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Osteocartilaginous Tumors in the Parapharyngeal Space Arising from Bone Exostoses

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Three osteocartilaginous tumors that arose from bone stalks were diagnosed by the demonstration, on multiplanar cross-sectional CT and MR images, of the site of stalk attachment to the adjacent mandible (one), skull base (one), and cervical vertebra (one). All three patients presented with signs and symptoms related to mass effect in the parapharyngeal region. Mass effect was more pronounced in the two cases of malignant degeneration (osteosarcoma, chondrosarcoma) than in the single case of benign osteocartilaginous exostosis.

Differentiation of benign from malignant osteocartilaginous tumor may be accomplished by appreciating characteristic CT and MR features of the cartilage cap. The differential diagnosis of mass lesions of the parapharyngeal space should include tumors of osteocartilaginous origin.

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Rarely, parapharyngeal masses originate from osteochondral elements derived from adjacent bone structures, such as the skull base, mandible, and vertebral column. These typically are destructive tumors that intimately involve the bone of origin. We present three cases of head and neck masses of osteochondral origin that projected into the parapharyngeal space and yet were derived from exostoses arising from adjacent vertebral, mandibular, or skull-base bone. These tumors were easily diagnosed once the relationship of tumor and bone stalk were appreciated on sectional images. Such tumors should be considered when listing a differential diagnosis of lesions involving this anatomic region. A specific diagnosis may be suggested by the characteristic findings described here.

Case Reports

Case 1

A 38-year-old woman presented with a history of left neck fullness and pain for several months. Lateral xeroradiography of the cervical spine (Fig. 1A) showed a calcified mass projecting ventral to the C2 and C3 vertebral bodies and a soft-tissue component projecting into the oropharynx and hypopharynx. Axial CT images confirmed the presence of a significant soft-tissue component (Figs. 1B and 1C) and revealed a peglike osseous continuity between the pharyngeal mass and the C2 vertebral body.

Histologic examination after an open biopsy and partial excision of the soft-tissue portion of the lesion showed malignant spindle cells with frequent mitoses in a bone and cartilaginous stroma. The final histologic diagnosis was fibroblastic variant of osteosarcoma (grade III), shown radiologically to arise from an osteocartilaginous exostosis at the C2 level.

Case 2

A 13-year-old boy presented with a 3-month history of decreased hearing on the left side, dysphagia, and speech difficulty. Physical examination revealed medial deviation of the left tonsil and palate.

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CT studies (Fig. 2) showed a mixed-attenuation soft-tissue mass with multiple calcifications, centered within the left parapharyngeal space, separated from the pharyngeal musculature by a displaced parapharyngeal fat pad. Coronal CT showed that the calcified soft-tissue mass clearly arose from an osseous stalk attached to the inferior surface of the left skull base.

At surgery, a transmandibular wide-field exposure was performed. The tumor was freed from its osseous pedicle at the skull base with a curved broad chisel. Grossly, the ovoid tumor (Fig. 3) was well circumscribed but not encapsulated, and on cut section it appeared firm and pinkish-white.

The final diagnosis, which was based on histologic and radiographic information, was grade II chondrosarcoma arising from the

osteocartilaginous stalk attached to the skull base (exostotic chondrosarcoma). CT follow-up 2 years later revealed no gross recurrence.

Case 3

A 61-year-old man presented with a 20-year history of a left-sided jaw mass and a 2½ month history of trismus. On physical examination, the patient could not open his mouth more than 0.5 cm.

Axial and coronal CT scans (Fig 4) showed sclerotic thickening and displacement of the left mandibular condyle, as well as hyperostosis of the adjacent left skull base and lateral pterygoid plate. A 5.7-cm calcific mass (without a significant soft-tissue component) could be identified in the left infratemporal and parapharyngeal regions.

