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PIGMENTED NEUROECTODERMAL TUMOR IN MAXILLA OF AN INFANT : REPORT OF A RARE NEOPLASM IN BANDUNG, INDONESIA

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Abstract
Introduction: Melanotic neuroectodermal tumor of infancy (MNTI) is a rare, locally invasive, fast-growing neoplasm with a high recurrence rate, which usually affecting children under 1 year of age. The tumor originated from the neural crest, most commonly found in the head and neck region, especially in the maxilla. Current standard for treatment is by surgical excision, but the high recurrency rate (10% - 60%) and the possiblity of malignant transformation makes the extent of excision debatable.
Case report: An 8 -months-old boy was referred to our department with a rapidly growing mass in the anterior of the maxilla since two months prior. The mass was bluish in color, firm, and non-ulcerative. We treated the patient with excision under general anesthesia. Pathology examination was done and MNTI was confirmed. There is no recurrency at 2 years follow-up.
Summary: Diagnosis and treatment of MNTI must be done meticulously due to the aggressive nature of the tumor. Wide surgical excision is the best treatment for this case, and routine follow-up must be done to detect any reccurency.
Keywords: melanotic neuroectodermal tumor; infancy; maxilla, surgical excision

Introduction

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare pigmented lesion, originating from the neural crest.[1] The diagnosis is usually made in the first year of life, although some cases were found in adults.[2, 3] First described by Krompecher in 1918, the tumor used to have different nomenclature due to the controversies of the origin.[1, 4] Borello and Gorlin (1966) found that the tumor was associated with an increased levels of vanillylmandelic acid (VMA) and suggested that the tumor is of neural origin.[4, 5] There was approximately 500 cases reported worldwide between 1918 until now.[3, 5] In Indonesia, there were only one case that has been reported in literature.[6]

Head and neck region have a great predilection of this tumor, about 67% of all reported cases, maxilla is the most common site affected. Other site has also been reported such as reproduction system, extremities, and skull.[1, 7, 8] Gender predilection is slightly higher in males. MNTI is recognized as a benign tumor, although it is locally aggressive and has a 3-6% possibility of distant metastases.[2, 9, 10]

The clinical presentation of the tumor is a rapidly growing, painless, non-ulcerative, bluish-black mass on the anterior side of the maxilla. Adjunctive examination such as CT scan, histopathological through incisional biopsy, and examination of vanillylmandelic acid (VMA) was done to confirm the diagnosis. There is no current consensus on the treatment of MNTI, but surgical resection was reported to be successful in numerous reports. Recurrency was observed in 20% of the cases. Recurrent MNTI is reported to have an excellent result with radiotherapy and chemotherapy.[4, 5, 11] We hereby present a case of MNTI treated with surgical resection without the use of any graft.

Case Report

An 8-month-old male infant was referred to Oral and Maxillofacial Surgery Department of Hasan Sadikin General Hospital, the parents noticed a growing mass on the maxilla in the last 2 months. The mass started as a little white lump in the size of a small pea on the maxilla gingiva, it has been rapidly growing especially in the last month before admission. The mass disrupted feeding process because of the difficulty of suction mechanism.

Extraoral examination showed an asymmetrical face with disappearance of the nasolabial sulcus, elevated alar base, and disruption of nose shape due to the tumor size on the right side (Fig 1a). Upon intraoral examination, a bluish projecting mass with 3x2x2cm in size was found on the alveolar ridge of the right maxilla, extending to the anterior palate. The mass was firm on palpation, and no ulceration was observed. There were no other abnormalities found on the oral mucosa other than the lesion site. The deciduous teeth were not yet erupted (Fig 1b)

