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Melanotic neuroectodermal tumour of infancy in the maxilla: a case report

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Summary. A 4·5-month-old Chinese boy presented with a bulging mass in the anterior right maxillary region. Clinical features and incisional biopsy examination confirmed the diagnosis of melanotic neuroectodermal tumour of infancy (MNTI). The lesion had first been noted a month before the biopsy. The extent of the lesion was defined via computed tomography, and surgical excision was accomplished through a partial maxillectomy under general anaesthesia. The patient has been irregularly followed-up over the past 8 years, and no evidence of recurrence has been found, either by clinical examination or serial computed tomography scans. A bony defect and a lack of alveolar process were noted on the right side of the anterior maxilla. The patient displayed social withdrawal at school as a result of his partial anodontia. There have only been a limited number of reports on the dental rehabilitation of postoperative MNTI. We report a case of maxillary MNTI in which the need for dental rehabilitation was emphasized.

Introduction

Melanotic neuroectodermal tumour of infancy (MNTI) is a rare neoplasm found mainly in children under one year of age. This lesion is generally accepted to be of neural crest origin. The lesion is most common in the anterior region of the maxilla, but it also can occur at other locations, including the skull, mandible, brain and epididymis [1,2]. Clinically, MNTIs are soft and rapidly growing pigmented swellings. They often destroy the underlying bone and may be associated with displacement of developing teeth. Clinical and radiological findings may suggest a diagnosis of MNTI, but histopathological examination is required for a definitive diagnosis. Melanotic neuroectodermal tumour of infancy generally follows a benign course, but the rate of recurrence is reported to be 15% within one year of the initial excision [2]. Most cases of recurrence can be cured by repeated surgery. Although extremely rare, malignant transformation has occasionally been reported [3,4]. Moreover, there have been only a limited number of reports on the dental rehabilitation of MNTI [5]. We report an additional case of maxillary MNTI in which the need for dental rehabilitation was emphasized.

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Case report

A 4·5-month-old Chinese boy initially presented at Veterans General Hospital, Kaohsiung, Taiwan, with a bulging mass in the anterior right maxillary region in March 1993. A biopsy of the lesion had already been performed at another institution. The initial diagnosis of MNTI was confirmed histologically. The lesion had first been noted a month before the biopsy by the boy's family, who reported progressive enlargement and difficulty in feeding. The patient's medical and family history was unremarkable.

Extra-oral examination revealed facial asymmetry with a firm fullness on the right cheek and raised alae on the nose. Intra-oral examination revealed a large, firm swelling fixed to the right maxilla. The lesion occupied the right maxillary alveolar ridge and right gingivo-buccal sulcus, and extended into

Fig. 1. Preoperative view: Note the firm fullness of the right cheek and the raised alae on the nose. Easy bleeding upon contact was noted.

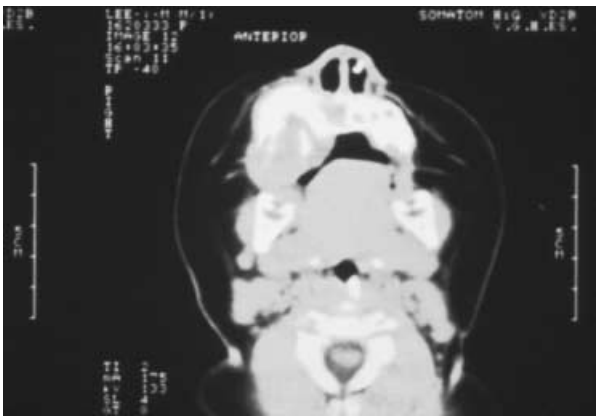

Fig. 2. Computed tomography scan showing a soft-tissue mass arising from the right maxillary bone with bony destruction, extending upward to the infra-temporal fossa.

the hard palate (Fig. 1). The lesion was not tender, and did not have a thrill or pulse. However, easy bleeding was noted upon contact.

Preoperative imaging studies included plain radiographs of the skull (postero-anterior and lateral projections), and computed tomography (CT) of the neck and larynx with contrast. The skull radiographs revealed a relative blurring of the bony structure of the right maxillary sinus. The CT scans demonstrated a soft-tissue mass arising from the right maxillary bone with bony destruction, extending upward to the infra-temporal fossa (Fig. 2). No encroachment on the nasal cavity was found. Neck lymphadenopathy was also noted.

Surgical excision was accomplished by a partial maxillectomy under general anaesthesia. The tumour

Fig. 3. Cut surface of the tumour showing black pigmentation surrounded by grayish-white fibrotic tissue.

appeared encapsulated and was covered by a layer of mucosal epithelium. Easy bleeding was noted on contact. The tumour did not invade the nasal cavity or the orbital floor. Several tooth germs were encountered and sacrificed. The gross specimen comprised a well-circumscribed soft mass, measuring $4{\cdot}5 \times 3 \times 2{\cdot}5$ cm, with a firm and rubbery consistency. The cut surface was greyish white in colour with marked black pigmentation (Fig. 3). Microscopically, the tumour comprised a moderately fibro-vascular stroma with nests and clusters of cells arranged in an alveolar pattern. The cells in the central parts of the alveoli were small and round with little cytoplasm and dark nuclei, whilst those at the periphery were cuboidal or flattened, and some contained cytoplasmic melanin (Fig. 4). Special stains for melanin were positive. Melanotic neuroectodermal tumour of infancy was diagnosed.

