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RESEARCH ARTICLE

RECURRENCE OF MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY AFTER 2 MONTHS OF COMPLETE ENUCLEATION IN A 3 MONTH OLD CHILD: A CASE REPORT

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ABSTRACT

Melanotic neuroectodermal tumour of infancy (MNTI), first reported by Krompecher in 1918, is an uncommon pigmented tumour affecting predominantly the craniofacial bones of the newborn infants [1]. Although classically benign, it is rapidly growing, locally aggressive and can follow a malignant course[2]. Infants in the first year of life are usually affected, compelling prompt diagnosis and treatment as well as close monitoring [3]. According to Krompecher, this tumor derives from epithelial nests evolved at the time of embryonic fusion of the facial processes. It has also been suggested that the tumor arises from the retinal anlage by a pinching-off process of neuroepithelium during the formation of embryonic eye[7]. Recurrences can be expected primarily because of incomplete excision, tumor dissemination during surgery, or multicentric nature . Approximately, a few hundred of these tumors have been reported in medical literature. We describe the case of a 3-month-old boy who presented with an enlarging swelling of left maxillary alveolus since 1 month of birth.

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INTRODUCTION

Melanotic neuroectodermal tumour of infancy (MNTI) is an uncommon, rapidly growing neoplasm that primarily develops in the jaws of infants. The lesion was originally described by Krompecher as a congenital melanocarcinoma[1]. MNTIs are derived from the neural crest, and appear in areas that develop from neuroectodermal pathways; the most common of these is the maxilla. Locations in other intraosseous and extraosseous structures have been characterized, including the mandible, skull, brain, and epididymis [3].Malignant behaviour has been reported in 6.5% of MNTIs (Kruse-Lösler et al.,2006) [1]. In the late 1966s, Borrello and Gorlin discovered the association between elevated levels of urinary vanillylmandelic acid in the affected infants and suggested that the lesion was of neural crest origin [4].

Histogenesis of this peculiar tumour comprises two distinct population of cells: melanin-containing large, polygonal cells resembling melanocytes; and small round blue cells [6].Neural crest cells display mesodermal and ectodermal morphologic features at different stages of their ontogeny, explaining the difficulty in deciphering the embryologic origin of these tumors and possibly explaining the biphasic cellular phenotype such tumors display [10]. These lesions enlarge rapidly ranging in size from 2cm to 5cm and may envelope developing deciduous teeth. The treatment of MNTI involves confirming histological diagnosis and surgical resection and use of adjuvant chemotherapy [9]. The clinical differential diagnosis of a rapidly growing mass of the anterior maxilla in an infant includes congenital epulis, teratoma, melanotic neurectodermal tumor, neuroblastoma, Ewing's sarcoma, rhabdomyosarcoma, melanoma, metastatic retinoblastoma, and lymphoma[8].

Although MNTI is classified as a benign lesion, it is often clinically worrisome because of its rapid onset and alarming local growth rate. The typical MNTI begins on the anterior aspect of the maxilla and rapidly expands to form a swelling or a tumescence that is cosmetically obvious to the parents of the infant [12]. Although the overall recurrence rate after surgery does not exceed to 15—20%, it has been reported to be as high as 60% after incomplete resection. In addition malignant transformation and metastases occur in fewer than 5% of cases despite the MNTI is accepted as a benign tumor. Unfortunately, it is almost impossible to predict clinically or pathologically which tumors have a more malignant phenotype at diagnosis [14].

CLINICAL REPORT: A 3-month old male infant was referred to the Department of Oral and Maxillofacial Surgery, Bharati Vidyapeeth Dental College,Pune in the month of August 2019 for treatment of a progressive swelling in the left maxillary alveolus (noted 1 month after his birth) which had been growing progressively for a duration of two months (Fig 1).

