

## Melanotic Neuroectodermal Tumor of Infancy

Satish Chandra Tripuraneni

### ABSTRACT

Melanotic neuroectodermal tumor of infancy (MNTI) is a relatively uncommon osteolytic-pigmented neoplasm that primarily affects the jaws of newborn infants. Here, we are reporting a case of MNTI presented at the age of 12 years. The objective of this report is to review the incidence, presentation, investigation protocols, management options and outcomes.

**Keywords:** Melanotic neuroectodermal tumor of infancy, Vanillylmandelic acid, Neural crest, Weber-Ferguson .

**How to cite this article:** Tripuraneni SC. Melanotic Neuroectodermal Tumor of Infancy. Clin Rhinol An Int J 2012;5(3):138-139.

**Source of support:** Nil

**Conflict of interest:** None declared

### INTRODUCTION

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare tumor mostly involving jaw bones with common age of presentation in early childhood, arising from neural crest cells. It is a slow growing benign tumor with pigmentation and associated with high urinary vanillylmandelic acid (VMA). Treatment of choice is surgical excision with clear margins followed by prosthetic support, if necessary.

### CASE REPORT

A 12-year-old boy presented with swelling of left maxilla since 3 months of age. He was asymptomatic except for cheek swelling and facial disfigurement. On examination, he was found to have diffused nontender bony hard swelling of left cheek along with left upper alveolus (Fig. 1). Malocclusion was there. Mucosa over the swelling was unremarkable.

Computed tomographic (CT) PNS was taken showing diffuse involvement of anterolateral wall of maxilla including alveolus, palatine process and frontal process with well maintained maxillary sinus. Radiological findings of mixed radiolucency and radiopacity with widening of diploic space with displacement of outer table were reported (Fig. 2). Missing tooth was there. Possibility of monostotic fibrous dysplasia was considered. Partial maxillectomy and facial contouring by sublabial approach was planned.

During the procedure after exposing the maxilla, we found pigmentation of bony maxilla, possibility of MNTI was considered and total lesion was excised including alveolus and hemipalate by Weber-Ferguson's incision (Fig. 3). Later histopathology revealed the lesion as MNTI.



Fig. 1: Preoperative photograph

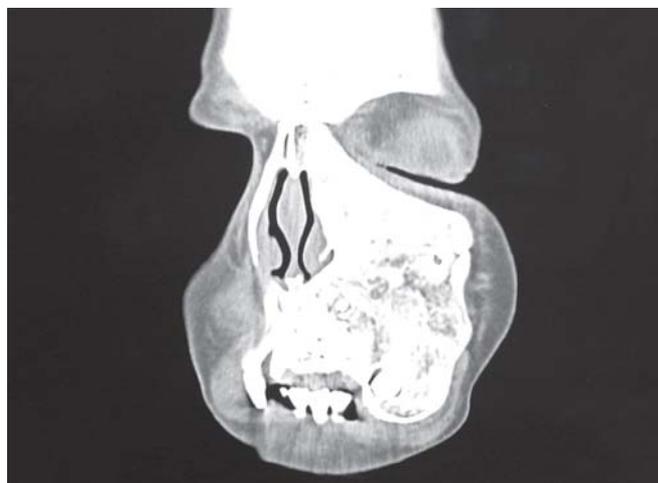


Fig. 2: Preoperative CT scan



Fig. 3: Specimen

After wound healing patient was given removable partial denture. Patient was followed up for 5 years without any recurrence.

## DISCUSSION

MNTI is a relatively uncommon osteolytic-pigmented neoplasm that primarily affects the jaws of newborn infants.<sup>1</sup>

Several patients with MNTI have demonstrated a high urinary excretion of VMA.<sup>2</sup> This finding adds credence to a neural crest origin because elevated VMA has been reported in neuroblastoma, ganglioneuroblastoma, pheochromocytoma and other neural crest tumors. However, the presence of urinary VMA is not diagnostic for MNTI.

Approximately 200 cases of MNTI have been reported in the literature. An exact number is difficult to discern because of the variety of terms that have been applied to the lesion in the past.<sup>3</sup>

The sexual predilection for MNTI is nearly equal, with a male-to-female ratio of 6:7.

