MNTI: Case Report of a Rare Neoplasm

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Background

Melanotic neuroectodermal tumor of infancy (MNTI) is an extremely rare neoplasm arising from the neural crest cells. It typically presents in the head and neck region in patients less than 1 years of age. Although it is considered a benign lesion, the lesion is often locally aggressive and can mimic a malignancy. Additionally, there are reports of metastatic spread and mortality associated with the tumor. Radiographs and lab values may be essential in diagnosis. Vanillylmandelic acid (VMA) levels of urine are often elevated due to a hypothesis of the tumor being of neural crest origin.

Case Description

We present a case report of a 16-week-old male who presented with a chief complaint of “a bump on his front lip”. Upon clinical examination, a ~3 cm mass was observed in the right maxilla associated with local bony destruction. The patient’s mother denied any symptoms including pain or discomfort. No drainage or purulence was noted over the last 2-3 weeks it had been present. Parents denied any issues with feeding or weight gain.

A maxillary occlusal film was exposed and revealed a large diffuse lesion, with “floating teeth”. Due to findings upon clinical exam, urine samples to measure VMA levels were taken to aid in diagnosis. A proper referral was made to Geisinger oral maxillofacial surgery for further evaluation and treatment. The patient was treated with surgical excision after the appropriate workup was completed and is currently being followed clinically for recurrence.

Differential Dx

1) MNTI
2) Osteosarcoma
3) Neuroblastoma
4) Alveolar Rhabdomyosarcoma
5) Ewing Sarcoma
6) Eruption Cyst

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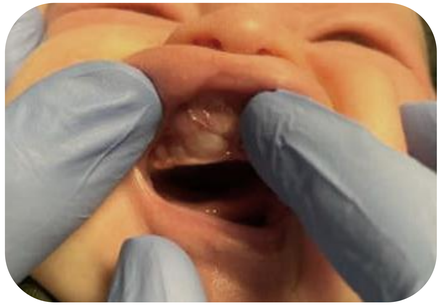

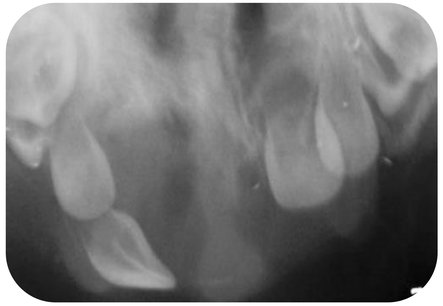

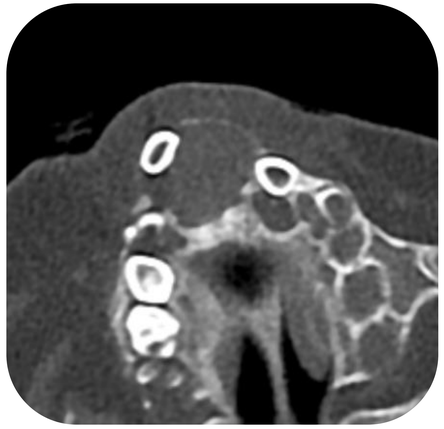

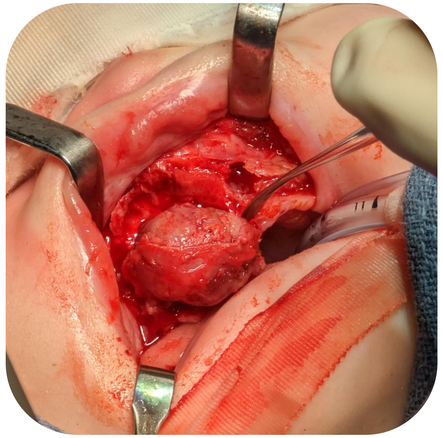

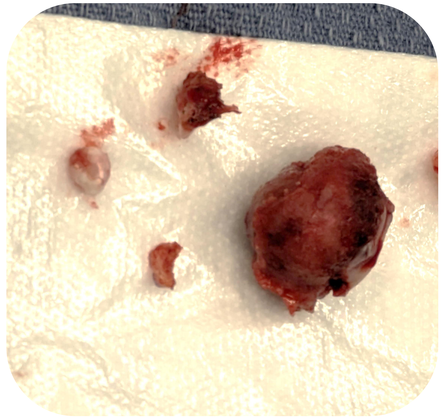

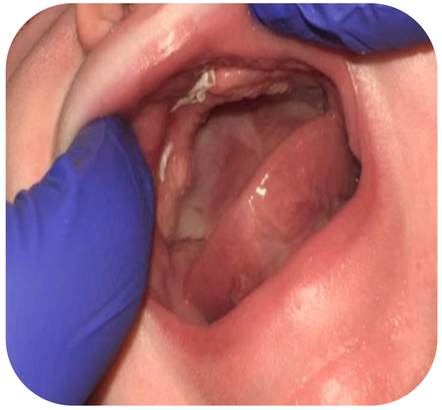

Treatment

For purposes of proper diagnosis, and incisional biopsy was completed. Final pathology report included “nests of tumor cells with biphasic cell population consisting of smaller cells with round, hyperchromatic nuclei and scant cytoplasm and larger cells with round to oval vesicular nuclei, prominent nucleoli and moderate eosinophilic cytoplasm.”

Patient was returned to the operating room for surgical enucleation of the lesion. Once the lesion was accessed, soft tissue and bony margins were outlined and included in enucleation to ensure full debridement of the lesion. Tooth buds of #D, #E, and #F were present within the margins outlined of the lesion and therefore extracted. A Geistlich Mucograft was then adapted to the alveolar defect, and then the site sutured.

Follow-up

Up to 15% of MNTI cases may develop recurrence. There is correlation found between age of diagnosis, and disease recurrence rate. Patients who are diagnosed with MNTI in the first 2 months of life had higher recurrence rates, whereas patients diagnosed after the 4.5-month mark tend to have the lowest recurrences. Close monitoring is suggested, for any required interceptive treatment, post the initial surgical course taken, again, particularly for patients diagnosed within the first 2 months of life.

Future Considerations

Due to the extent of such surgical treatment, facial asymmetry as well as premature loss of permanent dentition are also common complications needing a multidisciplinary approach in the years to come.

References

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