

Melanotic Neuroectodermal Tumor of Infancy

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ABSTRACT

Melanotic neuroectodermal tumor of infancy is a rare congenital neoplasm involving the head and neck region in young patients. A case of melanotic neuroectodermal tumor of infancy is presented. This tumor occurred in left maxillary alveolar ridge in a 5-month-old female. The present case showed an increased urinary level of vanillylmandelic acid, confirming that the tumor is originated from neural crest. Clinical assessment, histologic diagnosis and laboratory findings supported the diagnosis.

Keywords: Benign, Melanotic neuroectodermal tumor of infancy, Pediatric dentistry.

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INTRODUCTION

Melanotic neuroectodermal tumor of infancy (MNTI) is a relatively uncommon osteolytic-pigmented neoplasm that primarily affects the jaws of newborn infants. The lesion has had an interesting history since its initial description by Krompecher in 1918 as a congenital melanocarcinoma.^{1,2} Most of the tumors are located in the maxilla and become evident in the first year of life. Only a few cases occur in the neurocranium. Most patients were taken to a hospital because of progressive growth of a craniofacial mass, without associated neurological symptoms. The classic presentation is of a rapidly expanding mass that may have a blue, brown or black coloration.³ They present osteolytic lesions with associated dissolution and degeneration of surrounding bony tissues, often associated with displacement of developing teeth.³

We report a case of MNTI in a 5-month-old female infant showing biphasic tumor cell population with melanin pigmentation, increased urinary excretion of VMA level, emphasizing the need for appropriate treatment.

CASE REPORT

A 5-month-old female patient was referred to department of maxillofacial surgery and dentistry with a complaint of an expansile growth of the maxilla. The swelling was first noticed 2 months ago and slowly increased to the present size. Clinical examination showed a normally developed female infant. On the anterior region of the maxilla, a firm nonulcerated reddish-tumor was seen to be protruding through the lips and covered by the intact mucosa (Figs 1 and 2). Computed tomography (CT) scan showed a radiolucent lesion expanding the surrounding bone, and confirmed the displacement of the left and



Fig. 1: Extraoral view showing the bulge on left side



Fig. 2: Intraoral view showing bulge with intact mucosa

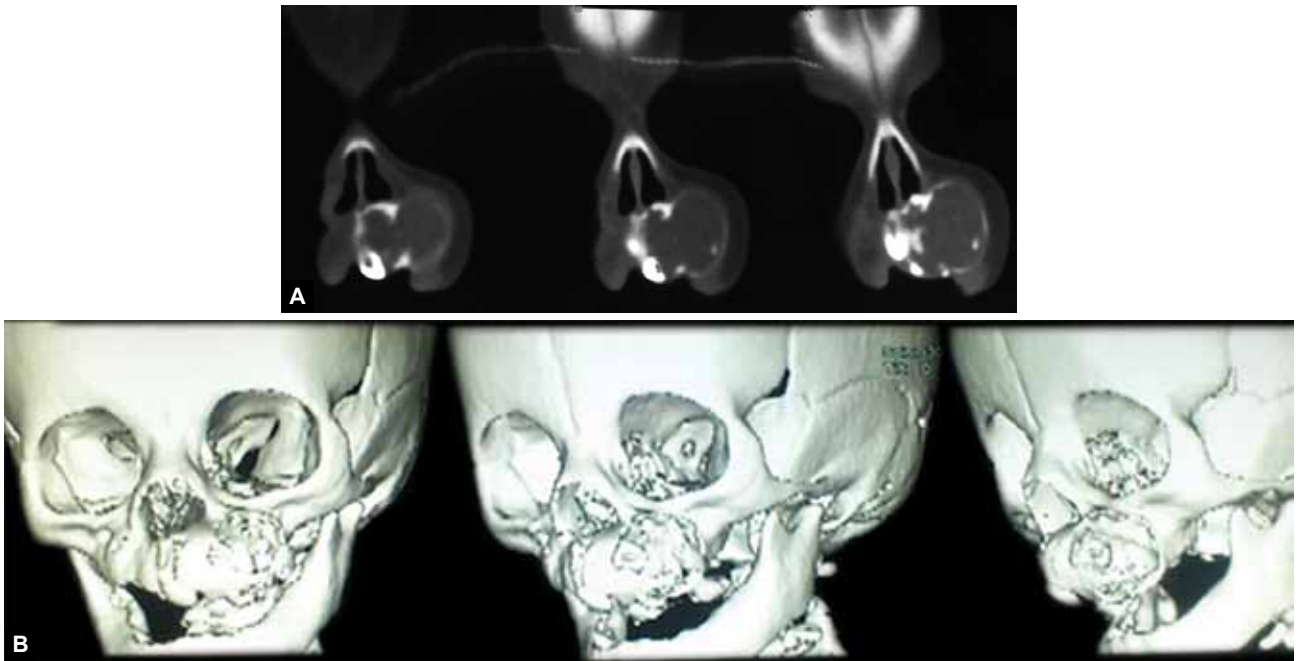
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Figs 3A and B: Computed tomography revealing an extensive mass resulting in expansion of anterior maxilla on the left side