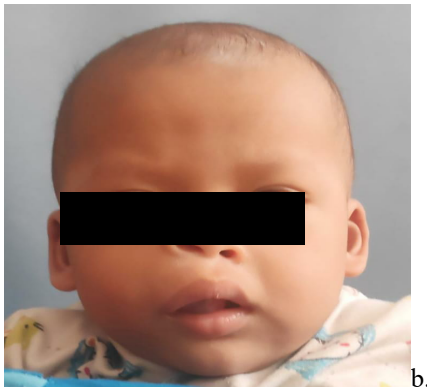
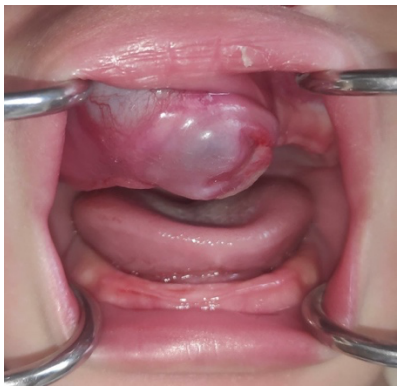
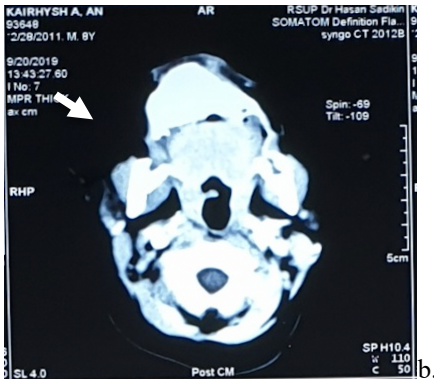
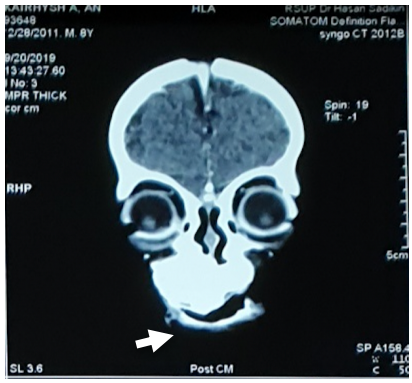
a.  b. 

Figure 1. Clinical manifestation. (a) Asymmetrical face with elevated right alar base; (b) Bluish mass projecting from right maxilla with 3x2x2cm in size

a.  b. 

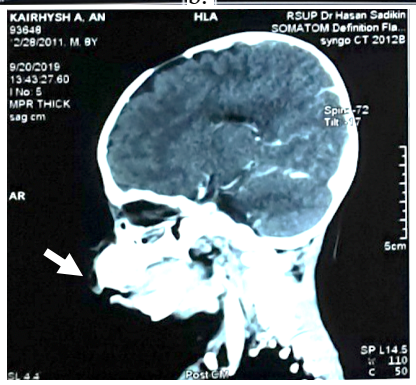
c. 

Figure 2. CT scan examination axial (a), coronal (b), and sagittal (c) view, a solid inhomogenic mass (white arrow) originating from maxilla, obliterating palate, right masticatory space, and infiltrate right maxillary sinus.

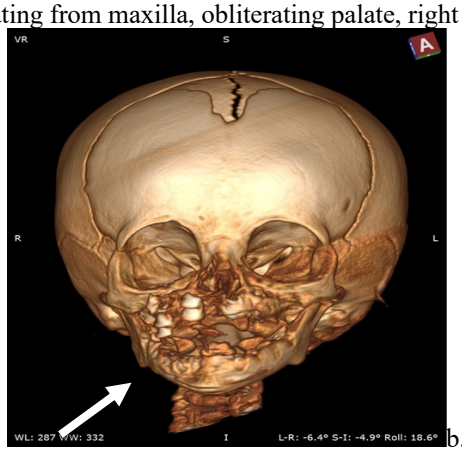
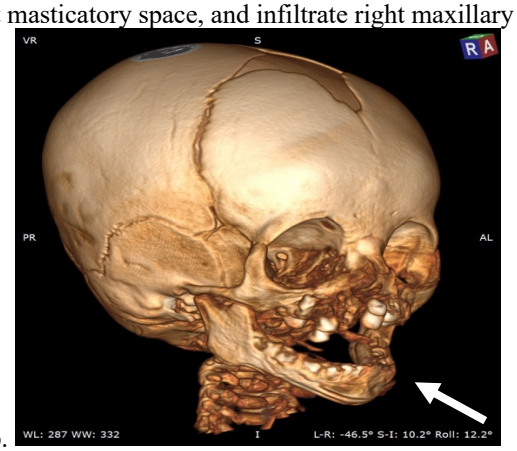
a.  b. 