Most patients, by some estimates more than 90%, present with the tumor in the first year of life, usually from age 1 to 6 months. The mean age of patients with MNTI is 4.3 months. Although extremely rare, a few cases of MNTI have been reported in adults, notably, a 23-year-old man, a 24-year-old woman and a 67-year-old woman.

- Although MNTI is classified as a benign lesion, it is often clinically worrisome because of its rapid onset and alarming local growth rate.
- More than 90% of MNTI occur in the head and neck region, with most on the anterior part of the maxillary ridge. Other common sites include the skull, the mandible, the epididymis and the brain.<sup>4</sup> Rare lesions have been reported in the shoulder, the skin, the femur, the mediastinum and the uterus.
- Plain dental radiography, CT scanning and MRI have been used to evaluate the content and the extent of MNTI.
- Grossly, the specimen has a gray, hard, rubbery consistency with foci of blue-black pigmentation. Additionally, entrapped developing tooth buds may be noted in the specimen as MNTI grows in and around the odontogenic apparatus.

The treatment of choice for MNTI is surgical excision, with a 5 mm healthy margin<sup>5</sup> and it is usually curative. This treatment can usually be accomplished with a partial maxillectomy by using a Weber-Ferguson incision and a facial degloving approach.

The possibility of local recurrence is a problem that has been documented to range from 10 to 60% of patients depending on the study quoted. Overall, the average recurrence rate is 15 to 20%. The recurrent lesions, possibly secondary to inadequate excision or multicentricity, usually become apparent within the first year after surgery. In instances of inoperable recurrence or where clear margins are impossible to obtain, radiation therapy and/or chemotherapy have been used, but too few examples exist for preferences to be established.<sup>6</sup>

Permanent reconstruction of the maxillary alveolus and missing dentition may have to be delayed until after growth is completed, often in the teenage years. In the interim, transitional removable partial dentures may be necessary. The skills of an orthodontist, prosthodontist, oral surgeon and/or dentist may be required, based on the extent of the missing structures, to correct any functional and cosmetic deformity.

Most MNTI are benign and effectively managed by aggressive surgical excision. Reconstruction may be challenging. Approximately 1% of tumors are malignant, with only rare tumors producing metastases.

In our case even though the child presented at the age of 12 years, the lesion was first appreciated by his mother at the age of 3 months. But that was gradually progressive till the age of 12 years where as the typical MNTI is a rapidly growing benign lesion. As the CT picture was suggestive of fibrous dysplasia we have not considered the possibility of MNTI preoperatively and we have not assessed the urine sample for VMA. But the intraoperative finding of pigmentation was clinching of MNTI, we have gone for a radical resection.

## CONCLUSION

MNTI should be considered for all the lesions of facial bones especially, if they start growing in infancy and early childhood and urinary VMA can be assessed along with radiological investigations for better planning of treatment.

## REFERENCES

1. Krompecher Z. Zur histogenese und morphologic den adamantinome und sonstiger kiefergeschwulste. Beitr Pathol Anat 1918;82:165-97.
2. Borello ED, Gorlin RJ. Melanotic neuroectodermal tumor of infancy—a neoplasm of neural crese origin. Report of a case associated with high urinary excretion of vanilmandelic acid. Cancer 1966 Feb;19(2):196-206.
3. Neville B, Damm D, Allen C. Melanotic neuroectodermal tumor of infancy. In: Oral and maxillofacial pathology (3rd ed). St Louis, MO: Saunders/Elsevier 2009:533-34.
4. Matsumoto M, Sakuma J, Suzuki K, et al. Melanotic neuroectodermal tumor of infancy in the skull: Case report and review of the literature. Surg Neurol 2005 Mar;63(3):275-80.
5. Dashti SR, Cohen ML, Cohen AR. Role of radical surgery for intracranial melanotic neuroectodermal tumor of infancy: Case report. Neurosurgery 1999 Jul;45(1):175-78.
6. Woessmann W, Neugebauer M, Gossen R, et al. Successful chemotherapy for melanotic neuroectodermal tumor of infancy in a baby. Med Pediatr Oncol 2003 Mar;40(3):198-99.

## ABOUT THE AUTHOR

### Satish Chandra Tripuraneni

Professor, Department of Otorhinolaryngology and Head and Neck Surgery, Dr Pinnamaneni Siddhartha Institute of Medical Sciences and Research Foundation, Gannavaram, Andhra Pradesh, India  
e-mail: sctripuraneni@yahoo.com