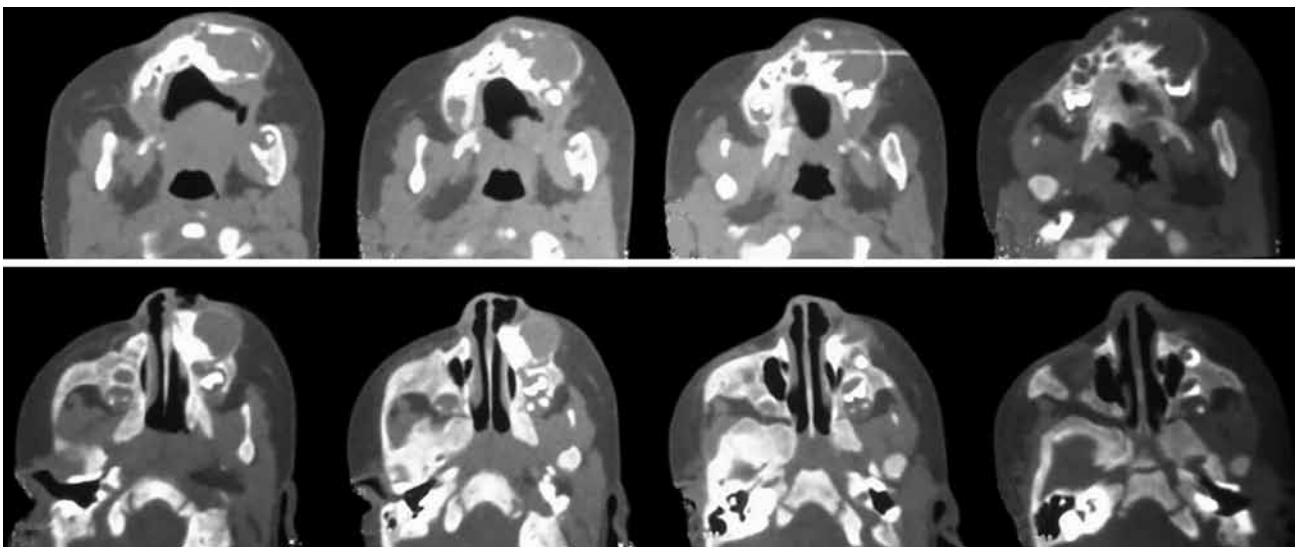


Fig. 4: Computed tomography showing displacement of deciduous teeth

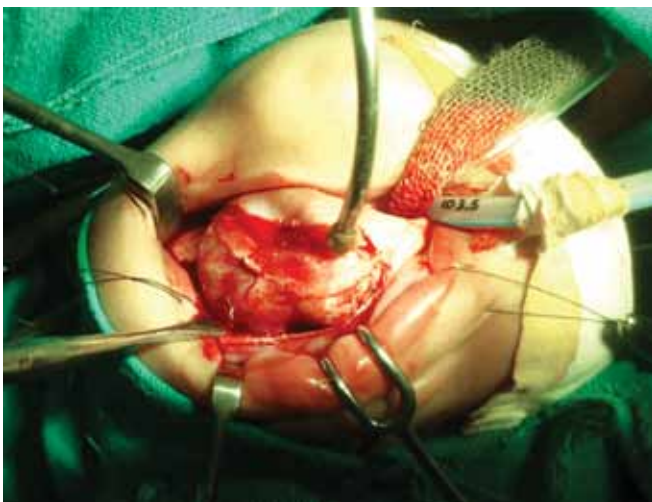


Fig. 5: Enucleation of the tumor

right upper deciduous central incisor and the left upper deciduous lateral incisor (Figs 3 and 4).

Surgical excision was performed under general anesthesia, with curettage of the bone (Fig. 5). The tumor was well demarcated. The specimen showed an intraosseous lesion that eroded the bone and was covered by intact overlying mucosa. Bone destruction was also seen at the periphery of the tumor while the center of the lesion did not contain any bone fragments. The background of the tumor was fibrotic and contained irregular islands of tumor cells (Fig. 6). There appear to be two population of tumor cells, the large melanotic (pigmented) type and the small (nonpigmented) type. Based on the above histopathologic findings, the lesion was finally diagnosed as MNTI (Fig. 7).

