Case Report

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Oral Melanotic Neuroectodermal Tumor of Infancy: A Case Report

Sachin Kumar[1*] Dhirja Goel[2] Prabhakar Gupta[3] Rajat Gupta[4] Nitin Bhagat[5] Rohit Sharma[6] Tanu Gupta[7]

[1]Reader, Department of Oral and Maxillofacial Surgery, School of Dental Sciences, Greater Noida, India.
[2]Reader, Department of Pedodontics, School of Dental Sciences, Greater Noida, India.
[3]Senior Lecturer, Department of Oral and Maxillofacial Surgery, School of Dental Sciences, Greater Noida, India.
[4]Senior Lecturer, Department of Oral and Maxillofacial Surgery, School of Dental Sciences, Greater Noida, India.
[5]Reader, Department of Oral and Maxillofacial Surgery, School of Dental Sciences, Greater Noida, India.
[6]Senior Lecturer, Department of Oral and Maxillofacial Surgery, School of Dental Sciences, Greater Noida, India.
[7]Senior Lecturer, Department of Prosthodontics, School of Dental Sciences, Greater Noida, India.

ABSTRACT

Background: Occurrence of oral benign tumor below 6 month of age is a very rare occurring phenomenon. This case report presents a rare oral tumor in a 2 month old infant which was present in anterior maxilla and was causing difficulty in feeding. The lesion was surgically removed and histopathologically diagnosed as melanotic neuroectodermal tumor. A 6 month follow up showed no recurrence.

Keywords: Neuroectodermal tumor, infant, maxilla.

INTRODUCTION

Oral melanotic neuroectodermal tumor of infancy (MNTI) is a rare, aggressive, non ulcerative osteolytic pigmented benign tumor of jaw [1]. It is characterized by occurrence in infants below the age of six months, rapid bone loss of jaw bone involved without pain, high recurrence rate and potential for malignant transformation [2]. Historically various designations were given and now World Health Organisation (WHO) has accepted the term neuroectodermal melanotic tumor of infancy based on neuroectodermal origin of the lesion. It can occur in different parts of the body including orofacial region mainly in maxilla. Classically it presents as a soft, pigmented mass in the maxillary anterior region. In this report, we present a case of MNTI of the anterior maxilla treated by surgical excision

CASE REPORT

A 2 month old male infant came to the Department of Dental Surgery with parents complaining of an expansile growth in the front part of upper jaw. The growth was causing difficulty in feeding. The parents noticed the swelling one month ago, it was initially small in size and had rapidly increased to the present size. On clinical examination the physical growth of the infant was adequate and weight was 4kg.

The maternal, birth and family histories were not significant. On extra oral examination there was facial asymmetry and a single well defined swelling extending from 1 cm away from the corner of mouth on right side (mesially) till corner of mouth on the left side (laterally) and from ala of nose till the vermilion border superior inferiorly. On palpation the swelling was hard to confirm in consistency and non mobile. On intraoral examination: A single well defined swelling measuring 3cmX 2cm extending from 52 tooth region till 63 tooth region (mesio-distally) and from depth of labial sulcus and extending beyond alveolar margin (superior-inferiorly). The color of the swelling was whitish pink with a bluish hue. On palpation the swelling was firm to hard in consistency and non mobile [Figure 1].

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*Correspondence Dr. Sachin Kumar.
Department of Oral and Maxillofacial Surgery, School of Dental Sciences, Greater Noida, India.
Email: sachin.kumar1@sharda.ac.in

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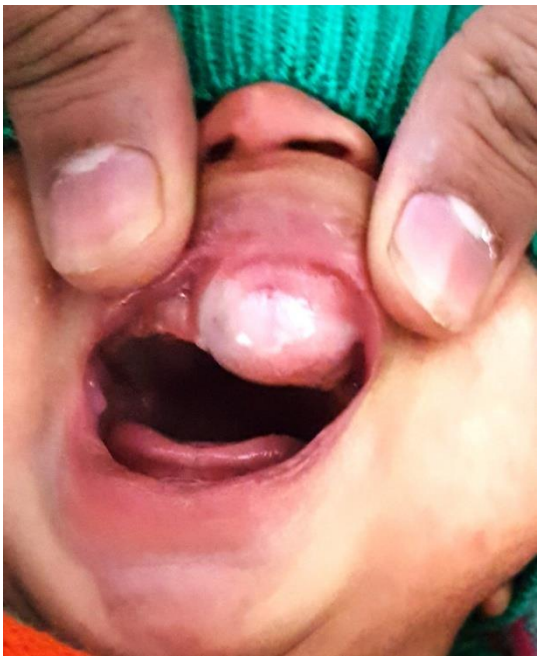

Fig 1: Intra-oral photograph showing well defined swelling.

Aspiration was negative. Based on clinical and radiographic examination a provisional diagnosis of benign tumor was made. The patient was treated under general anesthesia by wide local excision of the tumor mass and a good safety margin was provided to prevent recurrence. The gross specimen compromised of a 3cm X 2cm firm well circumscribed mass and a neonatal teeth. [Figure 2].

