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Mandibular Melanotic Prognoma: About A Case

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Abstract **Case Report**

Melanotic neuroectodermal tumor is a rare tumor, usually occurring in children under one year of age. The tumor is generally benign, locally aggressive and fast-growing. Mandibular localization is rare. Prognosis depends on complete surgical intervention, and remains favorable. Malignant transformation may occur, but remains exceptional. Long-term monitoring is recommended. In the light of an observation of melanotic progonoma in a 4-month-old infant at mandibular level.

Keywords: Melanotic progonoma, mandible, tumor.

INTRODUCTION

Melanotic neuroectodermal tumor of childhood (MNTI), is a rare locally aggressive tumor appearing most often in the maxilla during infancy. This tumor rarely appears in the mandible and cranial vault with significant intracranial extension.

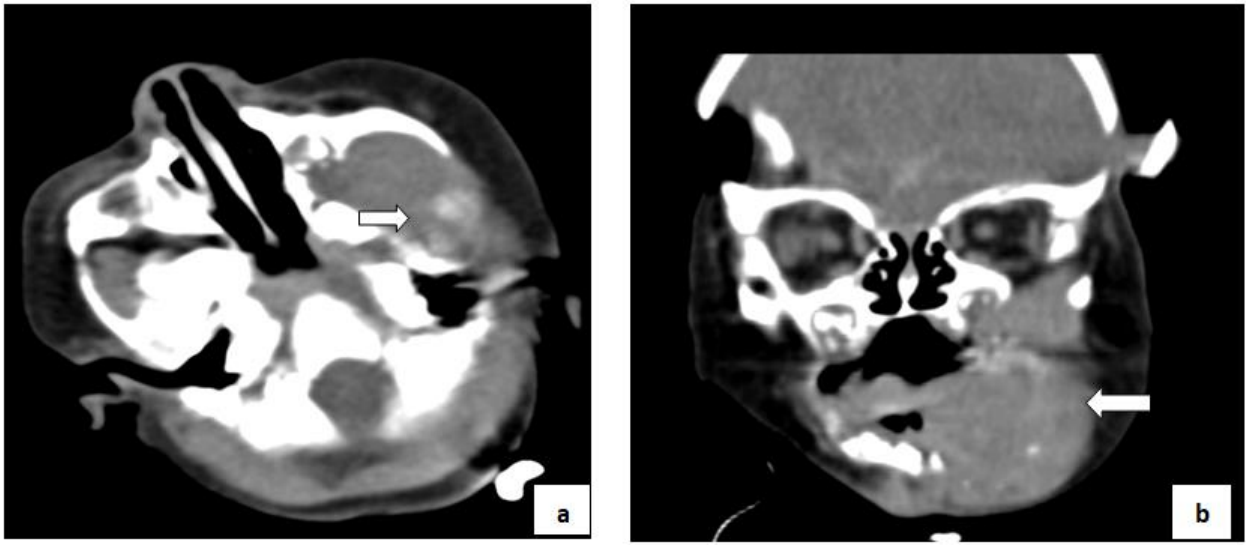
We present a case of mandibular melanotic prognoma in children and review the literature.

CASE REPORT

A 4-month-old female infant was admitted with a history of painless swelling of the left cheek since birth that progressively increased in size preventing the baby from suckling.

On examination, she presented with a large tumour approximately 3 cm in diameter occupying the left mandible and protruding into the oral cavity. The swelling appeared firm, was not warm to the touch and was neither tender nor mobile.

The facial CT scan showed a mixed bone lesional process of the left hemi-mandible with cortical rupture with grass-fire periosteal reaction.



