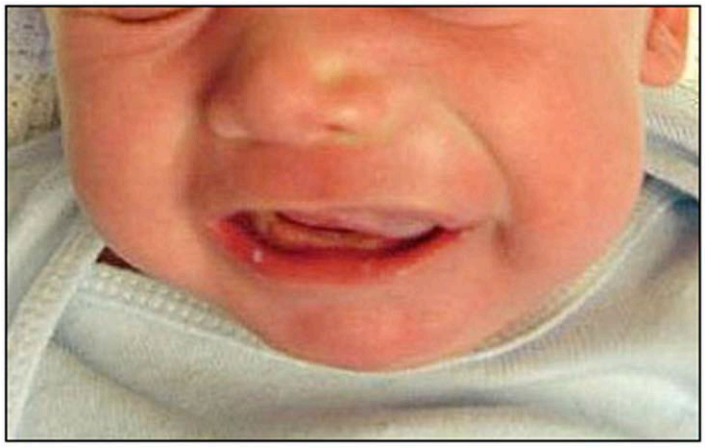
Maxillary Swelling in an Infant

James C. Burns, DDS, MS, PhD



The following Case Challenge is provided in conjunction with the American Academy of Oral and Maxillofacial Pathology.

Case Summary

This case challenge presents a two-month old infant demonstrating upper lip asymmetry.

An asymptomatic swelling was noted in the area of the left anterior maxilla of this two-month old infant by the child's parents. The swelling apparently was of rapid onset since the pediatrician at the one-month well baby check-up did not note it. The swelling caused an elevation of the left alae of the nose and slight facial asymmetry of the upper lip.

After you have finished reviewing the available diagnostic information, make the diagnosis.

Diagnostic Information

Clinical Findings

The intraoral lesion appeared as a 2 cm sessile, bluish mass located in the left anterior maxillary process. Palpation of the lesion did not seem to cause the infant any discomfort. A slight spongy feeling was suggestive of an absence of the buccal cortex. No thrill or pulse could be elicited from the mass. Although the lesion had supposedly expanded rapidly, the overlying mucosa was intact. (Figure 1)

Radiographic Findings

Under sedation, advanced imaging, using computed tomography (CT), was performed. The axial CT scan revealed a 2 cm radiolucent lesion with irregular borders. The space-occupying lesion was causing the displacement of teeth and interrupting their normal development. (Figure 2)

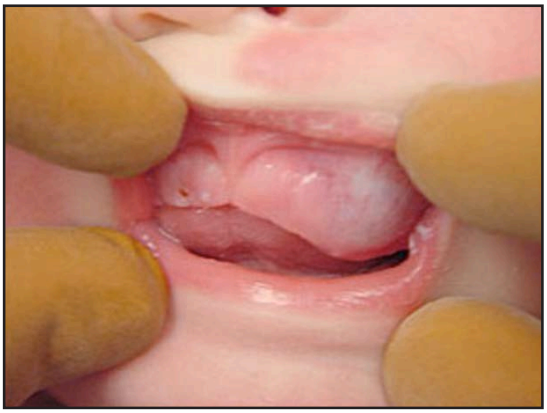


Figure 1. Intraoral examination reveals a 2 cm elevated bluish mass on the maxillary alveolar ridge.

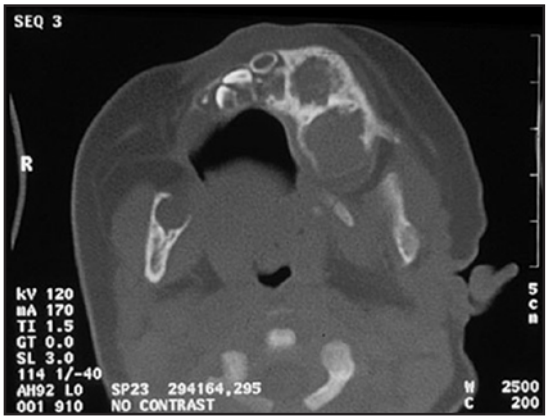


Figure 2. Axial CT scan reveals a 2 cm ill-defined radiolucency indicating evidence of the destruction of bone and displacement of the developing teeth.

