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RESEARCH ARTICLE

MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY IN 4 MONTHS OLD GIRL, A RARE CASE REPORT

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Abstract

Melanotic neuroectodermal tumour of infancy (MNTI) is an uncommon pigmented neoplasm of neural crest origin. A 4 months old female child presented with swelling at the upper gum , size measuring 2.5cm x 2.5 cm x2.0 cm. . USG and MRI suggestive of low grade lytic lesion- ? Congenital granular cell myeloblastoma/ ? Other lytic bony lesion /?congenital epulis of newborn. Microscopic examination showed dual population of cells , small round blue cells with hyperchromatic nuclei and scant cytoplasm in nodular arrangement surrounded by brown pigment containing epithelial cells and dense fibrosis with areas of ossification in the background. IHC showed positivity for HMB-45,NSE and synaptophysin .The final diagnosis was made as Melanotic neuroectodermal tumour of infancy (MNTI).

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Introduction:-

Melanotic neuroectodermal tumour of infancy (MNTI) is an uncommon pigmented neoplasm of neural crest origin. The term ‘Melanotic Neuroectodermal Tumour Of Infancy’ was introduced for the first time in 1966.(1) It was described for the first time in 1918 by Krompecher, and since the origin was unknown, it was described as congenital melanocarcinoma. Until 1966, this tumor was characterized by variable names (e.g., retinal anlage tumor, pigmented congenital epulis, melanotic progonoma, pigmented teratoma, atypical melanoblastoma, etc.). MNTI has a total of 500 cases reported since 1918, and it can cause significant morbidity.(3)

It is a rare tumor. Maxilla(62%) is the most common site. It also occurs in skull (15%) mandible(8%), long bones, epididymis, mediastinum ,soft tissue of extrmities ,testis and ovaries.(3) The patients are typically young infants less than 1 year old, with a mean age of approximately 7 months and a range of 3 months to 8 years. ,local recurrence has been observed in 10 to 15% of cases. Although usually benign, this tumor can grow rapidly and infiltrate adjacent bone, soft tissue, and the orbit but some cases with aggressive local behavior and a few resulted in distant metastases (3%). A systematic review of jaw lesions shows that the most common metastatic site is the orbit(3). Local recurrence has been observed in 10 to 15% of cases.Age at diagnosis is an important prognostic indicator, because younger age correlated with a higher recurrence rate(7).Borello and Gorlin found that the tumor produces vanillylmandelic acid (VMA), which is produced by other types of tumors that arise from the neural crest.[4]

The highest prevalence is in the United States, followed by India. There is a slightly higher predominance in males.(5) In older age cohorts >3 years of age, females have a higher prevalence.[6]

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Case Report

A 4 months old female child presented with swelling at the upper gum since the age of 2 months, gradually increasing in size and causing difficulty in mouth closing.All blood reports including CBC ,LFT and blood sugar were with in normal limits. USG and MRI were suggestive of low grade lytic lesion- ? Congenital granular cell myeloblastoma/ ? Other lytic bony lesion /?congenital epulis of newborn. Chest x- ray was normal. On clinical examination the growth was relatively hard with variable consistancy arising from the central and left side of upper maxillary alveolar arch containing two teeth. Excision of the growth was done and the specimen was sent for histopathological examination.

On gross examination the specimen consist of single greyish firm to hard nodular mass measuring 2.5cm x 2.5 cm x2.0 cm with one attached and one detatched tooth.Cut surface was greyish with blackish dot,like areas and bony area.

Multiple sections were given from firm and bony areas.Sections from bony areas were kept for decalcification . Microscopic examination of all the sections showed dual population of cells ,small round blue cells with hyperchromatic nuclei and scant cytoplasm in nodular arrangement surrounded by brown pigment containing epithelial cells. The background consist s of dense fibrosis with areas of ossification. Histomorphology was suggestive of Melanotic Neuroectodermal Tumour Of Infancy.However this is a rare tumor ,so immunohistochemistry for HMB-45, NSE,Synaptophysin, Chromogranin was done for final confirmation. In the biphasic tumor small round cell component showed positivity for synaptophysin and NSE while epithelial cells showed positivity for HMB-45.Chromogranin was negative.Thus IHC results were in favour of histomorphological diagnosis of Melanotic neuroectodermal tumour of infancy (MNTI).