The patient's recovery was uneventful and healing progressed satisfactorily. The patient has been irregularly followed-up for 8 years by the Section of Paediatric Surgery, and the Section of Oral and Maxillofacial Surgery at Veterans General Hospital, Kaohsiung. No recurrence of the lesion has been noted, as determined by clinical examination and serial CT scans.

The patient was referred to the Section of Paediatric Dentistry in May 2001 (at 8·5 years of age) for evaluation and dental rehabilitation. At the start of prosthetic treatment, extra-oral and cephalometric radiographic examinations revealed a concave facial profile with deficiency of the mid-face. Facial asymmetry with the chin deviating to the left was also noted on extra-oral examination. An intra-oral examination

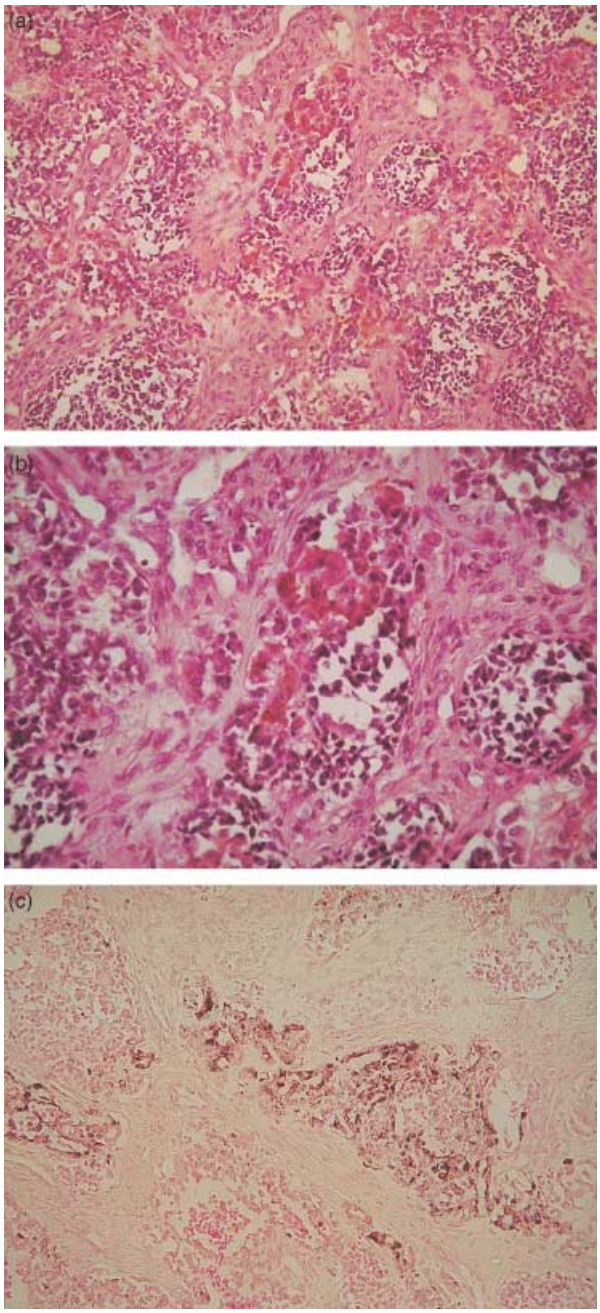

Fig. 4. Photomicrograph of an excised specimen: (a) Nests and clusters of cells arranged in an alveolar pattern (H&E, × 100). (b) Two cell types, i.e. small, hyperchromatic round cells and larger, epitheloid cells with vesicular nuclei. Some stippled melanin pigment is also present (H&E, × 200). (c) Large epitheloid cells containing melanin granules (Masson–Fontana stain, × 100).

showed the child to be in the mixed dentition stage with anterior cross-bite. A panoramic radiograph showed the absence of all primary and permanent teeth anterior to the permanent first molar in the right maxillary quadrant (Fig. 5). The upper dental mid-line deviated significantly to the right as a result of mesial drifting of the left maxillary incisors. The bony defect and lack of alveolar process on the right side of the anterior maxilla resulting from the surgical excision 8 years previously was also noted (Fig. 6). Since the patient was still growing, and also for economic reasons, a removable maxillary prosthesis has been used to improve aesthetics, and orthodontic treatment, bone grafting and implant placement have been delayed. Three wrought-wire clasps were used around bilateral permanent first molars and the left permanent central incisor for retention (Fig. 7). The removable prosthesis represents an interim treatment for improving psychosocial function and may require

Fig. 5. Postoperative panoramic radiograph (taken 8 years after surgery). Note the absence of all primary and permanent teeth anterior to the permanent first molar in the right maxillary quadrant.

Fig. 6. Postoperative intra-oral view (taken 8 years after surgery). Note the resulting bony defect and a lack of alveolar process on the right side of the anterior maxilla.

