Chronicling a Case of Melanotic Neuroectodermal Tumor of Infancy in a Pediatric Patient

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BACKGROUND

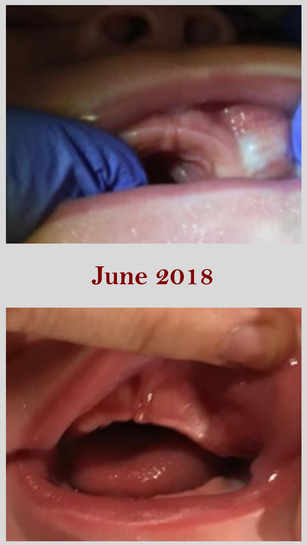
A 4-month-old white female patient initially presented to the Emergency Department at Riley Hospital in June of 2018 after her mother noticed a rapidly growing mass on her left maxilla. Her medical history at the time was essentially negative. Computed tomography (CT) radiographic imaging was acquired, revealing an abnormal, smooth-walled, expansile lesion involving the left maxillary alveolar ridge containing the unerupted maxillary left primary canine. A biopsy was performed under general anesthesia and the pathology report suggested a melanotic neuroectodermal tumor. The tumor was resected under general anesthesia just several days after the initial visit and submitted for pathological evaluation. In October of 2021, the patient returned to Riley Hospital Dental Clinic with chronic pain.

CLINICAL PRESENTATION

The initial description of the lesion was characterized as a non-fluctuant, firm mass measuring 3 mm across in its greatest dimension located on the left maxilla, between the alveolar ridge and buccal vestibule. The initial consultation with the on-call pediatric dentistry resident recommended a urine test for vanillylmandelic acid, which might suggest the lesion is of neuroectodermal origin.[1] Additionally, a complete blood count, serum calcium, and serum alkaline phosphatase labs were ordered. All lab results yielded no remarkable findings. The patient's mother had noted rapid growth of the lesion, reporting it nearly tripled in size in just a matter of days, which prompted a visit to the Riley Hospital Emergency Department and ultimately resulted in the patient being admitted for further workup.

BIOPSY and PATHOLOGY REPORT

An excisional biopsy was performed at Riley Hospital under general anesthesia by an attending oral and maxillofacial surgeon. The differential diagnosis included, along with melanotic neuroectodermal tumor of infancy, rhabdomyosarcoma, neuroblastoma, and Ewing sarcoma. A pathology report was completed from the Department of Pediatric Pathology at Riley Hospital. The pathology report stated "Histologic sections show portions of tooth and bone with associated fibrous connective tissue which shows a cellular proliferation consisting of poorly differentiated crushed small round blue cells and nests of more epithelioid-appearing tumor cells and associated pigment [...] The biphasic appearance of the tumor showing areas of pigmentation, with immunohistochemical staining with neuroendocrine markers and also with keratins and HMB45 are consistent with melanotic neuroectodermal tumor of infancy."



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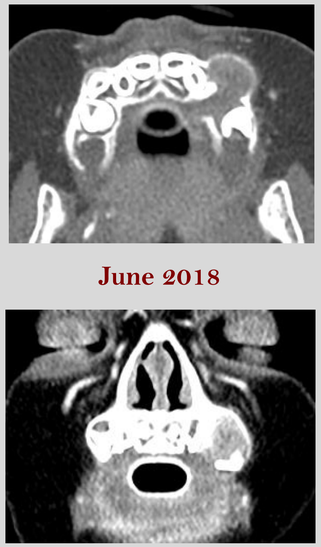

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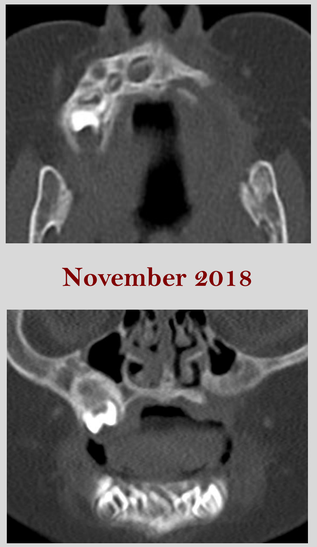

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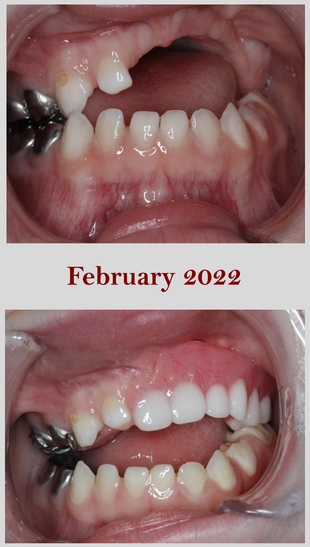

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DEFINITIVE TREATMENT and DIAGNOSIS

The confirmed diagnosis of melanotic neuroectodermal tumor of infancy warrants complete surgical excision.[2] Surgical removal under general anesthesia was completed which entailed excising the lesion and completing a peripheral ostectomy to ensure clear margins. Additionally, developing #I and F were located and determined to have been affected by the tumor spread and were also removed. All specimens were placed in separate containers containing 10 percent neutral buffered formalin solution and submitted to Riley Hospital Pediatric Pathology for definitive diagnosis. Receipt of the final pathology report confirms the lesion as a 1.5 cm melanotic neuroectodermal tumor of infancy. Proper surgical resection of MNTI is typically definitive, however local recurrence has been observed in 10-15% of documented cases."[3]

FOLLOW UP CARE

The patient had been carefully followed by several pediatric specialists, including a speech therapist, otolaryngologist, and nutritionist over the next 3 years. In October of 2021 at a return visit, the patient's mother reported a history of chronic pain that arises spontaneously and prevents sleeping. The onset of pain also triggers localized erythema along the maxillary branch of the left trigeminal nerve. Upon clinical examination, the area of tumor resection had healed, although #E, F, G, H, I, and J are all absent. Additional restorative work had also been completed by a local private-practice pediatric dentist. A maxillary acrylic-base partial denture was fabricated, and a prescription for Neurontin (gabapentin) was provided for short-term management of neuropathic pain. The patient's mother was encouraged to consult with an orofacial pain specialist for long-term solutions to neuropathic pain and familiarize her with wearing the fabricated prosthesis. Multiple follow up visits were required to make necessary adjustments to the patient's prosthesis and determine the correct dose of calcium channel blocker. A dose of 7.5 mg/kg given orally twice daily was determined to provide maximal analgesia while limiting undesired side effects.

REFERENCES

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