CASE REPORT

Melanotic Neuroectodermal Tumor of Infancy

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Abstract

Two baby's, one was 6 months old and another was 2 months old presented with a rapidly progressing melanotic neuroectodermal tumor of infancy (MNTI) in the maxilla. The tumor extended from the midline to the retromolar area of the left maxilla. It was confirmed as a melanotic neuroectodermal tumor through, radiology, cytology and histopathological examinations. The encapsulated tumor was excised .No recurrence was seen in the initial follow up nearly one year.

Introduction

Melanotic neuroectodermal tumor of infancy (MNTI) is a relatively uncommon osteolytic -pigmented neoplasm that primarily affects the jaw of newborn infants.The lesions have had an interesting history since its initial description by Krompecher in 1918 as a congenital melanocarcinoma and approximately 200 cases have been published upto 2001.[1] Borello and gorlin suggested the term Melanotic Neuroectodermal Tumor of Infancy (MNTI).[2] Its a rare variety; locally aggressive and rarely metastasizes. In this case report, we described two rare cases of MNTI as an aggressive tumor in the Maxilla of a 6 months and 2 months old infant that was excised and showed no recurrence.

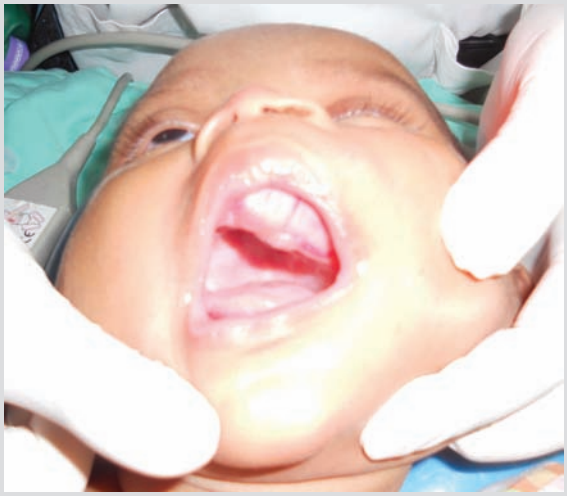


Figure 1/A : Pre-op

Case 1

A 6-months old, female baby got admitted on 14/01/2012 in Oral and Maxillofacial Surgery Department of Apollo Hospitals Dhaka with the complaints of a rapidly progressing swelling on the left side of the face almost since birth and difficulty in feeding and sucking. According to the statement of her mother, a nodular intra-oral swelling was first noticed after 4 weeks of birth. She was then seen by local doctor in Rangpur but the swelling was increasing in size. She was then referred to Dhaka and admitted in Apollo Hospitals Dhaka for proper treatment.

Figure 1/A : Pre-op

1. Senior consultant, Oral & Maxillofacial Surgery, Apollo Hospitals Dhaka 2. Registrar, Oral & Maxillofacial Surgery Apollo Hospitals Dhaka, 3. Consultant Histopathology, Apollo Hospitals Dhaka

Past history of mother and baby nothing contributory. The pregnancy and delivery were normal, there was no history of medication during pregnancy and growth of the baby was normal in relation to her age.

On examination, the firm, non-tender, non-fluctuant swelling extended from the midline of the maxilla to the left retromolar area measuring 6X4 sq cm in diameter. The overlying mucosa and skin were intact, not ulcerated and bluish in color. No cervical palpable lymph nodes were found.

Fine needle aspiration cytology (FNAC) was done. A few drops dark of pigment was aspirated and sent for cytology. Microscopically, the smear showed small oval cells in clusters as well as singly where some cells contained melanin pigmentation.The cells had hyperchromatic nucleus with scanty cytoplasm.The diagnosis was made as melanotic neuroectodermal tumor of infancy (MNTI).

CT scan of the facial bone was done which showed a large bony growth and well defined margin with the involvement of the maxillary sinus.