Fig. 7. Intra-oral photograph showing a removable partial prosthesis with three wrought-wire clasps around the bilateral permanent first molars and the left permanent central incisor for retention.

several revisions before the patient can achieve dental and skeletal maturation, and more definitive restorative treatment be provided.

Discussion

A total of 195 MNTI cases have been reported [1,2]. The most common sites of origin were the maxilla (61%), skull (13%), mandible (7%), brain (6%) and epididymis (5·8%). Surgical excision has been the typical treatment for MNTI, but individuals with MNTI which has not been amenable to surgical management have received chemotherapy, chemotherapy with radiation therapy and radiation therapy [6]. Computed tomography can accurately define the extent of the lesion, and this greatly assists surgical planning [7]. Wide excision with a resulting large defect and partial anodontia in the maxillofacial region remains a significant concern for both the growing patient and the dentist. Although they may be considered, the use of implants in growing patients is particularly controversial [8–11]. Implant use should be carefully considered in partially dentate children because submergence of the implant may occur [10]. However, some form of treatment is needed. A large bony defect, lack of alveolar process on the right side of the anterior maxilla and a deficiency of mid-face were obvious in the patient reported here and a cause of social difficulty. A conventional removable prosthesis without bone grafting and orthodontic treatment was used for this patient. Poor retention, frequent adjustment and repair, and occasional remaking of the conventional prosthesis in response to growth and development are potential problems associated with a conventional removable prosthesis. However, the use of a conventional prosthesis to meet the immediate psychological and functional requirements of growing patients is highly recommended as an interim measure [8]. Whilst the prosthesis is in use, the definitive treatment plan, including dental implants, orthognathic surgery and orthodontic treatment may be constantly reassessed and improved so that it may be more effectively carried out when growth is at or near completion.

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Résumé. Un garçon chinois de 4 ans $^1/_2$ présentait une masse bombée dans la région maxillaire antérieure droite. Les caractéristiques cliniques et l'examen de la biopsie ont confirmé le diagnostic de tumeur neurectodermique de l'enfant (MNTI). La lésion avait été diagnostiquée dans un premier temps un mois avant la biopsie. L'étendue de la tumeur a été définie par tomographie numérisée, et une excision chirurgicale a été effectuée par maxillectomie partielle sous anesthésie générale. Le patient a été suivi irrégulièrement durant les huit dernières années, sans récurrence détectée que ce soit à l'examen clinique ou lors de scanners de tomographie numérisée sériée. Un défaut osseux et un manque de procès alvéolaire ont été notés du côté droit du maxillaire antérieur. Le patient présentait à l'école un repli sur lui du fait de son anodontie partielle. Peu de cas ont été décrits de réhabilitation dentaire en post-opératoire de MNTI. Nous décrivons un cas de MNTI maxillaire dans lequel l'accent est mis sur le besoin de réhabilitation dentaire.

Zusammenfassung. Eine Fall eines $4^1/_2$ jährigen chinesischen Jungen mit einer Auftreibung des rechten vorderen Oberkiefers wird vorgestellt. Klinische Symptome und eine histologische Absicherung ergaben eine Diagnose eines infantilen melanotischen neuroektodermalen Tumors (MNTI). Ein Monat vor Biopsie wurden die ersten Veränderungen bemerkt. Eine Computertomographie zur Bestimmung der Tumorausdehnung wurde durchgeführt, In Vollnarkose wurde eine partielle Resektion des Oberkiefers

durchgeführt. Der Patient wurde unregelmäßig zu Nachuntersuchungen vorgestellt, während 8 Jahren zeigte sich kein Hinweis auf ein Rezidiv, weder klinisch noch im CT. Ein Defekt an Knochen und Alveolarfortsatz im Operationsgebiet blieb bestehen. Die teilweise Zahnlosigkeit führte zu Nachteilen bei Sozialkontakten in der Schule. Es gibt nur wenig Literatur zur Rehabilitation nach MNTI-Operation. Der vorliegende Fall verdeutlicht die Relevanz der Rehabilitation bei MNTI-Operationsresiduen.

Resumen. Un niño chino de $4^1/_2$ meses se presentó con una masa tumoral en la región anterior maxilar derecha. Los signos clínicos y el examen de biopsia incisional confirmaron el diagnóstico de tumor melanótico neuroectodérmico de la infancia (TMNI). La lesión se había apercibido un mes antes de la biposia. La extensión de la lesión se definió por tomografía computarizada y la escisión quirúrgica se realizó a través de una maxilectomía parcial bajo anestesia general. El paciente se siguió de forma irregular durante los ocho años anteriores y no se encontró ninguna evidencia de recurrencia, tanto por el examen clínico como por los exámenes con escáner de tomografía computarizada seriada. Se notaba en el lado derecho del maxilar anterior un defecto óseo y la falta de proceso alveolar. El paciente encontró rechazo social en la escuela como resultado de su anodoncia parcial. Ha habido un limitado número de informes sobre rehabilitación dental en el postoperatorio de TNMI. Informamos de un caso de TNMI maxilar en que se enfatiza la necesidad de una rehabilitación dental.

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