tender immobile swelling involving maxillary alveolus. Biopsy and immuno-histochemistry were suggestive of MNTI and surgical excision with 5mm safe margins was carried out. Both infants had uneventful recovery and were started on oral feed and were discharged.

Keywords: Maxillary mass, Melanotic neuro-ecto-dermal tumor of infancy

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Introduction:

Melanotic neuro ectodermal tumor of infancy was first described by Krompecher in 1918.[1] It is a melanin containing tumor derived from neural crest cells,[2,3] also called as melanoic progonoma, melano-ameloblastoma or retinalanlage tumor. It frequently arises from the maxilla. Radical surgery is the recommended treatment. Radical surgery have also been reported in some cases.[2] We are documenting two cases of MNTI, managed in our institute.

Case 1:

A 4 month boy weighing 5 kg, rapidly increasing intraoral swelling of 3 months duration, causing difficulty in taking feed and bleeding for last few days. On examination, child appeared pale with N/G tube and tracheostomy with large tumor about 7 cm in maximum diameter occupying right maxilla, protruding in to whole oral cavity with ulcerated overlying mucosa.

Patient had Hb of 8.9gm/dl and WBC count of 4.3 x 109 /L. MRI done previously revealed an inflammatory mass 2.8x2.2 cm involving alveolar process of maxilla and extending into right maxillary sinus. A fresh MRI at one month's interval showed a significant increase in the size of tumor, 5.5x5.7 cm in maximum diameter on gingivo-buccal aspect of right cheek. Posteriorly extending up to the angle of mandible with minimal encroachment of infra temporal fossa with no intra-ocular or intra-cranial extensions. Metastatic workup was also negative.

A Fergusson's incision under general anesthesia was made. Tumor was completely engulfing alveolar process, palate, maxillary sinus and right zygomatic complex. It was excised with safe margins of 0.5mm leaving infraorbital rim completely intact. Histology confirmed tumor free margin. Patient had uneventful recovery, was allowed N/G feeding 6 hrs post-operatively and later shift to oral feed. Trial for tracheostomy tube removal was done twice but was not possible due to apnec episodes so was discharged with tracheostomy on 15th post-operative day.

Case 2:

A 6 months male child weighing 7 kg, admitted through emergency department with gradually increasing right cheek swelling since birth, and bleeding from that swelling for a few days. On examination, a pale child with 3x3cm purplish

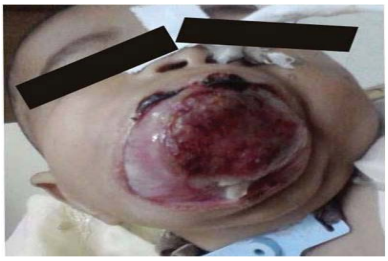
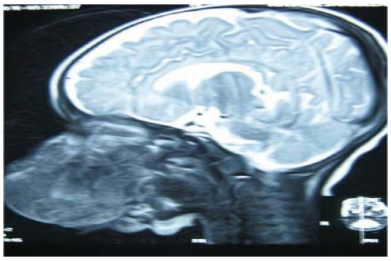


Figure 1:

Figure 2:



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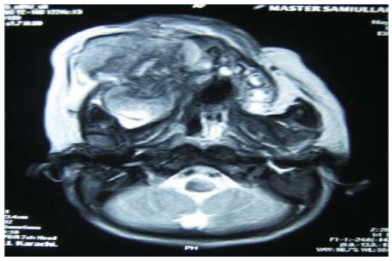

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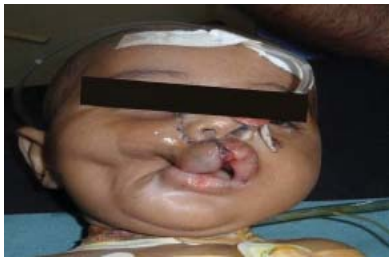

Figure 5:

swelling with eroded surface inside mouth on right upper alveolus, hard in consistency involving hard palate. Initial investigations revealed Hb 6.5 gm/dl, TLC 15400/cmm, Platelets 454000/cmm. Surgical excision of mass was done after optimizing the patient, under general anesthesia. It was 3x4 cm blackish firm, vascular mass arising from right upper alveolar margin containing calcification and teeth, extending into maxillary sinus.

Histopathology revealed fragments of a neoplastic lesion showing dual population of cells, Immuno-histochemistry revealed HMB-45 positive in epithelial cells, synaptophysin, chromogranin, CD 99, CD 56 positive in rounded cells making the features consistent with MNTI.

Post-operatively patient remained well, allowed orally on 3rd day and was discharged on 5th post-operative day. Metastatic workup was negative. At 1 month follow up patient was doing well no local recurrence.

Discussion:

Melanotic neuro-ecto-dermal tumor of infancy has been described as neuro-ecto-dermal in origin on basis of ultrastructural, immunocytochemical and electron microscopic studies.[1] 90% of the tumor involves head and neck region, maxilla being the most common site (68.8%)[4] Our both patients presented with maxillary alveolar mass. Age ranges between 3-6 months. MNTI is associated with an increase in urinary excretion of free form of vanille lmandelic acid (VMA),[3] but it was within limit in our cases.

Histologically there is biphasic proliferation of small neuroblastic cells and large melanin pigmented epithelioid cells, Immuno-histochemical markers like HMB45, synaptophysin and CD56 are predominantly reactive for Melanotic neuro-ecto-dermal tumor of infancy in literature. In frequent to rare positive cases are reported for S100, CD99 and chromogranin.[6] Our first case was positive for neuron specific enolase, synaptophysin, cytokeratin while second case was positive for HMB45, synaptophysin, chromogranin, CD56 and CD99.

Local recurrence is reported to be 15% and malignant transformation 6.6%. Metastasis although very rare can occur in liver, bone, lymphnodes, spinal cord or brain is mentioned in literature.[3,6] Recommended chemotherapy for malignant MNTI includes alternate cycles of cyclophosphamide and doxorubicin every 21 days for four months.[4]

Melanotic neuroectodermal tumor of infancy is a rare, benign but locally aggressive tumor, usually diagnosed clinically due to the classical physical findings and characteristic radiological features. It is important to investigate and diagnose histologically so as to get a correct diagnosis and early radical resection of the tumor with clear margins can be performed

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Role and contribution of authors:

Kanwal Zia, collected the data, referecnes and wrote the article.

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