Pathologic Findings

Histopathologic examination revealed a nonencapsulated tumor showing local infiltration into the adjacent bone. The lesion showed a mixture of large polygonal cells and small hyperchromatic round cells which were arranged in sheets or alveolar structures. (Figure 3) A number of the larger cells contained melanin pigment. (Figure 4)

Laboratory Studies

The histopathologic appearance of the specimen led the clinician to order a urinalysis. These results were normal for a young child, except for significantly elevated levels of vanillylmandelic acid (VMA). The VMA value was 3 standard deviations from the mean for a child in this age group.

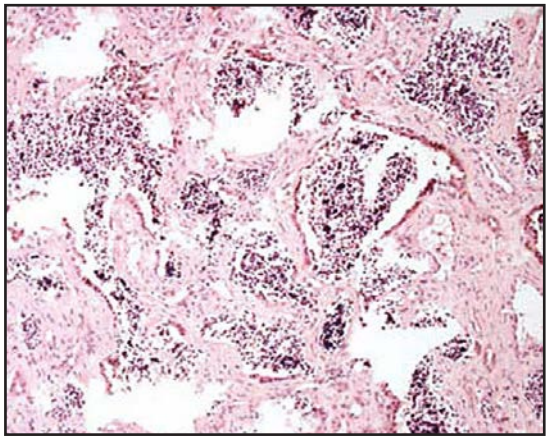


Figure 3. Low-power photomicrograph of the tumor showing infiltrating islands of tumor cells.

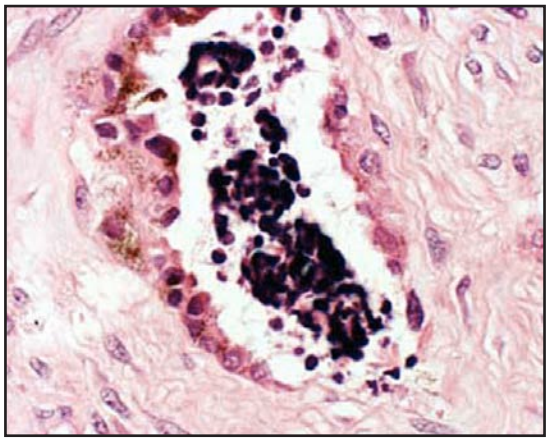
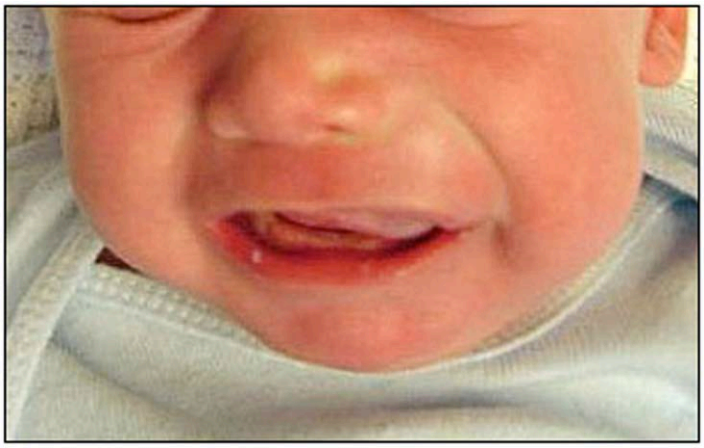


Figure 4. High-power photomicrograph of the tumor showing central round hyperchromatic cells surrounded by larger polygonal cells containing brown melanin pigment.

Can you make the diagnosis?

This case challenge presents a two-month old infant demonstrating upper lip asymmetry.



