Case Report

Surgical Management of Melanotic Neuroectodermal Tumor of Infancy: A case Report

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Abstract

Melanotic neuroectodermal tumour of infancy (MNTI) is rare, rapidly growing, pigmented neoplasm of neural crest origin. It is generally accepted as a benign tumour despite of its rapid and locally destructive growth. It primarily affects the maxilla of infants during the first year of life. Surgical excision is considered as the treatment of choice. The recurrence rate varies between 10% and 15%, and malignant behaviour has been reported in 6.5% of cases. We report a 15-month old male who presented with a 2-month history of a rapidly growing mass in the anterior. A biopsy showed melanotic neuroectodermal tumor, and complete resection with negative margins was subsequently achieved.

Keywords: Melanotic neuroectodermal tumor, infancy.

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Introduction

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare pigmented neoplasm of neural crest origin that most commonly presents in infants during the first year of life. It is predominant in males and most commonly arise in the maxillary region. In the past, it has been known by a variety of names, such as congenital melanocarcinoma, melanotic epithelial odontoma, melanotic ameloblastoma, retinal anlage tumor, melanotic progonoma, pigmented adamantinoma, congenital pigmented epulis, and melanocytoma. Like other tumors of neuroectodermal origin, MNTI is frequently associated with elevated urinary excretion of vanillylmandelic acid (VMA), a metabolite of epinephrine and norepinephrine. Although an increase in VMA is helpful, this symptom alone is not diagnostic of MNTI.

A small number of cases have been reported in older children and adults. This tumor grows very fast and originates in the neural crest. MNTI generally originates from the soft tissue overlying the maxilla (68–80%), but it can occasionally arise in the skull (10.8%), mandible (5.8%) or brain (4.3%)⁴. In addition to the head and neck region, other sites can be affected by the condition less frequently, including the femur, epididymis, ovaries, uterus and mediastinum. Clinically, MNTI is soft and reddish-blue tumor that often destroys the underlying bone and prevents teeth development. Clinical and radiological findings can suggest a diagnosis of MNTI.

The treatment of choice for this tumor is surgical excision. Because of its relatively high recurrence rate, cases of MNTI should be monitored closely during the post-resection period and beyond. Although locally invasive, the risk of tumor metastasis is approximately 5%⁵,⁶.

We report a case of melanotic neuroectodermal tumor of infancy who was successfully treated at Hue Central Hospital.

Case report

A 15-month old boy presented at Hue Central Hospital due to the tumor at his maxilla that had been growing since 2 months of age. Facial asymmetry with the tumor in the anterior maxilla and hard palate was noted on exam. The tumor was oval, non-ulcerative, non-tender, and had a reddish-blue color. The tumor protruded through the lip, with a size of about 2×3×4 cm (Fig. 1). The patient could not suction and had difficulties feeding. The remainder of the physical examination was normal.

The magnetic resonance imaging (MRI) findings showed one heterogenous lesion at the right anterior maxilla mesuring 22×28×35 mm. The lesion showed hy-
perintense signal on T2W images. The lesion destroyed the maxilla and hard palate (Fig. 2). A chest and abdomen computed tomography (CT) showed no other sites of disease.

A needle biopsy showed a proliferation of a dual population of cells. Masson Fontana staining revealed positivity for melanin in peripheral epithelioid cells. Immunohistochemical analysis showed: 1) polygonal pigmented cells that were positive for HMB45 and EMA; and 2) small round cells groups with neurofilament background, that were positive for Synaptophysin and CD56 (Fig. 3). These findings were consistent with the diagnosis of melanotic neuroectodermal tumor of infancy.

A multidisciplinary team discussion was held. They all agreed with the final diagnosis of MNTI after ruling out the other possibility such as rhabdomyosarcoma, neuroblastoma, melanoma and lymphoma. For treatment, surgical resection was recommended. The procedure...
included an intra-oral resection of the tumor which involved the alveolus from approximately the first premolar tooth to the opposite premolar tooth. The posterior extent of the tumor was the incisive foramen. Gross specimen showing a well-circumscribed soft mass, measuring 2.0 cm × 3 cm × 3.5 cm (Fig. 4). Future surgical plans include the reconstruction of the bony deficit, probably with a cranial bone full thickness graft and soft tissue coverage with a temporals muscle flap. This would hopefully allow insertion of integrated dental implants to complete the reconstruction.

After surgery, the baby recovered soon. Two months later, he was checked with MRI (Fig. 5). The result showed no residual tumor. Ten months after surgery, there was not any sign of recurrence (Fig. 6).

Discussion

The first case of MNTI ever reported in the literature was designated as “congenital melanocarcinoma” by Krompecher in 1918. He described a pigmented tumor of the maxilla in a two-month-old infant. Since then it has been known by a variety of names, such as melanotic epithelial odontoma, melanotic ameloblastoma, retinal anlage tumor, melanotic progonoma, pigmented adamantinoma, congenital pigmented epulis, and melanocytoma. The term “melanotic neuroectodermal tumor of infancy” was recommended by Borello and Gorlin in 1966, when they reported a case of melanotic tumor in the maxilla of a three-month-old child who had increased urinary excretion of VMA before surgery, suggesting a neural crest origin.

MNTI generally occurs in the craniofacial region with a predominance for the maxilla (68–80%), followed by the skull (10.8%), mandible (5.8%). Other sites can be affected, such as: brain (4.3%), epididymis, mediastinum, ovary, uterus and peripheral bone. The MNTI is seldom congenital. It emerges within the 1st year of life, mostly...
below 6 months of age. Infants present with a painless, non-ulcerative bluish black gingival mass that is often confused with an eruption cyst. It may appear to be malignant due to its rapid growth potential\(^9\). Metastatic disease is rare, occurring in only 3% of patients. When metastases develop, the smaller neuroblastic cells predominate in the secondary deposits, and histology therefore resembles neuroblastoma more than MNTI\(^10\).

MNTI has a significant destructive nature with high growth potential. The treatment of choice for MNTI is surgical excision, and it is usually curative. Existing teeth and developing teeth must be sacrificed when they lie within the lesion or near the borders of an MNTI. Some controversy still exists regarding the amount of adjacent bone that needs to be removed during the surgical procedure. Some clinicians suggest that peripheral excision with 2–5 mm margin were generally considered as ideal. Others advocate for only enucleation of the tumor followed by curettage of the bone cavity. However, a recent systematic review claims that no differences in recurrence rates were observed between curettage and resection\(^11\). In this patient, when doing surgery, we chose the tumor removal with 2 mm margin from the tumor.

The overall incidence of local recurrence is approximately 10–15%. Currently, age at manifestation is considered to be a strong prognostic indicator in MNTI. Infants who present within the first two months of birth have a higher risk of recurrence which generally occurs within 6 months from treatment. In contrast, diagnosis from 2.5 to 4 months is associated with an intermediate risk, and after 4.5 months of age the risk of recurrence appears to be minimal\(^12\),\(^13\). And most of recurrence cases could be saved by extensive resection. As documented by our case, a complete resection may be curative. Chemotherapy is usually recommended only in cases of unresectable or metastatic disease.

**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 10 months without any recurrence.

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**Patient consent:**

Consent to publish the case report was obtained. This report does not contain any personal information that could lead to the identification of the patient.

**Conflict of interest:**

The following authors have no financial disclosures.

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