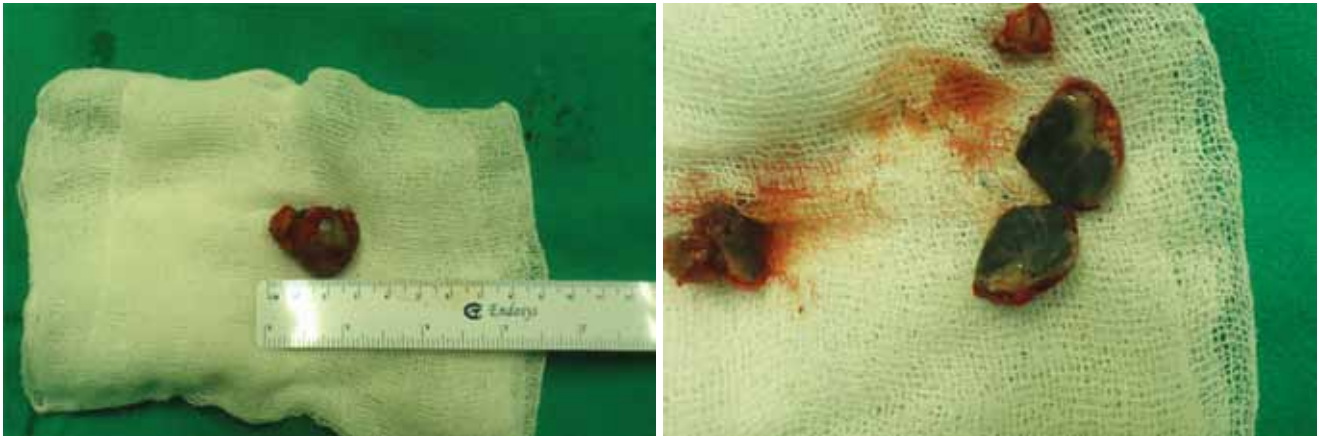


Fig. 6: Enucleated tumor having black pigmented appearance

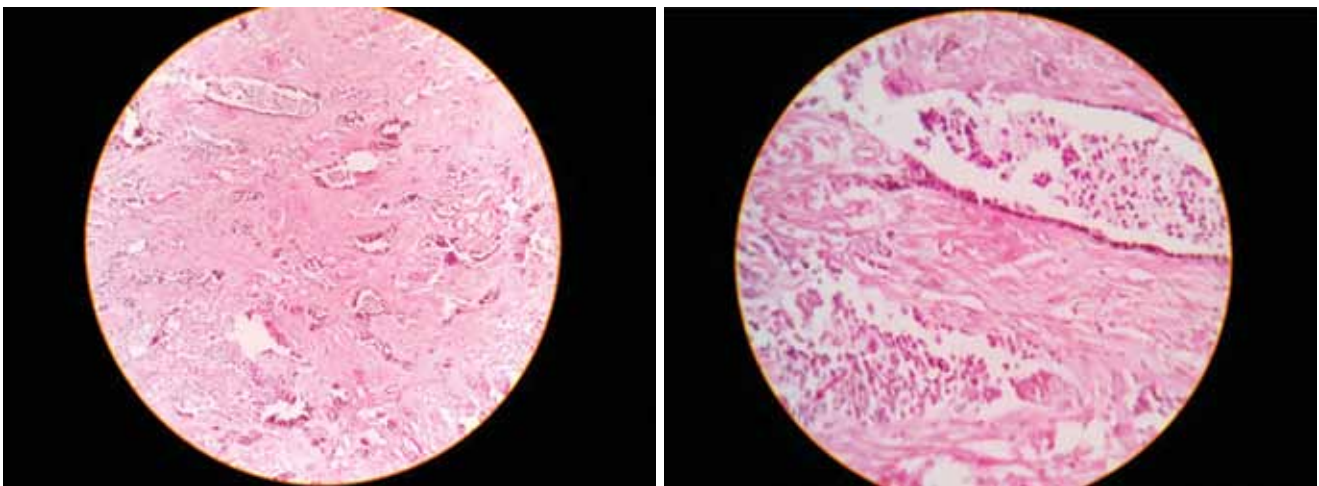


Fig. 7: Histopathologically showing biphasic tumor population

DISCUSSION

First described by Krompecher in 1918, the MNTI is a rare tumor, and approximately 200 cases have been reported.⁴⁻⁶ It is usually benign, though locally aggressive and fast growing. Most of the patients are infants, although a few adult patients have been reported. Male to female ratio is almost equal (6:7) and the mean age of the patients is 4.3 months. Of the reported tumors, 68.6% are located in the maxilla, followed by the skull (10.6%), mandible (7.3%), intracranial structures (5.3%), epididymis (4%), soft tissues, uterus and mediastinum.⁷

Several theories have been proposed to explain the pathogenesis of this neoplasm which recapitulates the early stages of retinal development (retinal anlage tumor). A congenital dysembryogenetic neoplasm arising from neural crest cells is the posited theory best supported by embryologic, ultrastructural, biochemical, immunohistochemical, electron microscopic and molecular genetic studies.⁸⁻¹⁰ Support for this proposed neuroectodermal origin is given with secretion of vanillylmandelic acid or other catecholamines by the neoplastic cells, a finding characteristic of other tumors of neural crest origin, such

as pheochromocytoma, neuroblastoma and ganglioneuroblastoma.

Radiographic examination of MNTIs usually reveals a radiolucency with or without regular margins.¹¹ In some cases, an associated osteogenic reaction, which exhibits a 'sun ray' radiographic pattern, may be seen and may lead to a misdiagnosis of osteosarcoma.³ Computed tomography scanning tends to show hyperdense masses and can accurately define the extent of the lesion, showing tooth displacement, as seen in this case.^{11,12} Magnetic resonance imaging with gadolinium contrast provides optimal tissue characterization while avoiding the radiation exposure of a CT scan.¹² The ectodermal band that ultimately gives rise to the dental lamina is intimately associated with neuroectodermal cells which are theorized to be the origin of the MNTI.

The treatment of choice consists of complete surgical excision. Individuals with MNTI that are not amenable to surgical management alone may receive other modes of treatment.¹³ In general, this may be chemotherapy alone, chemotherapy with radiotherapy, chemotherapy before and after the surgical treatment, radiotherapy and surgical treatment or a combination of all. Chemotherapy may

serve as an alternative or adjuvant option in the treatment of widely extended MNTIs.¹⁴

CONCLUSION

Although MNTI behaves in a benign fashion, 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 the present case, early diagnosis and treatment prevented further complications and the patient was followed-up for 12 months without any recurrence.

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