Figure 3. CT scan 3D reconstruction frontal (a) and lateral oblique (b) view. Bone destruction observed at right maxilla region with tooth bud displacement (white arrow).

CT scan examination of the head and neck region showed an inhomogenic solid mass originating from the maxilla (Fig 2,3). The mass obliterated the soft palate, right masticatory space, and infiltrated the right maxillary sinus. There were no bleeding, ischemic lesions, and vascular malformations observed. Laboratory examination showed slight thrombocytosis. VMA level was not examined due to the urgency of the case.

Melanotic neuroectodermal tumor of infancy was diagnosed based on clinical and radiographic examinations. Multidisciplinary discussion was conducted prior to surgery, surgical resection under general anesthesia with 3mm of healthy tissue margin resection was decided (Fig 4). The tumor specimen was sent to the Pathological Anatomy Department for histological examination.

Based on the pathology examination results, MNTI diagnosis was confirmed. There was no adjuvant treatment done before or after surgical treatment. Two years follow-up showed no signs of recurrency, no significant stunt of maxillary growth other than no primary teeth is erupting at the tumor site (Fig 5).

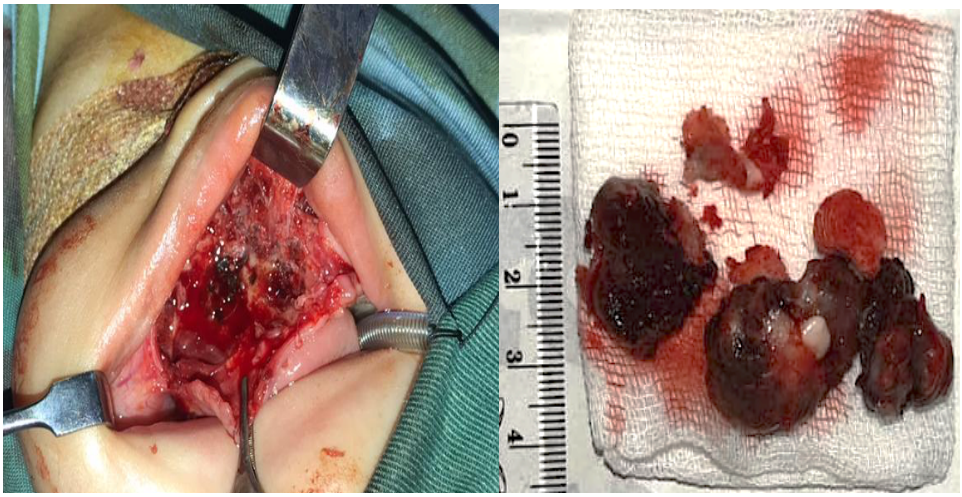


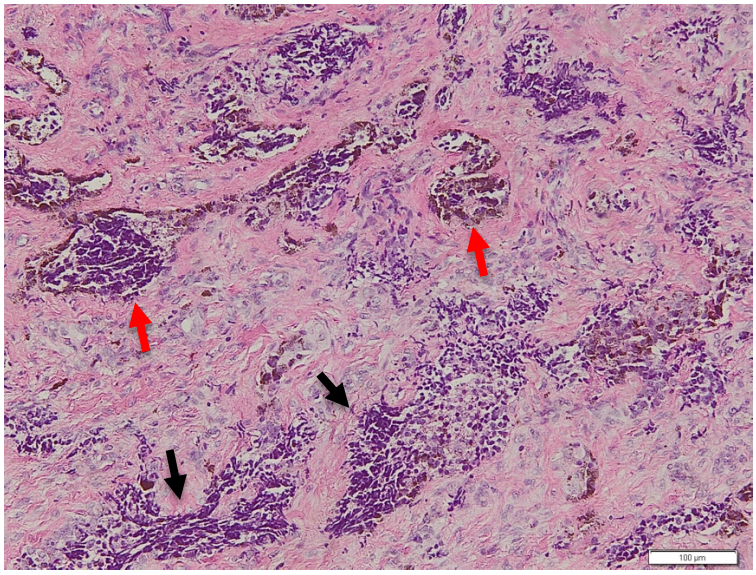
Figure 4. Intraoperative findings.