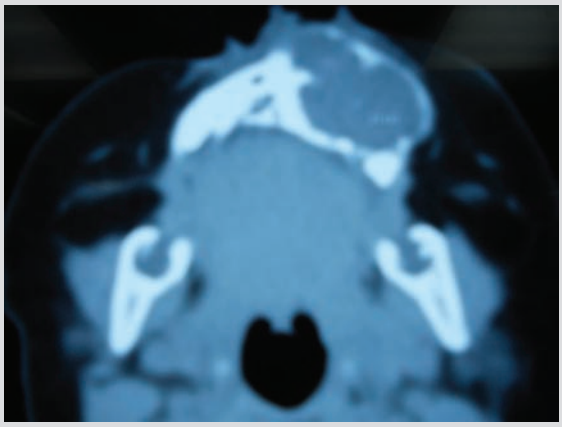


Figure 1/B : CT Scan of Maxillofacial region

Other routine examinations were done. The haemoglobin level was 10.0 gm/dl. Operation was done on 15/01/2012. All preoperative precaution were taken. After proper scrubbing and painting drapping was done. Intra-oral procedure was done. 2% lignocaine with adrenaline was given on the operating site. Intraoral sulcus incision was given, periosteal flap and mucosa were elevated from the lesion of the left sided maxilla. After proper exposure the lesion was excised intact with its lining. All bleeding points were secured. Wound was closed by layer with 4-0 vicryl after placing a small drain. The patient was discharged on 18/01/2012 with satisfactory post operative condition.

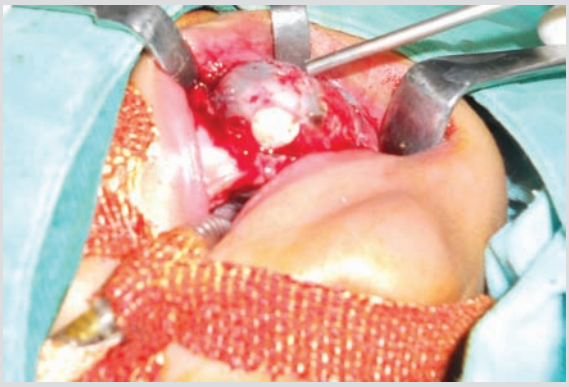


Figure 2/A : Per operative

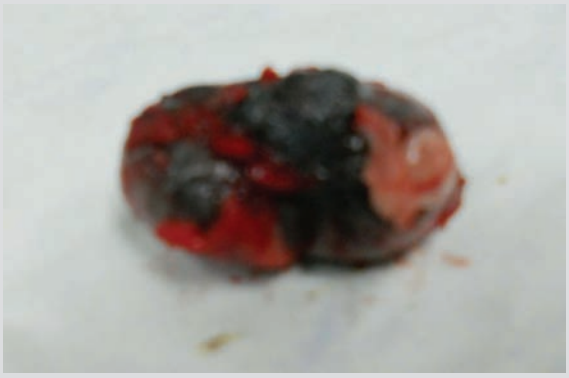
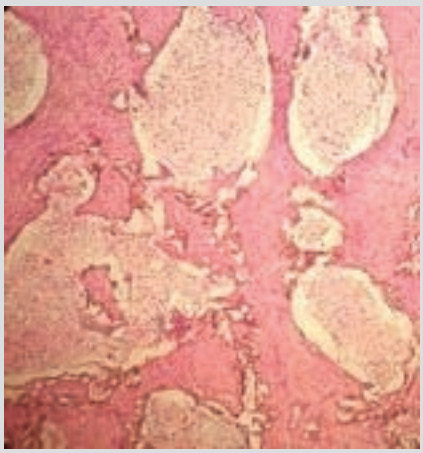


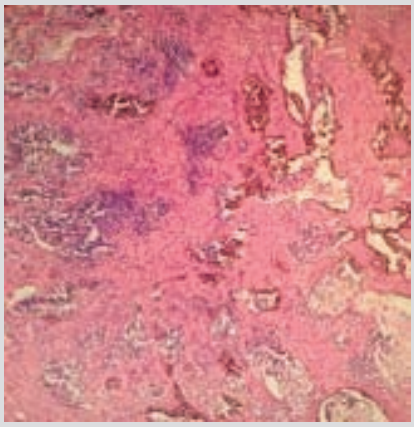
Figure 3/B : Excised tumor

The excised specimen was sent for histopathological examination. The histopathological report revealed round, small to intermediate sized cells having scanty

cytoplasm, arranged in nests and separated by fibrous tissue. Melanin pigment-containing cells were also found. Finally it was diagnosed as a melanotic neuroectodermal tumor of infancy.