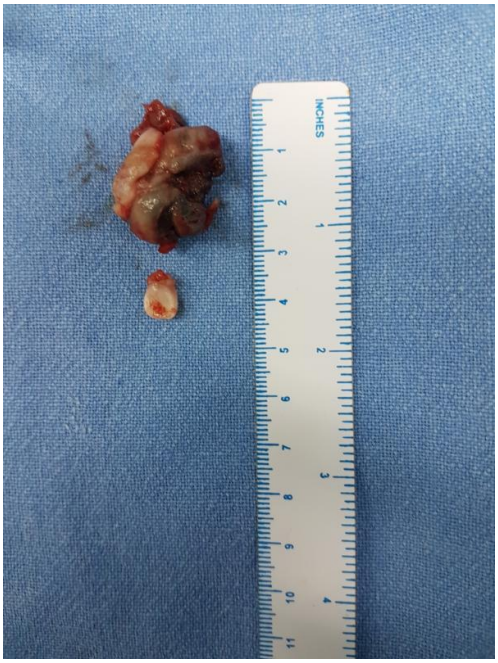

Fig 2: Gross specimen.

The cut surface was grayish white in color with black pigmented areas. The tissue was submitted for histopathological examination.

Histopathological findings:

Section showed a benign cyst lining in subdermis. Beneath the cyst a biphasic tumor was noted, composed of a large epithelioid melanin pigment containing cells with abundant cytoplasm, vesicular nuclei surrounding rest of small round cells with stippled chromatin and scant cytoplasm. Lymphovesicular emboli were also seen. A diagnosis of pigmented neuroectodermal tumor of infancy was made [Figure 3].

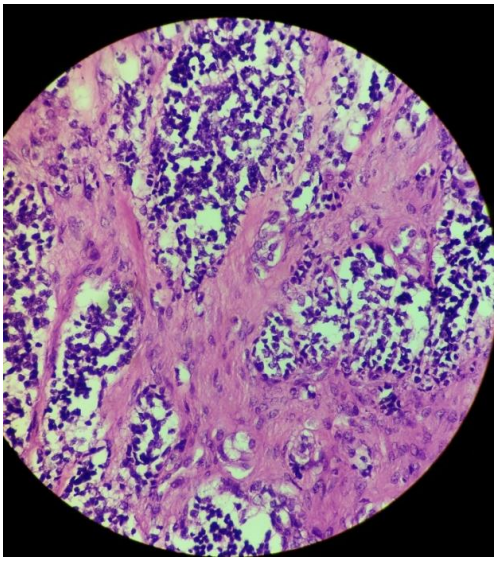

Fig 3: Histopathologic picture.

Post-operative picture [Figure 4] and X-ray [Figure 5] shows no evidence of the lesion.

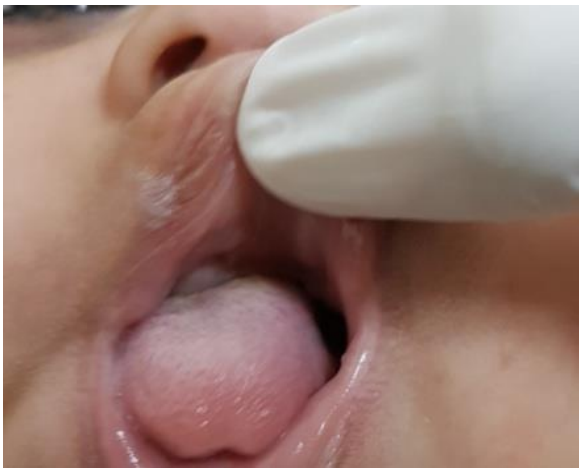

Fig 4: Post-operative picture.

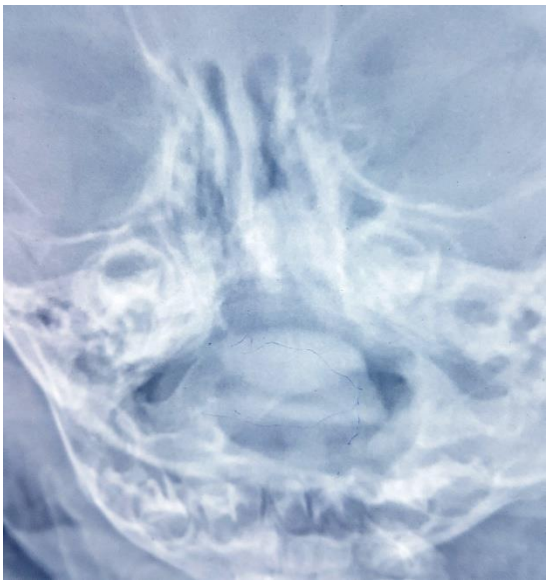

Fig 5: Postoperative radiograph with no evidence of lesion.