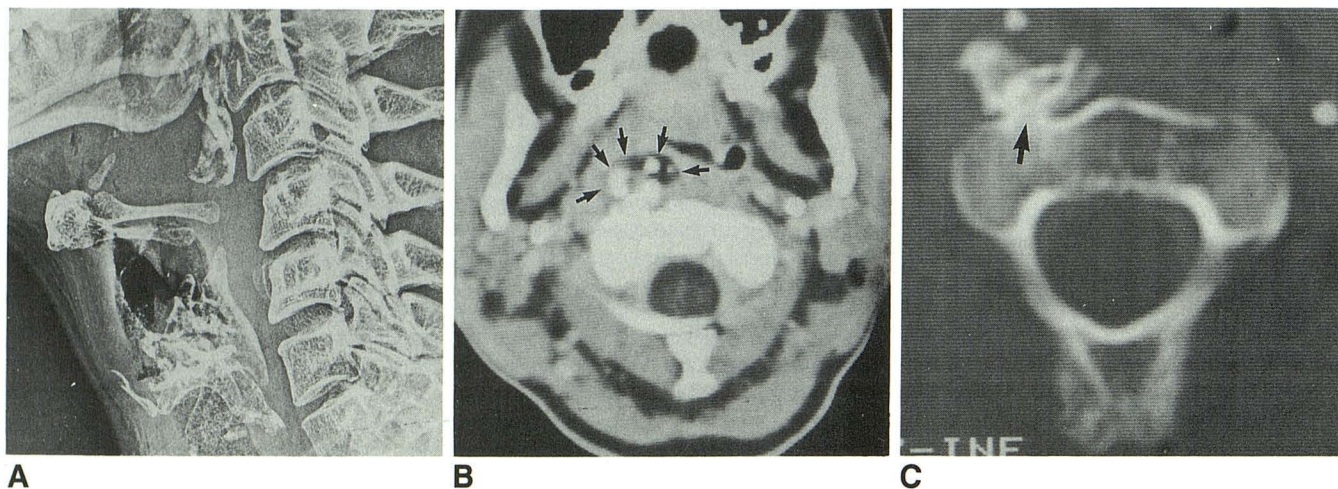


Fig. 1.—Case 1: 38-year-old woman with painful left neck mass. Fibroblastic osteosarcoma (III) arising from C2 vertebral body exostosis. A, Lateral xeroradiograph of the neck shows dense calcification parallel to C2 and C3 vertebrae. A nonmineralized soft-tissue component extends ventrally into the hypopharynx. B and C, Axial CT images through C2 vertebral body show heavily mineralized mass extending into parapharyngeal and retropharyngeal spaces at junction of naso- and oropharynx (arrows in B). The mass arises from a thin exostosis, continuous with right side of C2 vertebra (arrow in C).