Histopathological Slide 1

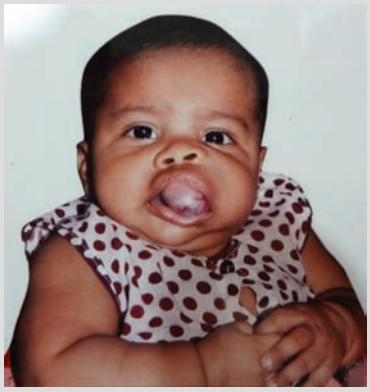


Histopathological Slide 1

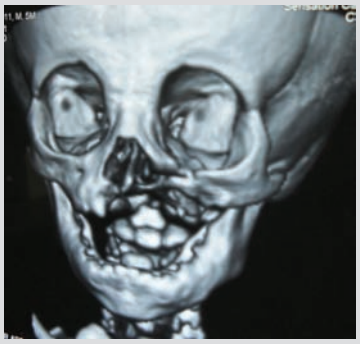
Follow up was done after four weeks, after eight months and after one year no further growth or recurrence was noted with a normal face profile.

Case 2

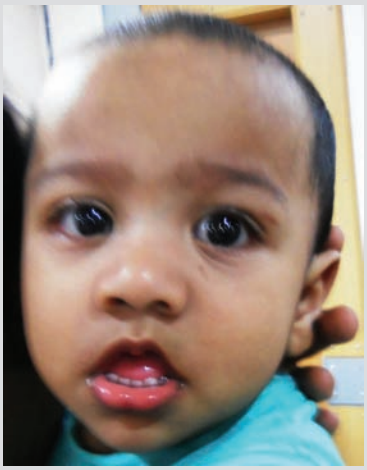
A 2 months old baby boy Master Abdullah was coming from Khulna in Oral and Maxillofacial Surgery Department of Anowar Khan Modern Medical College and Hospital with the complain of swelling of the anterior maxilla and also complain of bleeding from the swelling on 24/07/2011. Fine needle aspiration cytology (FNAC) was done. A few drops dark of pigment was aspirated and sent for cytology. Microscopically, the smear showed small oval cells in clusters as well as singly where some cells contained melanin pigmentation. The cells had hyperchromatic nucleus with scanty cytoplasm. The diagnosis was made as melanotic neuroectodermal tumor of infancy (MNTI). CT Scan of facial bone was also done and shows a large bony growth on anterior maxilla. Excision of the tumor with partial maxillectomy was done on 17/09/2011 and sent for histopathological examination and the report revealed a case of melanotic neuroectodermal tumor. Patient was followed-up for one year and there is no sign of recurrence was seen.



Pre-operative view



CT Scan of facial bone



Post-operative view after one year follow-up

Discussion:

More than 90% of the parents presents with the tumor in the first year of life, usually from age 1-6 months. The mean age of patients with neuroectodermal tumor of infancy (MNTI) is 4.3 months. But a few cases have been reported in adults and may occasionally develop as congenital.

Though MNTI is classified as a benign lesion, but can be aggressive locally and invades the bone marrow without forming any fibrous capsule. Malignant transformations are also reported in about 3.2 % cases.[3] When metastases develop, the smaller neuroblastic cells predominate in the secondary deposits in histology and may resemble neuroblastoma .[4]

More than 90% of MNTI occur in the head and neck regions.Typically, MNTI develops in the maxilla and less commonly in the mandible, but extramaxillary locations have been reported in the skull, long bones, epididymis, mediatinum, soft tissues of extremities, cheek, and rarely brain.[5] Joseph C et all reported a case of MNTI of skull base recurring in the posterior fossa following metastasis to the subarachnoid space.

The MNTI reported recurrence rate of 10-20% and multicentric character of the lesion may be responsible for recurrence. Some of the cases were reported as distant metastasis.[6]

Some authors advocated the necessity of post-operative chemotherapy to prevent distant metastasis and aggressiveness that was estimated less than 5%.[7]

The tumor appears as a sessile or slightly pedunculated, lobulated, firm mass typically deep blue or with a dark discoloured surface. It is usually 2-4cm in diameter but may show a huge involvement showing a well defined or a poorly defined destructive radiolucency of the bone with a “sunburst” appearance as mild calcification along vessels radiating from the centre of the tumor.

This tumor may show pseudo capsulation perhaps with reactive bone formation at the periphery of the lesion. Developing tooth buds may be encircled by the tumor leading to misdiagnosis as an odontogenic tumor. Besides the histopathology, immunohistochemical study and urinary vanillylmandelic acid (VMA) production confirm the neural origin of MNTI. Increased serum level of adrenaline, nor adrenaline, urinary VMA remain normal after surgery and chemotherapy.[8]

Conclusion

Although MNTI behaves as a benign lesion, recurrences can occur especially within the first 6 months with the need for close follow-up postoperatively. Early detection and treatment will avoid further complications and may support a favorable outcome for the patient. In this case, early diagnosis and treatment prevented further complications and the patient was followed up for 6 months without any evidence of recurrence.

References:

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