Discussion:-

The key features of MNTI used to distinguish it from other entities are the clinical presentation, biphasic population of epithelial melanogenic cells and primitive neurogenic cells, and its characteristic immunohistochemical findings.

1. Neuroblastoma:, It shows rosette formation , which is absent in MNTI. Additionally, the large pigmented epithelial cells that are present in MNTI are not found in the neuroblastoma, so it's negative for cytokeratin and HMB-45.
2. Ewing sarcoma: compared to MNTI, it lacks the biphasic appearance of the MNTI. It demonstrates characteristic diffuse strong membranous immunostaining for CD99 .
3. Alveolar rhabdomyosarcoma: Considering its muscular differentiation, this tumor is positive for desmin and myogenin. Necrosis is also present in this tumor, which is rarely present in MNTI.
4. Malignant melanoma :it would be negative for cytokeratins but positive for melanoma markers .

During the evaluation of a pigmented mass in the head and neck, the differential diagnosis list should also include lymphomas and clear cell sarcoma of soft tissue.[6] The age of presentation of MNTI may overlap with congenital granular cell tumor of the newborn (congenital epulis), but the characteristic histology would help rule this entity out. Odontogenic tumors of childhood may be considered clinically due to the location and presentation, but they rarely occur before 6 years of age.

Figure no. 1:- (A) Nodular mass measuring 2.5cm x 2.5 cm x2.0 cm with one attached. (B)cut surface was greyish with blackish dot,like areas and bony area.

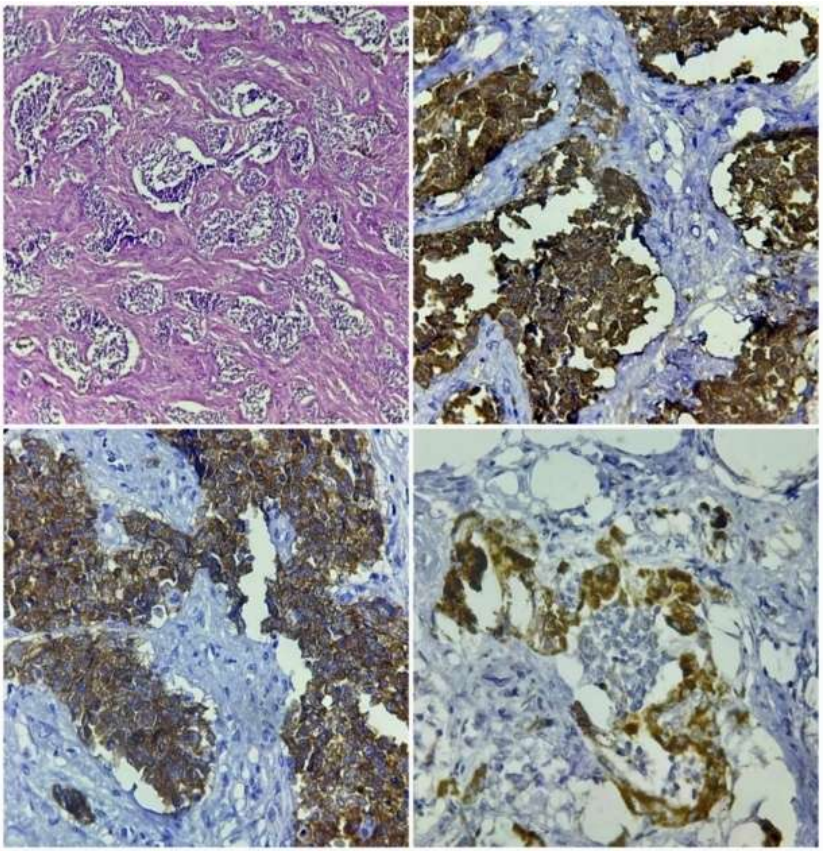

Figure no .2:- Microscopy shows dual population of cells , small round blue cells with hyperchromatic nuclei and scant cytoplasm in nodular arrangement surrounded by brown pigment containing epithelial cells, fibrosis and ossification(A;H& E,100x) positive staining for NSE (B;400x), positive staining for synaptophysin(C;400x), positive staining for HMB45(D;400x).

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