Rehabilitation following surgical management of mneti: a splint in time saves nine!

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Received date: January 15, 2021; Accepted date: January 20, 2021; Published date: March 05, 2021

Citation: Kumar S, Kulkarni V, Misra CS (2021) Rehabilitation following surgical management of mneti: a splint in time saves nine! J. Surg Case Rep and Imag 4(1); DOI: 10.31579/2690-1897/060

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Abstract

Introduction: Melanotic neuroectodermal tumour of infancy (MNTI) is a rare, rapidly growing pigmented neoplasm of neural crest origin generally arising in infants during the first year of life. The rehabilitation of the anterior defect is necessary to circumvent feeding problems. The area is also a site for primary growth of maxilla and placing a hard splint would restrict the same. We have used a novel way to use a soft splint to overcome this issue with follow up and re-fabrication.

Case report: We report a 7 months old male who presented with a rapidly growing mass history of a rapidly growing mass in the upper front part of the jaw a biopsy showed melanotic neuroectodermal tumour, and complete resection with negative margins was subsequently achieved. The patient was reviewed at follow up with a magnetic resonance imaging, which showed no residual remnants. Rehabilitation was done with a soft splint to avoid restriction of maxillary growth.

Conclusion: Due to its rapid growth potential and locally destructive behaviour, early diagnosis is extremely important to limit local expansion. The treatment of choice for melanotic neuroectodermal tumour of infancy (MNTI) is surgical excision. Rehabilitation is also important with due attention to avoiding restriction maxillary growth, this was overcome by use of soft splint which has scanty been reported in literature.

Keywords: Melanotic neuroectodermal tumour of infancy; maxilla; rehabilitation

Introduction

Melanotic neuroectodermal tumor of infancy (MNTI) manifests as a very rare benign neoplasm within the first year of birth [1]. A small number of cases have been reported in older children and adults as well. This tumor grows very fast and originates in the neural creast. MNTI generally originates from the soft tissue overlying the maxilla (68–80%), but other studies support evidence of the lesion arising in the skull (10.8%), the mandible (5.8%), or brain (4.3%) [2, 3]. The predominant site although is the head and neck, other sites such as the femur, epididymis, ovaries, uterus and mediastinum have also been reported, but are seldom. Clinically, MNTI is soft and reddish-blue tumour that often destroys the underlying bone and prevents teeth development. Clinical and radiological findings can suggest a diagnosis of MNTI. The mean age of patients at diagnosis is 4.3 months, with a near equal male to female ratio of 6/7. The best choice for treatment is surgical excision. The rate of recurrence ranges from 10 to 15%, and metastases rate of 6.5% is reported in the literature [4]. The mainstay of management is surgical removal of the pathology which invariably leads to defect in the premaxilla, which in turn is a vital structure in primary growth horizontally in the first decade of development [5]. Post-surgical rehabilitation involves the use of an acrylic splint to aid in feeding. The authors felt that a rigid splint would hamper the growth of premaxilla thereby jeopardizing growth and development. Hence a novel way was devised by fabrication of soft splint to circumvent the problem.

Case report

A seven months old male child reported to our hospital with slowly growing painless mass over the anterior part of upper jaw, which made feeding the child difficult (Fig-1).
On examination, there was a non-tender painless mass over the anterior part of the maxilla with displaced upper anteriors and flaring of ala of nose. Computed Tomogram was suggestive of ovoid mass in the anterior part of hard palate abutting the nasal floor and displacing the deciduous incisors (Fig-2, 3), routine lab investigations were in normal range, along with normal levels of vanillin metallic acid (VMA). The case was taken up under general anaesthesia for complete removal of the lesion with removal of displaced anteriors (Fig-4, 5). Post operative biopsy confirmed the diagnosis of MNTI. Post-operative magnetic resonance was obtained to rule out any remnants of the lesion. The challenge faced by the clinical team was post-operative rehabilitation to aid in feeding of the patient. The presence of primary growth centre in the anterior part of the maxilla which is very necessary for the primary displacement of maxilla during growth and development would be in jeopardy if an acrylic plate were to be placed to aid in feeding. Hence it was decided to fabricate a soft split (Biostar Inc.) which would aid in feeding and speech but would also provide enough relief to aid in appropriate growth of the anterior maxilla (Fig-6). This contraption is planned to be refabricated every month to accommodate the growing maxilla. The case is planned to be taken up for fabrication of functional prosthesis till the eruption of permanent teeth.

**Discussion**

MNETI although a rare tumour, is locally regressive and may develop into a fantastic size in short time. The review conducted by Chrcanovic and Gomez [6] suggests that MNETIs were slightly more prevalent in males and were markedly more prevalent in the maxilla, and most of the lesions were asymptomatic, presenting cortical bone perforation and tooth displacement. The fact that about 10% of the lesions were observed at birth supports the hypothesis of an embryonic origin of the tumour. Some authors have suggested additional examinations to help in the diagnosis or to assist in the determination of the most suitable therapy for each case.

High levels of urinary excretion of VMA and serum AFP are characteristic of this tumour. A return of urinary VMA levels to normal after surgery was observed in some of the studies (e.g., the cases reported by Johnson et al. [7] and Howell and Cohen [8], among others). In some cases, this examination was found to be useful to monitor the patient when the level was high at the initial examination. However, it was found that in most patients, the presurgical values were normal. Although this test is indicated as a complement to the diagnosis, only a third of the patients showed high levels of urinary excretion of VMA, as was seen our case where the VMA values were absolutely normal. Concerning the treatment of MNTIs, some authors have suggested that wide surgical margins may not be necessary because tumour islets may regress spontaneously after incomplete excision. Shafer and Frissell reported that at the time of the operation it was impossible to tell whether the tumour had been completely excised, and their case was followed up for 66 months with no recurrence [9]. Marginal resection and segmental resection were found to be associated with lower recurrence rates. Although resection results in higher morbidity, the vast majority (99.2%) of the cases treated by enucleation resulted in concomitant removal of the involved teeth [1]. As both marginal and segmental resection are related to higher morbidity,
enucleation with or without complementary treatment (curettage or peripheral osteotomy) would appear to be the most indicated therapy. Hence we considered this mode of therapy.

There has been scant literature that discusses on the post-operative management of MNETI. It is prudent to consider that the premaxillary segment. Premaxilla growth is closely related to the development of human face, yet, with respect to premaxillary growth and development, it has not been defined yet the period in which the premaxilla/maxilla suture closure occurs, so that they can constitute a single bone. Growth of the premaxilla in relation to the maxilla may be correlated to malformation, such as prognathism, deep bite and protrusion. Formation of the premaxilla acts as a stabilization element inside the facial skeleton, comparable to the cornerstone of a Roman arch. In other words, premaxilla is closely related to the development of human face and its abnormal growth may be related to characteristic malformations [10]. The advocating of hard splint may cause pressure on a vital growth center thereby hindering the horizontal growth of maxilla. Hence, we in this case advocated the use of soft split to minimize the stress on premaxilla thereby aiding in growth and minimise the development of hypo plastic maxilla.

References