Figure 5. Two years follow-up showed no signs of recurrency, no significant stunt of maxillary growth other than no primary teeth is erupting at the tumor site.

Histological findings

The tumor volume was 3x1.7x1cm on the largest piece, and 1.7x1.2x1 cm on the smallest, there was a tooth found with no tumor invasion to the tooth. Microscopic examination showed a biphasic population of cells (Fig 6a). A neuroblastic-like appearance consists of small round, oval cells with pleomorphic and hyperchromatic nuclei and little cytoplasm. The other cell type is round, oval cells in cord and nested formation. Melanin pigment was found in the cytoplasm (Fig 6b). Immunohistochemistry study was performed. The result was positive for cytokeratin and synaptophysin staining (Fig 7). This founding confirmed the MNTI diagnosis.

A.



B.

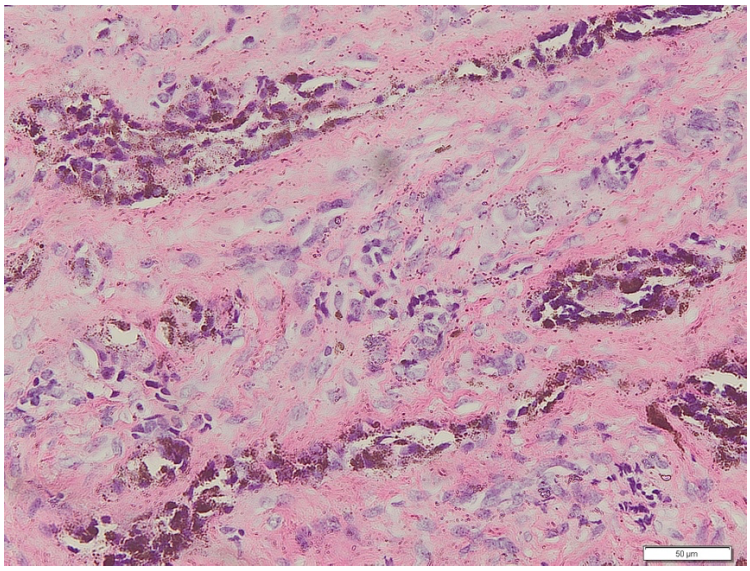
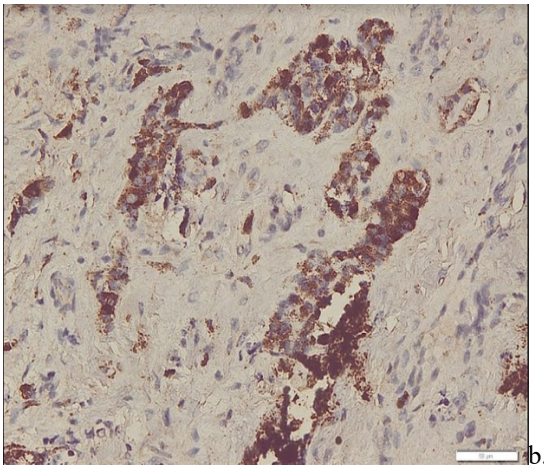


Figure 6. A. Biphasic cell population, “neuroblastic-like” (black arrow) and nested cells (red arrow). B. Melanin pigment was found in the cytoplasm.

a.



b.

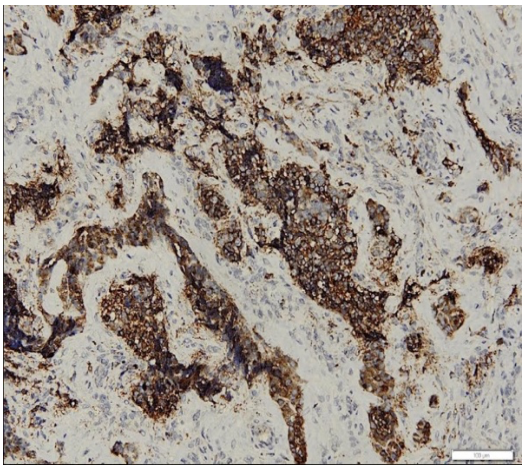


Figure 7. (a) Cytokeratin and (b) synaptophysin staining positive.