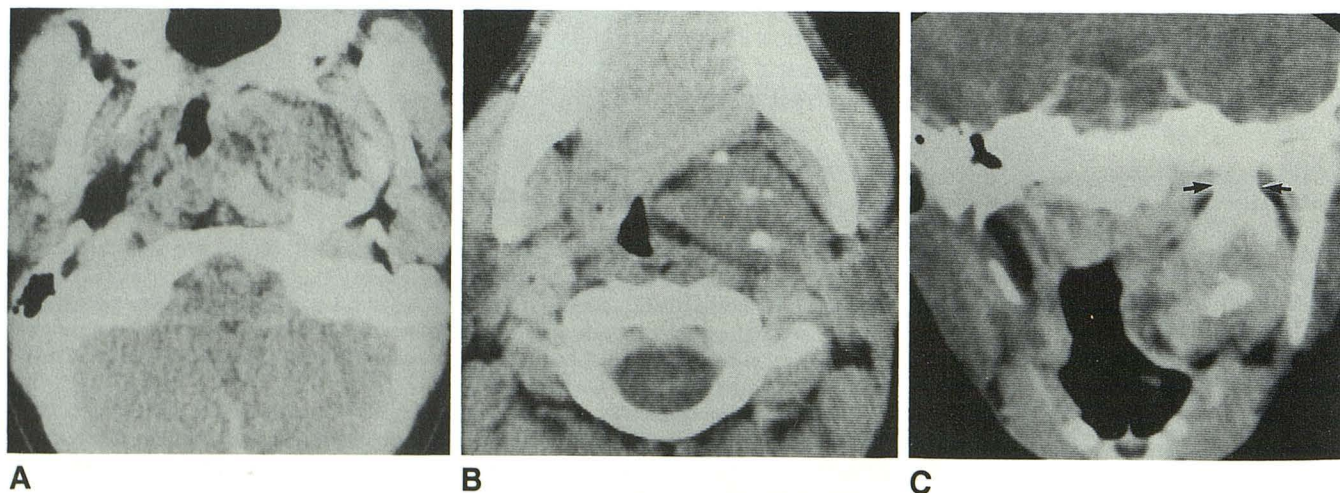


Fig. 2.—Case 2: 13-year-old boy with dysphagia and left conductive hearing loss. Chondrosarcoma (II) arising from an exostosis of the undersurface of the skull base.

A and B, Axial CT scans define a well-margined mass apparently centered within left parapharyngeal space. Parapharyngeal fat surrounds the mass, medially separating it from the pharyngeal musculature. Scattered areas of mineralization are noted.

C, Coronal CT scan reveals that a portion of the mineralization seen on the axial studies is in fact an osseous stalk (exostosis, arrows), arising from the undersurface of the sphenoid bone at the middle cranial fossa floor. Note circumferential distribution of parapharyngeal fat.

MR images (Fig. 5) demonstrated a mixed iso- and hypointense mass projecting into the parapharyngeal space, distorting fascial planes and flattening the retroantral portion of the infratemporal fossa fat pad. The coronal images demonstrated contiguity of the mass with the mandible and the region of peglike reactive sclerosis at the skull base.

Surgical excision of the tumor and the left mandibular condyle was performed en bloc. A rock-hard mass could not be totally resected, and a thin layer of tumor remained firmly fixed to the skull base.

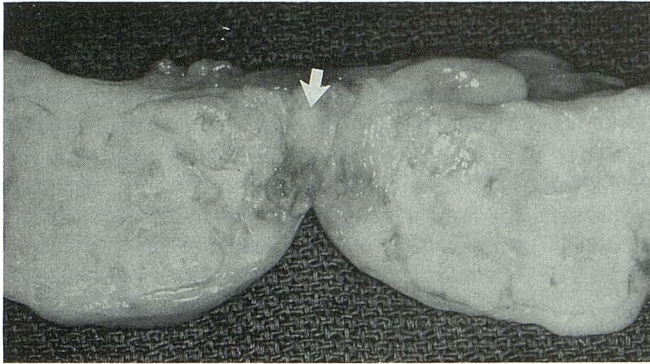


Fig. 3.—Case 2: Pathology photograph shows bivalved chondrosarcoma, with site of origin from exostosis indicated by arrow.

postoperatively. Histologic examination showed bone, hyaline cartilage, and fibrovascular tissue, consistent with a diagnosis of benign osteochondroma.

Discussion

Tumors of the parapharyngeal space are uncommon and represent only 0.5% of head and neck tumors [1–4]. Of these, 50% originate from salivary glands and 30% from neural elements [1]. The remaining 20% are of miscellaneous origin, with metastatic carcinoma and direct extension of tonsillar or nasopharyngeal carcinoma being most common [1]. The four most common tumors found in this location, in descending order of occurrence, are deep-lobe parotid tumors, tumors of minor salivary gland origin, paragangliomas, and neuromas (vagal or sympathetic chain origin).