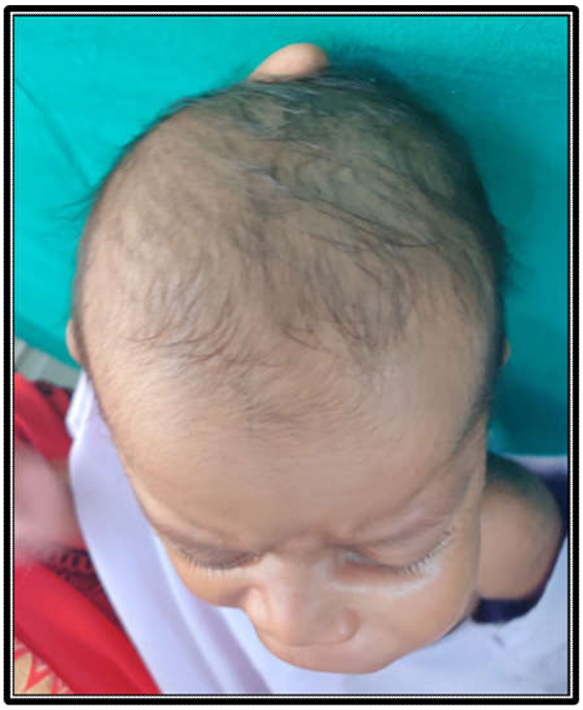

Fig 1. Extraoral view of the lesion

An extraoral examination revealed facial asymmetry, deletion of the left nasolabial folds, and elevation of the left nasal alar base [16]. The patient's medical and family history was unremarkable. There was a history of difficulty in feeding. He had no history of nasal discharge, bleeding, airway obstruction. He had no fever. Examination revealed a large and sessile mass occupying the maxillary alveolar ridge, the buccogingival sulcus with extension out of the mouth. The patient was in good general health condition [2]. On evaluation, physical examination showed a painless pale-pinkish mass involving the left maxillary alveolus extending from midline of the alveolar ridge to the right posterior portion of the maxillary alveolus (Fig 3) Extra-oral examination revealed a lesion elevating the upper lip and causing incompetence. On intraoral examination, there was a round swelling expanding the alveolar ridge, the overlying mucosa had areas of black pigmentation with no ulceration. It was firm, non-tender and the child was unable to approximate the alveolar ridges.

Fig. Profile View of Patient

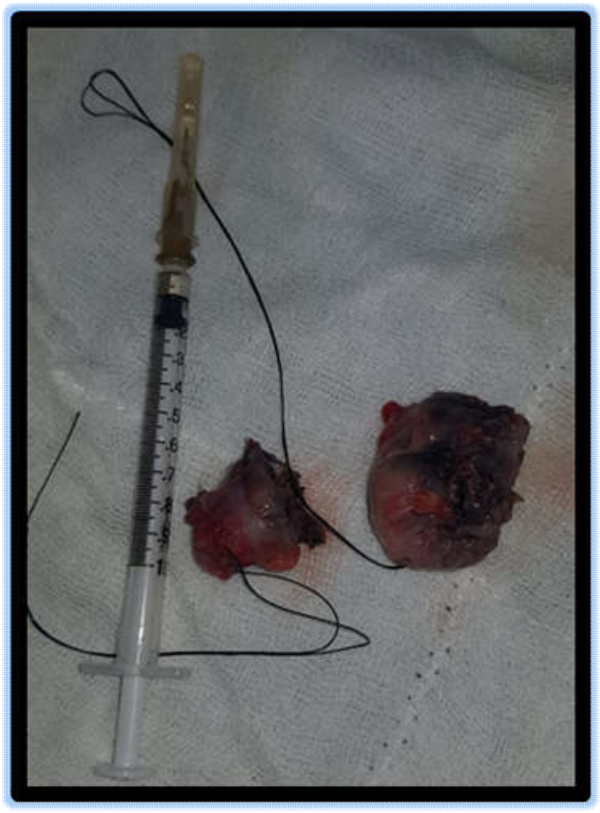

Fig 2. Excised Lesion

Histopathological sections (taken outside) showed a fibrocellular tumor covered by squamous epithelium.The stroma showed cords, clusters and nests of odontogenic epithelium embedded in a fibrous to fibro-cellular stroma with no evidence of malignancy.

