Pediatric Maxillary and Mandibular Tumors

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KEYWORDS

• Maxilla • Mandible • Tumor • Odontogenic cyst • Osteomyelitis • Ameloblastoma • Langerhans cell histiocytosis • Osteosarcoma

KEY POINTS

- Pediatric masses of the jaw can include both benign and malignant processes of both odontogenic and nonodontogenic origin, as well as infectious processes.
- High-resolution imaging with computed tomography and/or MRI can be suggestive, but often a definitive diagnosis requires tissue for histopathology.
- Treatment is usually surgical and must take into account the growth potential of the facial bones, the aggressiveness of the lesion, and the rate of recurrence.
- The clinical presentation of certain benign jaw lesions may suggest an underlying genetic disorder.

INTRODUCTION

Pediatric maxillary and mandibular tumors encompass a broad range of causes, histopathologies, and clinical behaviors. Although many benign lesions are asymptomatic and are discovered only on a routine dental radiograph, locally aggressive lesions and malignancies can cause pain, facial swelling, and trismus. The work-up is often challenging because many of the tumors have nonspecific radiographic findings; open or excisional biopsy is often necessary for a definitive diagnosis. Treatments range from simple enucleation to a large segmental resection and reconstruction. The clinician must be cognizant of both odontogenic and nonodontogenic processes that can be involved in order to prevent either an unnecessarily aggressive resection with resultant facial deformity or an overly conservative resection that leads to recurrence.

NONODONTOGENIC BENIGN TUMORS

Osteoma

Osteomas are slow-growing benign osteogenic tumors most commonly discovered in late adolescents and young adults. They are composed of well-differentiated lamellar

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Abbreviations	
BL	Burkitt lymphoma
CGCG	Central giant cell granuloma
CT	Computed tomography
ES	Ewing sarcoma
FD	Fibrous dysplasia
LCH	Langerhans cell histiocytosis
MRI	Magnetic resonance imaging
NBCCS	Nevoid basal cell carcinoma syndrome
OKC	Odontogenic keratocyst
PCO	Primary chronic osteomyelitis
PFD	Polyostotic fibrous dysplasia
PTCH	Patched
XRT	Radiotherapy

or compact bone.[1,2] Although small endosteal (derived from bone marrow) lesions are often asymptomatic, larger lesions can cause a progressive enlargement of the affected area with impingement on adjacent structures. Paranasal sinus osteomas are more common than mandibular osteomas, and most of these arise in the frontal or ethmoid sinuses.[3] Maxillary osteomas are rare, and can remain asymptomatic for long periods of time. Mandibular lesions are commonly found in the posterior body or mandibular condyle, with the latter causing a progressive shift in occlusion.[1] Treatment includes either watchful waiting for smaller or asymptomatic maxillary tumors or conservative surgical excision for symptomatic tumors.

In addition to the differential from more aggressive bone-forming tumors, the discovery of a craniofacial osteoma is significant because of the association with Gardner syndrome, which is a triad consisting of colonic polyps, multiple osteomas, and soft tissue tumors such as epidermoid cysts and desmoid tumors.[4] Gardner syndrome is an autosomal dominant disorder with near 100% penetrance caused by a genetic mutation at chromosome 5. Colonic polyps begin to manifest during puberty and have a high rate of malignant transformation. The onset of multiple osteomas during puberty should alert the clinician to the possibility of Gardner's syndrome.

Osteoblastoma

Osteoblastomas are benign neoplasms of osteoblasts. In the pediatric population they are usually discovered during adolescence. Osteoblastomas often present with pain and swelling that is not relieved with nonsteroidal antiinflammatory drugs. Craniofacial osteoblastomas most commonly involve the medullary bone of the posterior mandible, appearing as radiopaque or radiolucent lesions with patchy areas of mineralization.[1] Most cases are treated successfully by local excision or curettage; however, an aggressive variant of osteoblastoma with irregular histologic features has been described with a recurrence rate of up to 50%.[2]

Fibrous Dysplasia

Fibrous dysplasia (FD) is a nonneoplastic disease caused by the replacement of normal bony architecture by fibrous connective tissue in a loose, whorled pattern intermixed with irregular areas of bony trabeculae.[1,5] It is caused by a sporadic mutation in the GNAS1 gene on chromosome 20, resulting in abnormal osteoblast differentiation and irregular bone formation.[5] The clinical presentation is that of painless, slowly

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