Discussion

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare, benign tumor mostly developed in the first year of life, although there were some congenital and adult cases reported. The first case of MNTI was reported by Krompecher in 1918 as a congenital melanocarcinoma, he described a pigmented tumor of the maxilla associated with a developing teeth in an infant.[12] The origin of this tumor was a controversy in the early time, hence the many names suggested, such as pigmented epulis, congenital melanocarcinoma, pigmented adamantinoma, retinal anlage tumor, and pigmented tumor of the jaw of infants.[3, 5, 12] In 1966, Borello and Gorlin reported a melanotic tumor in an infant and found an elevated level of urinary vanillylmandelic acid (VMA), the VMA level was returned to normal after the removal of the tumor. High level of VMA is useful for diagnosing a tumor of neural crest origin. Thus, it was determined that MNTI is a tumor of neural crest origin.[12, 13] The tumor mostly affected men compared to women, most commonly affecting the head and neck region, maxilla is the most common site affected (68-80%), skull and mandible were also reported as the site predilection of this tumor.[14] Other places such as brain, peripheral bones, mediastinum, ovary, and testis has also been reported on literature.[7, 15]

Recurrent case of MNTI was observed on 15-27% of cases, the recurrency was found as early as 4 weeks after surgical resection. The recurrent lesion may have been caused by incomplete resection and the occurrence of multicentric tumors. To prevent recurrency of the tumor, early diagnosis and treatment is needed, but the low incidence of the tumor may delay the diagnosis and treatment.[3, 5] Malignant transformation and metastases was also reported.[9, 16]

Clinical manifestation of the tumor is a rapidly growing, painless, expansile, blueish-black pigmented mass, although the pigmentation may sometimes invisible due to the tissue covering it. It is considered as a benign tumor by the World Health Organization, despite its locally aggressive nature. In CT examination, the lesion is intraosseus expansive lytic lesions with irregular margins and may envelop or displace adjacent teeth. MRI can also be used to examine the extraosseous lesion. Radiographic examination is useful in determining the surgical design. [10, 16, 17]

Histologically, the H&E stained specimen showed a biphasic population of cells forming nests, tubules, or alveolar structures within a fibro cellular connective stroma. The alveolar and tubular structures are lined by a cuboidal epithelioid cell with granules of melanin pigment. The other cell types are small, round, and neuroblast-like cells with hyperchromatic nuclei and little cytoplasm.[1, 4] Immunohistochemistry studies can be helpful to assist with diagnosis.[18] Cytokeratin, synaptophysin and HMB-45 are widely used for IHC markers, Ki-67 and CD99 are also used to determine tumor aggressiveness.[3, 19] The pathological anatomy and immunohistochemistry findings of our patient were consistent with the one reported in the literature, thus the MNTI diagnosis was made.

The treatment of this tumor is mainly surgical resection, although there is still no consensus on the treatment guideline.[3] Radiotherapy and chemotherapy have been successful as adjuvant or neoadjuvant therapy, and for the treatment of the unresectable and malignant cases.[16, 20]. High recurrence rate of MNTI makes it important to have a clear margin upon the resection of the tumor.[3, 16] Wide margin resection is debatable for the head and neck region due to the proximity of vital structures, some literature suggests 2-5mm healthy tissue margin to prevent recurrency.[4, 5, 15,21] Early detected recurrency may easily be managed by resection.[16] In our patient, we resected a 3 mm healthy tissue margin, and there was no recurrent lesion observed at the two years follow-up.

Conclusion

Melanotic neuroectodermal tumor of infancy is a rare benign but aggressive tumor that mostly affects the head and neck region. Although benign, there is a high recurrence rate and possibility of metastases. Malignant transformation of the lesion also has been reported and generally fatal. Early diagnosis and treatment is important to prevent future recurrency. Surgical resection is the standard of treatment, but there is still no consensus on the treatment. Chemotherapy and radioteraphy were also found effective as adjuvant treatment. Rarity of the case may delay the diagnosis and resulting in a less desirable outcome. Close follow-up after the procedure is needed to detect any recurrency early.

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