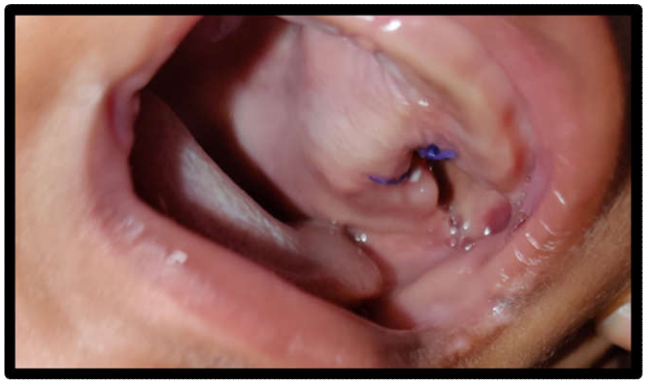

Fig. 4. Primary closure of the lesion

This was suggestive of Ameloblastic Fibroma. Computed tomography (CT) of the head and neck displayed a well-demarcated soft tissue mass [3]. Under orotracheal intubation the lesion was excised with three of the developing tooth buds in situ. Two fragments were removed one measuring 4.5 cm X 3.0 cm X 2.5 cm and the other 3.0 cm X 2.5 cm X 2.0 cm (Fig 2). The resulting defect was primarily closed (Fig 4). Microscopic examination revealed biphasic pattern of tumor cells.There were groups of small round neuroblast like cells possessing fibrillar cytoplasm surrounded by larger cells arranged in tubular and alveolar pattern and having pale nuclei with abundant cytoplasm containing melanin. Few mitotic figures were also seen .Tumor cells were surrounded by fibrous stroma.The tumor was seen invading the bone and cartilage (suggestive of Melanotic Neuroectodermal Tumor of Infancy). At two months post-operative follow-up the patient was doing well and remains under regular review [4].

DISCUSSION

Melanotic neuroectodermal tumours of infancy are usually characterized as rapidly expanding and painless tumors, invading bone and displacing adjacent tissue. There is no sex predilection, although some authors describe a tendency towards male predilection. They most frequently occur in the first 6 months of life; only 8.9% occur in patients older than 1 year [1, 2]. Lesions in the maxilla are the most common by far and are usually localized in the anterior portion. Typically, the stretched mucosa over the lesion shows a bluish tinge [2, 3] [15]. To date many histogenetic theories to explain the origin of this tumor have been proposed. This is the reason of so many names being given to the tumor. This tumor usually shows a benign course. However, there are some reports considering the tumor potentially malignant in literature because of the high recurrence rates of 50% and high metastases rates of 5—10% in these studies [6,8]. In a careful review of the literature, it was revealed that 64% of all recurrences appeared in children who were diagnosed at 12 weeks of age or younger and mostly localized in maxilla [9][14]. Conventional radiographs of bony lesions usually show a radiolucency with or without irregular margins. It is typical of the CT scans to reveal hyperdense masses, but hypodense variants have been reported as well. The CT can accurately define the extent of the lesion and thus provides a good basis for surgical planning. The histologic appearance of MNTI is unique, and a distinct biphasic pattern also exists. One portion of the lesion has large polygonal cells that contain melanin pigments that give the MNTI its blue-black clinical appearance. The cuboidal polygonal cells are at the periphery of the alveolar spaces, whereas the central portion contains the small lymphocyte-like cells [13]. The treatment of choice for MNTI is surgical excision, and it is usually curative. This treatment can usually be accomplished with a partial maxillectomy, by using a WeberFergusson incision and a facial degloving approach. Permanent reconstruction of the maxillary alveolus and missing dentition may have to be delayed until after growth is completed, often in the teenaged years. The skills of an orthodontist, prosthodontist, oral surgeon, and/or dentist may be required, based on the extent of the missing structures to correct any functional and cosmetic deformity [12]. To safeguard against leaving a microscopic tumor at the surgical margins, curettage of the cut bone has been previously advocated. It is interesting to note the extent of surgical excision. Most investigators now favor a surgical approach that is conservative via a buccogingival sulcus incision or lateral rhinotomy approach. If technically possible, complete resection with a 5-mm margin is recommended, but when radical surgery would cause mutilation, local excision with curettage of the underlying bone is usually sufficient. The most commonly reported range of recurrence varies between 10% and 20%, but recurrence rates up to 60% have been documented (9,10). Differences in treatment and the length and manner of follow-up (clinical vs radiographic surveillance) may account for the wide range of reported recurrent disease [18].The case described herein shows features typically associated with this tumor and further supports the neural crest origin theory while, at the same time, adding to the limited reports affecting the maxilla [11]. We conclude that early conservative surgical excision provides an excellent result with good prognosis for patients with melanotic neuroectodermal tumor of infancy [20].

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