3. Discussion

MNETI is a rare neoplasm of infancy. It was first described by Krompecher in 1918 as congenital melanocarcinoma.[2] This tumour is known by various other names like retinal analage tumour, melanotic ameloblastoma and melanotic prognoma because the histogenesis of this tumour was not clearly known previously and was thought to arise either from the odontogenic epithelium or from developing retinal epithelial cells.[3] Borelleo et al. thought it to be a tumour of neural crest origin based on elevated vanillylmandellic acid (VMA) levels and named this tumour as melanotic neuroectodermal tumour of infancy which is the current accepted histogenesis and preferred name of this tumour.[4] MNETI is a tumour of infantcy with 95% of cases presenting within 1st year of life with a female predominance.[5] In the present study, both cases presented in first 3 months of age and one was male and other was female child. Clinically the child presents with rapidly growing non ulcerated mass. Head and neck is the commonest region with 70% of the cases occuring in the maxilla. Other less commonly involved sites are mandible, skull, neurocranial dura or brain, epididymis, skin, mediastinum and uterus.[5,6] Temporal bone involvement is very rare with only few case reports.[7,8] In a review of seven cases of MNETI, Johnson et al. found one case involving the temporal bone.[9] In the present study, one child had right maxillary involvement and the other child had left temporal bone involvement. VMA levels may be elevated or within normal limits and in our both cases it was within normal limit. On radiological examination, CT scan shows well demarcated hyperdense lesion and magnetic resonance imaging (MRI) shows hyperintense images on T1 and hypodense images on T2.[10] Cytological examination shows scanty to moderately cellular smears with dual population of larger epithelial cells and smaller neuroblastic cells. In the present study, the smears were cellular and 2nd case showed rosette formation as described in other case reports having temporal bone involvement.[8,9] Histopathological examination shows alveolar pattern of the tumour having large epithelial pigmented cells in the periphery and small neuroblastic cells in the center.[1] Immunohistochemical examination shows positivity for epithelial (cytokeratin CK) and melanocytic (HMB-45) markers in the large epithelial cells. Neural markers (neuron specific enolase, NSE) are positive in the large epithelial and small neuroblastic cells both. S-100 is negative. Neurosecretory granules and melanosomes are seen on electron microscopic examination in small and large cells respectively. MNETI has benign clinical course and complete surgical excision is the treatment of choice. Recurrence can be seen in 10-15% of the cases and metastasis in 7% of cases.[6,10] The differentitial diagnosis includes other malignant small round cell tumours like Ewings sarcoma/primitive neuroectodermal tumour (PNET), alveolar rhabdomyosarcoma and metastatic neuroblastoma. Characteristic dual population of cells and immunohistochemical positivity for epithelial, melanotic and neural markers differentiate MNETI from other small round cell tumours.

In conclusion, MNETI should be considered in infants presenting with mass in maxilla or other head and neck regions. FNAC is a rapid, inexpensive and relatively less invasive technique to diagnose MNETI, however histopathological & immunohistochemical examination is essential to differentiate it from other malignant small round cell tumours of infancy for proper management.

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