Select the Correct Diagnosis

A. Congenital Epulis
B. Melanoma
C. Neuroblastoma
D. Melanotic Neuroectodermal Tumor of Infancy

Congenital Epulis

Choice A. Sorry, this is not the correct diagnosis.

Although the location of this lesion and the age of the patient are consistent with a diagnosis of congenital epulis, the color of the lesion and its histopathologic appearance do not support this entity. Congenital epulis is a lesion that occurs almost exclusively in female newborns on their alveolar ridges, although a few cases have been described on the tongue. The congenital epulis usually presents as a pink elevation or mass on the newborn's alveolar ridge. The maxillary ridge is involved more often than the mandible. The histopathologic characteristics include uniform sheets of large, rounded, eosinophilic cells with an absence of alveolar-like structures and melanin. Treatment for the congenital epulis is usually surgical excision, although spontaneous regression has been documented in a few cases.[1]

Please re-evaluate the information about this case.

Melanoma

Choice B. Sorry, this is not the correct diagnosis.

The most common age for patients with intraoral melanoma is during the sixth and seventh decades of life. Although oral melanoma is often nodular and located on the palate or maxillary alveolus, the age of the patient and the histopathologic appearance in this case would rule out the diagnosis of melanoma. The color of the typical oral melanoma is often described as a brown to black lesion with irregular borders. Ulceration may develop in some cases. Pain is not a common feature, nor is intraosseous development since melanoma derives its origin from the epithelium. However, the underlying bone may show radiographic evidence of surface erosion. Treatment for melanoma is wide surgical excision that is often supplemented with chemotherapy or immunotherapy.[2]

Please re-evaluate the information about this case.

Neuroblaastoma

Choice C. Sorry, this is not the correct diagnosis.

The neuroblastoma is one of the most common childhood malignancies. It accounts for about 10% of cancer deaths in children. The majority of the patients are under five years of age, with 40% of the cases developing within their first year of life. Neuroblastomas often arise within the abdomen, arising in the adrenal glands or from the paravertebral autonomic ganglia. Since the neuroblastoma is of neuroectodermal origin, it can produce elevated levels of vanillylmandelic acid in the urine. Microscopically, the tumor grows in solid sheets of primitive-looking cells with large, hyperchromatic nuclei surrounded by scant cytoplasm. Often, special immunohistochemical studies are necessary to identify this primitive cell's origin. Prognosis is based upon stage of disease, age of the patient, histology, and Myc-N (oncogene). Depending on defined risk groups and clinicopathologic staging, survival rates range as high as 90% for low-risk groups, but less than 15% for high-risk groups.[3]

Please re-evaluate the information about this case.

Melanotic Neuroectodermal Tumor of Infancy

Choice D. Congratulations! You are correct.

The melanotic neuroectodermal tumor of infancy (MNTI) is a relatively uncommon osteolytic, pigmented neoplasm that primarily affects the jaws of newborn infants. The sexual predilection is nearly equal, and most patients present with the tumor in their first year of life. Although MNTI arises from neural crest cells and is usually a benign lesion, it is clinically worrisome because of its rapid onset and alarming local growth rate. The typical MNTI begins as a nonulcerated, lightly pigmented, blue or black lesion on the anterior aspect of the maxilla and rapidly expands to form a swelling or a tumescence that is cosmetically obvious to the parents of the infant. The intraoral lesion appears as a sessile, lobulated mass, often reaching 2-4 cm in diameter by the time of diagnosis. Bone destruction and displacement of teeth often occur because of the intraosseous location in the maxilla. More than 90% of MNTI occur in the head and neck region, with most lesions located on the anterior part of the maxillary ridge. Other common sites include the skull, the mandible, the epididymis, and the brain. Most examples are solitary lesions and treated with surgical excision. Recurrence has been reported in about 15% of cases. In addition, there are a few reported cases that have acted in a malignant fashion, resulting in metastasis and death.[4-6]

References

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About the Author

Note: Bio information was provided at the time the case challenge was developed.

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