ORIGINAL RESEARCH PAPER

Plastic Surgery

MELANOCYTIC NEUROENDOCRINE TUMOUR OF INFANCY, A RARE PRESENTATION OF MAXILLARY SWELLING.

KEY WORDS:
Neuroectodermal, Melanotic, maxillary neoplasm.

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ABSTRACT

Melanotic neuroectodermal tumour of infancy (MNTI) is rare, rapidly growing, pigmented neoplasm of neural crest origin. It is generally accepted as a benign tumour despite its rapid and locally destructive growth. It primarily affects the maxilla of infants during the first year of life. Surgical excision is considered as the treatment of choice. The recurrence rate varies between 10-15% and malignant behaviour has been reported in 6.5% of cases. We report a case of MNTI, associated with an erupted primary tooth in a 3 month old female child. We discuss the clinical, radiographic and histologic features of this rare tumour, as well as its surgical management and follow-up.

INTRODUCTION:

The melanotic neuroectodermal tumour of infancy (MNTI) is a rare, pigmented neoplasm of neural crest origin occurring in the first year of life.[1] It was first described by Krompecher in 1918, who named it congenital melanocarcinoma.[2] More than 90% of MNTIs occur in the head and neck region, maxilla being the most commonly affected site followed by the skull, mandible and brain.[3]

A variety of nomenclatures have been used depending on the origin of this tumor such as Congenital melanocarcinoma, retinal angle tumor, pigmented congenital epulis, or melanotic progonoma. In 1966, Borello and Gorlin identified elevated levels of urine vanillylmandelic acid (VMA), a marker of neurogenic tumors, in MNTI patients.[4]

The treatment of choice is surgical excision. Because of its high recurrence rate, cases of MNTI should be monitored closely during the post resection period. Although locally invasive, the risk of tumor metastasis is 6.5%. The benefits of adjuvant chemotherapy and radiation therapy to prevent recurrence of the tumor are unproven.[5]

We report a case of MNTI, associated with an erupted primary tooth in a 3 month old female child. We discuss the clinical, radiographic and histologic features of this rare tumour, as well as its surgical management and the follow-up.

CASE REPORT:

A 3-month-old girl presented with swelling of the maxillary anterior alveolar ridge for 4 weeks, causing feeding difficulties. There were no congenital anomalies at birth. On clinical examination, there was superior displacement of the left paranasal region and of the left upper lip. Intraoral examination revealed a firm, reddish-blue mass measuring approximately 4cm, from the left alveolar ridge, covered by an intact mucosa. (Figure 1) Needle aspiration produced negative results. The computed tomography scan showed a well-circumscribed osteolytic expansive mass in the left anterior maxilla causing expansion and destruction of the buccal cortical bone and involvement of the primary deciduous incisive tooth (Figure 2).

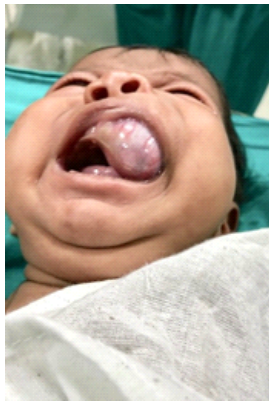

Figure 1: superior displacement of the left paranasal region and of the left upper lip. Intraoral examination revealed a firm, reddish-blue mass measuring approximately 4cm, from the left alveolar ridge, covered by an intact mucosa.

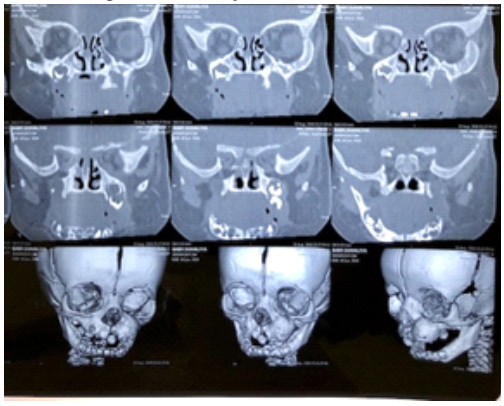

Figure 2: The computed tomography scan showed a well-circumscribed osteolytic expansive mass in the left anterior maxilla causing expansion and destruction of the buccal cortical bone and involvement of the primary deciduous incisive tooth

The diagnoses of dentigerous cyst, adenomatoid odontogenic tumor, MNTI or rhabdomyosarcoma were suggested based on the clinical and imaging findings. For the definitive diagnosis, the patient underwent an incisional biopsy in the operating room. Microscopically, the lesion showed biphasic proliferation

of small rounded neuroblast-like cells, and epithelioid cells with eosinophilic cytoplasm containing variable amounts of melanin.(Figure 3) No features of malignancy were observed. Immunohistochemically, the melanocyte- like component was strongly and diffusely positive for HMB-45.(Figure 4)

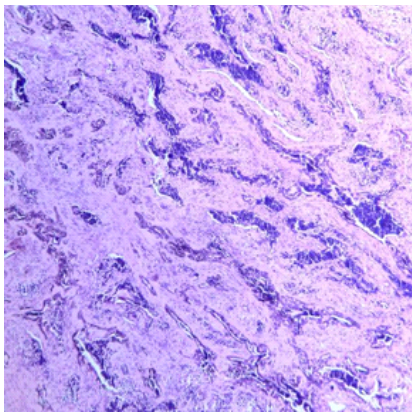

Figure 3: Microscopically on H & E stain, the lesion showed biphasic proliferation of small rounded neuroblast-like cells, and epithelioid cells with eosinophilic cytoplasm containing variable amounts of melanin.

