



MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY


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ABSTRACT

Melanotic neuroecto dermal tumor of infancy (MNTI) is a rare benign, non-ulcerative, painless, rapid expansile tumefaction of neural crest origin with a high recurrence rate (10%-60%) and risk of malignant transformation as much as 6.6%. Approximately 92.8% of MNTIs occur in the head and neck region of which 68-80% occurs in the maxilla. In this case report we describe a 7 month-old child with MNTI involving the anterior region of maxilla, with the aim to highlight the diagnostic assets of the disease.

Key words: Infancy, Melanotic neuroecto dermal tumor, Vanillylmandelic acid.

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INTRODUCTION

Melanotic neuroecto dermal tumor of infancy (MNTI) is a rare benign, non-ulcerative, painless, rapid expansile tumefaction of neural crest origin with a high recurrence rate (10%-60%) [1]. It is also known as congenital melanocarcinoma, melanotic epithelial odontome, melanotic ameloblastoma, retinal anlage tumor, melanotic progonoma, pigmented adamantinoma, congenital pigmented epulis and melanocytoma [2-4]. The terminological variations reported illustrate the uncertainties about its histogenesis [5].

Approximately 92.8% of MNTIs occur in the head and neck region[1] of which 68-80% occur in the maxilla, 10.8% in the skull, 5.8% in the mandible and 4.3% in brain [1,6]. It generally occurs in infants before the age of 1year [7,8]. The mean age of patients at diagnosis is 4.3 months [9], with no significant gender predilection reported [6]. In

addition to the head and neck region, other sites affected by the condition less frequently, include the femur, soft tissues of extremities, shoulder, thighs, epididymis, ovaries, uterus and mediastinum [1,6,8]. Its risk of malignant transformation is 6.6% [6].

Similar to other tumors of neuroectodermal origin, MNTI is frequently associated with elevated urinary excretion of vanillylmandelic acid (VMA), a metabolite of epinephrine and norepinephrine; though this symptom alone is not diagnostic of MNTI [2].

Its histopathological features are distinctive, with tubular or alveolar formation of large melanin-containing cells around nests of smaller neuroblastic cells possessing fibrillar cytoplasm [7]. Here we present a case of MNTI in 7-month-old female diagnosed on the basis of histopathology and biochemical analysis.

CASE HISTORY

A 7-month-old female infant presented to our department with a swelling in maxillary anterior region. As reported by her parents, initially the

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swelling was small and since they associated it with eruption of tooth no medical advice was taken. The swelling rapidly increased in size over 1 month leading to difficulty in feeding. This raised concern amongst her parents for which they approached oral medicine and radiology department. Patient's pre- and perinatal histories were unremarkable. Growth and development of the infant was adequate for her age. No history of trauma to the region was reported. No airway difficulties were noted.

Extraoral examination revealed swelling over left side of face that obliterated the left nasolabial fold, and elevated the left nasal alar base. The lower eyelid of ipsilateral eye was raised, with red, ulcerated punctum over medial side without active pus discharge [Figure 1]. Intraorally a swelling measuring about $4 \times 4 \times 5$ cm was seen extending from midpalatine raphe to 5 cm buccally. Swelling was smooth with intact, bluish overlying mucosa [Figure 2]. Swelling was bony hard, fixed, non-fluctuant, non-pulsatile, non-purulent causing no discomfort on palpation. Tooth 61 was the only maxillary tooth clinically visible; it was noncarious, displaced medially and extremely mobile. No lymph nodes were palpable.

Considering the age, site, duration and clinical findings, sarcomatous growth was considered as the provisional diagnosis; although, well defined, round, smooth swelling was favouring swelling of benign nature like congenital epulis, neuroblastoma, teratoma, melanoma, lymphoma and hemangioma.

Maxillary occlusal radiograph revealed radiolucent expansile lesion over the left side with thin cortical margins having tooth buds of 51-55, 62 [Figure 3]. Computed tomography (CT) of para-nasal sinus revealed approximately $4.5 \times 3.7 \times 5$ cm sized expansile bony lesion with enhancing soft tissue component and cortical thinning arising from superior alveolar arch on left side that bulged inferiorly into the oral cavity, caused obliteration of left nasal cavity, and elevated the floor of left orbit causing proptosis of left eye, compression and displacement of left maxillary sinus supero-medially and displacement of the teeth around the lesion [Figure 4]. Magnetic resonance imaging (MRI) revealed an expansile, isointense (T1WI) lesion which was heterogeneously hyperintense compared to muscle (T2WI) [Figure 5]. Radiographic findings were suggestive of benign tumor.

Histopathology findings revealed loose to dense fibrocellular connective tissue mass with aggregates of small and round cells showing large dark nucleus and scanty cytoplasm with some aggregates exhibiting melanin pigmentation inside

and around these round cells suggestive of MNTI [Figure 6].

For further confirmation, 24 hour urinary excretion of VMA was done and was found to be increased (2.6 mg/24 h) confirming the diagnosis. Also levels of alkaline phosphatase (191IU/L), lactate dehydrogenase (860U/L) and serum uric acid (10.2mg/dl) were raised.

Surgical excision of the lesion was advised however, patient's parents showed negligence and did not give consent for the treatment.