DISCUSSION

MNTI is a very rare benign neoplasm of infants. It was first described by Krompecher in 1918. The age of occurrence is generally between 3 to 6 months without any gender predilection. Patients with MNTI as late as 67year old had also been reported [3]. Earlier the origin was presumed of a odontogenic or retinal rests and this lead to a variety of names including retinal anlage tumor, retinoblastic teratoma , pigmented ameloblastoma, melanotic epithelial odontoma, atypical melanoblastoma, melanotic progonoma, pigmented teratoma, pigmented epulis and many other names [4]. These different names were suggested according to the doubtful etiology of the lesion in the past. Borello and Gorlin were first to suggest the neural crestal origin and coined the term " Melanotic Neuroectodermal Tumor of Infancy. Recently immunohistochemical analysis and ultrastructural studies have proved the neuronal origin of the tumor [5]. The tumor is associated with an increased urinary VMA (Vanillylmandelic acid) levels in most of the infants. In most of the cases the VMA levels are normalised after complete removal of the tumor. Also a negative rate VMA does not rule out diagnosis of MNTI because all the neural crests are not involved in the metabolism of catecholamines. The elevated values of VMA have little clinical applicability except that it can helps in diagnosis. VMA levels do not reflect the biological behaviour of the MNTI [3]. The MNTI can occur in different part of the body but most commonly it occurs in head and neck region. Anterior maxilla is the most commonly involved region. Clinical presentation includes: a painless expansile pigmented mass, commonly occur singly. Biological behavior includes a very aggressive behaviour with high reoccurence rate. The recurrence is usually seen during first 6 months. Also the tumor has potential for malignant transformation. The rate usually seen is 6.5% for all and around 2% for maxillary tumor [6]. Therefore long follow up is advised. Diagnosis of the lesion is basically based on clinical presentation during first 6 months of life and site of occurrence. Also the clinical appearance of the lesion gives a clue in diagnosis. Radiographic examination includes: traditional radiography and CT scan of the lesion. Radiographic presentation usually reveals radiolucency with or without regular margins. CT scans usually show a hyperdense mass and provides a definitive extent of the lesion and helps in surgical planning. Magnetic resonance imaging shows a hypodense mass with focal areas of hyperdensity. Histopathological findings are typical for the lesion consisting of small neuroblastic cells and large epithelial cells containing melanin. Immunohistogical markers which are positive for MNTI are HMB-45 and synaptophysin and provides confirm diagnosis of the lesion [7]. The treatment of choice is complete surgical excision with safe margins to avoid recurrence. A minimum of 5mm safe margins have been recommended by various authors [3, 4, 5]. Lesions which cannot be surgically treated such as those occurring at skull base or malignant MNTI, chemotherapy and radiotherapy are potential alternatives [5]. Differential diagnosis include: eruption cyst, hemangioma, Ewing's sarcoma, small cell malignancies (neuroblastoma, melanoma), , rhabdomyosarcoma, desmoplastic small round cell tumor peripheral neuroepithelioma, peripheral primitive neuroectodermal tumor and lymphoma [6].

The case presented in the article had all the clinical features leading to diagnosis of the lesion. Although a provisional diagnosis of an odontogenic tumor was made initially based upon clinical and radiographic findings. The confirm diagnosis was provided by the histopathology report only. Post operatively VMA urine analysis was done was it was under normal range. During surgical removal the

lesion was well defined from the palatal and labial mucosa and was involving the bone. Surgical excision was done with a safe margin of at least 5mm. Post excision the surgical site was primarily closed and post operatively healing was satisfactory. The child was under observation and during 2 year follow up there was no recurrence seen.

CONCLUSION

Occurrence of intra oral lesion involving bone in infants is a rare phenomenon. Whenever such a lesion is seen a differential diagnosis of MNTI should be included. The diagnosis of MNTI is mostly histopathological. MNTI should be treated as early as possible because of its aggressive nature and aggressive surgical treatment is better than conservative to prevent recurrence. Post operative a long follow up is recommended.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Hamilton S, Macrae D, Agrawal S, Matic D. Melanotic neuroectodermal tumour of infancy. Can J Plast Surg 2008;16:41-4.

2. Johnson RE, Scheithauer BW, Dahlin DC. Melanotic neuroectodermal tumor of infancy. A review of seven cases. Cancer 1983;52:661-6.

3. Tripuraneni SC. Melanotic Neuroectodermal Tumor of Infancy. Clin Rhinol An Int J 2012;5:138-9.

4. Singh NN, Singh M, Sreedhar G, Singhal P. Melanotic neuroectodermal tumor of infancy: Presentation of a case. SRM J Res Dent Sci 2014;5:123-6.

5.Chaudhary S, Manuja N, Ravishankar CT, Sinha A, Vijayran M, Singh M. Oral melanotic neuroectodermal tumor of infancy. J Indian Soc Pedod Prev Dent 2014;32:71-3.

6. da Fonseca MA, Thikkurissy S. Maxillary Melanotic Neuroectodermal Tumor of Infancy. Int J Clin Pediatr Dent. 2009 ;2:61-4.

7. Barrett AW, Morgan M, Ramsay AD, Farthing PM, Newman L, Speight PM. A clinicopathologic and immunohistochemical analysis of melanotic neuroectodermal tumor of infancy. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2002; 93:688-98.