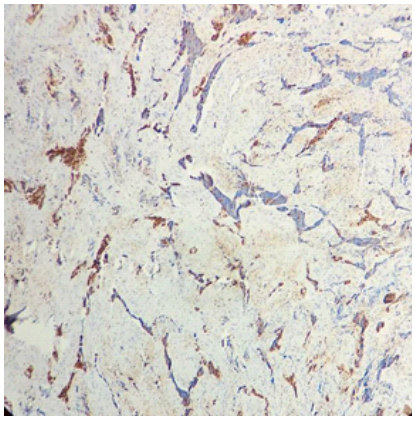

Figure 4: On immunuhistology with HMB- 45 stain, the melanocyte like component was strongly and diffusely positive.

Thereafter, the lesion was easily enucleated and the bony cavity was carefully curetted and washed. The primary (deciduous) incisor was extracted since it was displaced buccally and had no osseous support. (Figure 5) No postoperative morbidity was observed. Microscopic examination of the surgical specimen confirmed the diagnosis of MNTI.

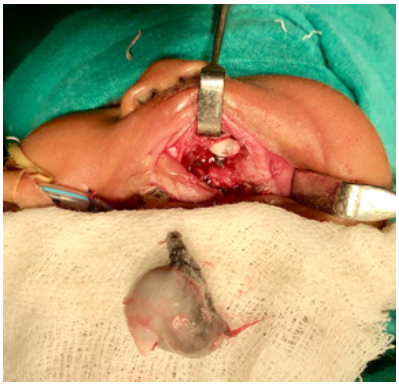

Figure 5: The lesion was easily enucleated and the bony cavity was carefully curetted and washed. The primary (deciduous) incisor was extracted since it was displaced buccally and had no osseous support.

Discussion:

MNTI was first described by Krompecher in 1918; he called it congenital melanocarcinoma. Since then, there has been uncertainty about its cellular origin. [6,7,8] The presence of Vanillylmandellic acid is pathognomonic for the neural crest component.[2]

Up to 1990, roughly 200 cases had been reported in the literature; that has increased to roughly 486 cases reported to date. [2,9]The lesion has a slight male predilection and an affinity for the head and neck. MNTI most commonly arises in the anterior maxilla (69%), skull (11%), mandible (6%), the upper limb, thigh and epididymis.[2]

Recurrences are seen in 10%-15% cases; however, metastasis is rare. It is a rapidly expansile tumour with tendency to invade local structures, and presenting with a short clinical history. If diagnosed early, it can be cured by complete resection. Aggressive chemotherapy and radiotherapy required for other paediatric small round cell tumours are not indicated in patients with completely excised tumours, thus avoiding their significant side effects in these young patients. An accurate and early pre-operative diagnosis by FNAC, thus facilitates appropriate management of these patients.[10]

Mummery and Pitts reported a case of pigmented maxillary tumor in a 3 month old baby girl that arose from aberration of dental epithelium and was termed as melanotic epithelial odontoma. Hal pert and Patzer (1947) reported a similar tumor that contained pigmented epithelium and was suggestive of ciliary body of the eye. Misugs and colleagues (1965) suggested that the growth is derived from the neural crest. Borello and Gorlin reported a case of MNTI in the maxilla in a 3-month-old boy where there was increased excretion of VMA, which returned to normal after the tumor was removed.

Histopathologically, the MNTI is similar to other tumors that contain small round cells. Clinically, the differential diagnosis of MNTI includes congenital epulis, neuroblastoma, Ewing´s sarcoma, rhabdomyosarcoma, eruption cyst, Burkitt lymphoma, Langerhans cell histiocytosis and hemangioma. All these lesions are able to generate oral manifestations as swelling in the anterior maxilla.[3]

Radiographic exams, such as CT scan, can contribute to diagnosis, in addition to providing relevant information for surgical planning. Intraosseous MNTI lesions are commonly characterized by a well- circumscribed hypodense mass, and advanced stage tumors show excessive bone destruction. The present case showed a well-defined, unilocular, osteolytic lesion causing expansion and destruction of the maxillary cortical bone.[4]

Conventional radiographs of bony lesions usually reveal radiolucency with or without irregular margins. CT scan reveals hyperdense masses, but hypodense variants have also been reported. CT can accurately define lesion extent and provide a good basis for surgical planning. Melanotic neuroectodermal tumors of infancy tend to be isointense on T1-weighted images and slightly hyperintense on T2-weighted images. Furthermore, there may be an inhomogeneous increase in signal on T1-weighted images, which also may reflect intratumoral melanin, whereas T2-weighted images visualize the tumor as isointense or hypointense to gray matter. Contrast enhancement is usually quite marked.[5]

The primary treatment is surgical resection, although there are examples of MNTI treatment using chemotherapy alone. However, it is generally agreed that it 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.[7]

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