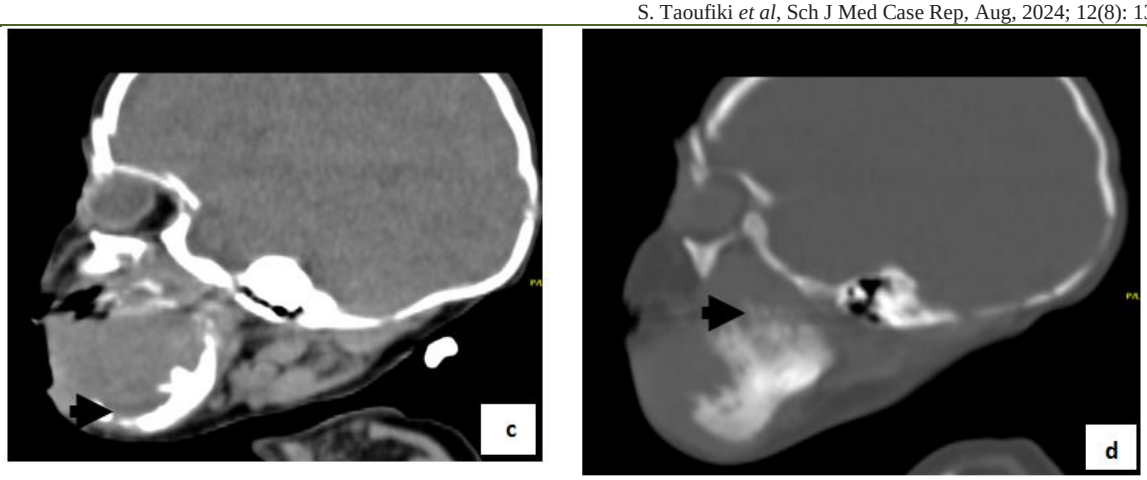


Figure 1: Facial CT scan axial, coronal and sagittal sections bone and soft tissue window illustrating a lesional process of the left hemi-mandible (a, b) (arrows) with cortical fracture and grass fire periosteal reaction (c, d) (arrowheads)

Histological study revealed reworked and ulcerated fibrous tissue infiltrated by dense cell clusters with brownish pigmented cytoplasm.

The patient underwent surgery and local excision. Recovery was uneventful. The patient is under close follow-up with proposed bone grafting. Facial appearance has improved markedly.

DISCUSSION

Infantile melanotic neuroectodermal tumors (IMNT), or melanotic progonomas, are rare and mostly benign tumors of the young child [1]. Ninety-two percent of cases occur in children under one year of age [3]. MNTIs are mainly located in the cervico-facial region and cranial vault, but many other sites have been described (testis, epididymis, mediastinum, femur, soft tissue, brain, etc.).

The nature of these lesions is uncertain, and they present both the characteristics of a hamartoma and a true tumor, the histogenesis of which is doubtful. The hypothesis that these pigmented neoplasms originate from the neural crest is supported by ultrastructural studies and histochemical and biochemical evidence of the secretion of catecholamine-related hormones [4, 5]. The cranial CT showed hyperdense and osteolytic tumor. At magnetic resonance imaging (MRI), it was hypo-intense on T1 **T2:** hyperintense to muscle, **T1 C+ (Gd):** enhancement is usually present but can be difficult to assess if very T1 hyperintense. Imaging does not confirm the diagnosis; however it allows to evaluate the intracranial extension [6].

The differential diagnosis of MNTI involves other pediatric "small round cell" neoplasms. These include neuroblastoma, Ewing's sarcoma, rhabdomyosarcoma, peripheral neuroepithelioma, desmoplastic small round cell tumor, malignant melanoma, peripheral primitive neuroectodermal tumor, and lymphoma [7].

Surgical excision is the typical treatment for progonoma, that are not amenable to surgical management alone may receive other modes of treatment.102 In general, this may be chemotherapy alone,106 chemotherapy with radiotherapy, chemotherapy before or after surgical treatment,38,46,64,80,102 radiotherapy and surgical treatment, 8 or a combination of all [8-12].

This lesion rapidly progresses to the oral cavity and facial mass, leading to deformation of the lateronasal region. The vast majority of children are cured by local resection, without the need for radiotherapy or chemotherapy.

CONCLUSION

Mandibular melanotic prognoma is a rare tumor of early age. The prognosis depends on a complete surgical procedure, and remains favorable.

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