DISCUSSION




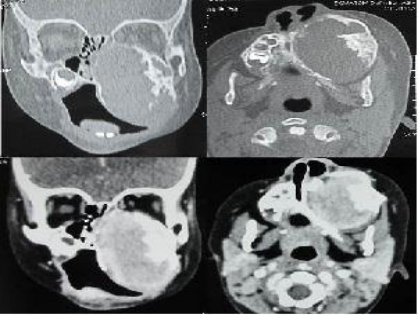
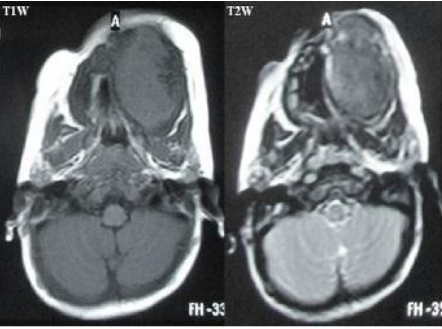
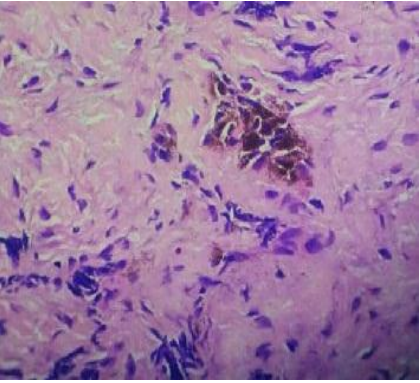
The most common site of occurrence of MNTI is the anterior maxillary alveolar ridge (70% cases) [7]. The classic clinical presentation is a sessile, firm swelling involving the upper anterior alveolar ridge and anterior hard palate. The presence of melanin gives it a bluish hue, often mimicking a vascular malformation. It may result in facial asymmetry, displacement of teeth and difficulty in feeding [8]. These findings were consistent with that of presented case.

Some benign lesions tend to have an alarming growth rate which may lead to misdiagnosing them clinically as malignant lesions. Owing to the age of the patient and the alarming growth rate of the swelling, the presented case was clinically diagnosed as sarcoma. However, radiographic findings favoured benign lesion. Histopathology and raised level of VMA helped to arrive at a definitive diagnosis as MNTI.

Though the primary treatment for MNTI is surgical resection though, it is generally agreed that chemotherapy is indicated for patients not amenable to surgical treatment, or for use as an adjuvant therapy prior to and following surgery. Radical resection may reduce the risk of relapse for a fast growing tumor, and extended resection is often applied to reduce the risk of malignant transformation. However, the effects of radical resection on post-operative growth and development should be taken into consideration to minimize any loss of tissue function [10].

High recurrence rate of these lesions [6], can be attributed to the fact that MNTI is a tumor with no envelope or multicenter growth and tumor edge invades into the bone causing difficulty in complete resection [10].

Owing to its rapid onset, alarming local growth rate, their malignant potential, and high rate of recurrence, it is essential to diagnose this type of tumor at an early stage.

<p>Fig 1. Swelling over left side of face</p> 	<p>Fig 2. Bony hard swelling over left side of palate with surface mucosa having bluish tinge</p> 
<p>Fig 3. Maxillary cross-sectional occlusal radiograph</p> 	<p>Fig 4. Computed tomography of para-nasal sinus showing approximately 4.5 × 3.7 × 5cm sized expansile bony lesion with enhancing soft tissue component and cortical thinning arising from superior alveolar arch on left side</p> 
<p>Fig 5. Magnetic resonance image showing an expansile, isointense (T1WI) lesion which is heterogeneously hyperintense compared to muscle (T2WI)</p> 	<p>Fig 6. Histopathology (H/E stain) showing melanotic pigment inside and around round cells</p> 

CONFLICTING INTEREST

No interest

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REFERENCES

1. Bangi BB and Tejasvi ML. (2012). Melanotic neuroectodermal tumor of infancy: A rare case report with differential diagnosis and review of the literature. *Contemp Clin Dent*, 3, 108-12.
2. Tam J, Cheung WS, Senger C, Reichman M, Campbell KM. (2015). Melanotic neuroectodermal tumour of infancy: A case report. *J Can Dent Assoc*, 81, f7.
3. Agarwal P, Saxena S, Kumar S, Gupta R. (2010). Melanotic neuroectodermal tumor of infancy: Presentation of a case affecting the maxilla. *J Oral Maxillofac Pathol*, 14, 29-32.
4. Lee CH, Hong SP, Lim CY, Chi JG. (1986). Melanotic neuroectodermal tumor of infancy. *J Korean Med Sci*, 1, 63-7.

5. Madrid C, Aziza J, Hlali A, Bouferrache K, Abarca M. (2010). Melanotic neuroectodermal tumour of infancy: A case report and review of the aetiopathogenic hypotheses. *Med Oral Patol Oral Cir Bucal*, 15, 739-42.
6. Souza DFM, Sendyk DI, Seo J, da Fonseca EV, Deboni MCZ. (2013). Melanotic Neuroectodermal Tumour of Infancy in the Maxilla. *Case Rep Dent.*, 20, 1-5.
7. Tandon PN, Sah K, Kale A, Kadam A, Shah H, Chandra S. (2011). Melanotic neuroectodermal tumor of infancy: Report of a case associated with high urinary excretion of vanilmandelic acid. *Contemp Clin Dent*, 2,337-41.
8. Chaudhary S, Manuja N, Ravishankar CT, Sinha A, Vijayran M, Singh M. (2014). Oral melanotic neuroectodermal tumor of infancy. *J Indian Soc Pedod Prev Dent*, 32, 71-3.
9. Hamilton S, Macrae D, Agrawal S, Matic D. (2008). Melanotic neuroectodermal tumour of infancy. *Can J PlastSurg*, 16, 41-4.
10. Cui Y, Mao Z, Liao C. (2015). Melanotic neuroectodermal tumor of infancy : A case report and review of the surgical treatment. *OncolLett*, 9, 29-34.

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