The three osteochondral origin lesions presented here are examples of tumors arising adjacent to the parapharyngeal fat space and secondarily extending within it. All patients presented with symptoms referable to this compartment. Osteochondral lesions rarely present in the parapharyngeal region. In a review of 91 cases of tumors of the parapharyngeal space by Som et al. [4], there was reference to only one chondrosarcoma. This tumor arose directly from the skull base and was not illustrated. Other authors [5–8] have re-

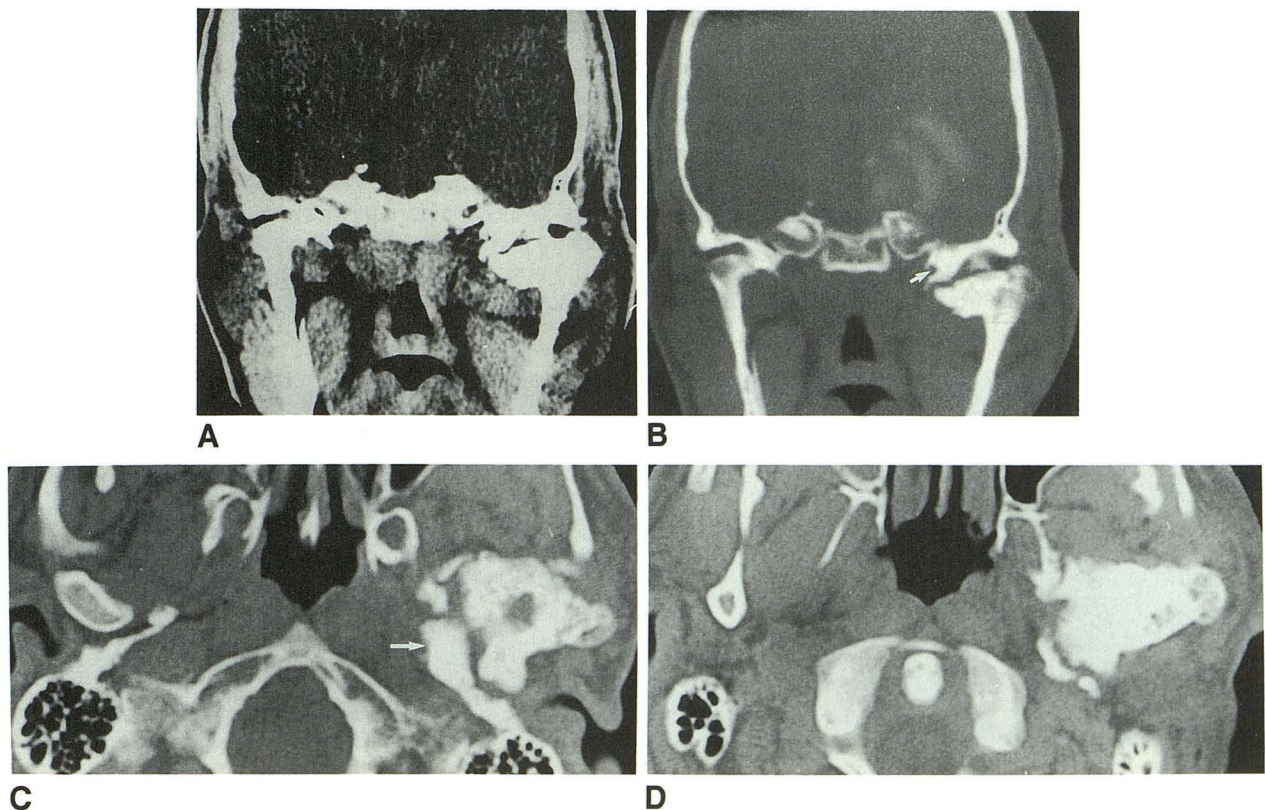


Fig. 4.—Case 3: 61-year-old man with 20-year history of left jaw mass, with increasing trismus. Osteocartilaginous exostosis. A–D, Coronal (A and B) and axial (C and D) CT scans reveal extensive mineralization and thickening of left mandibular condyle. Note absence of significant soft-tissue mass and normal local fat planes adjacent to pterygoid muscle (A). A secondary area of localized hyperostosis, most likely related to chronic impaction, is seen to arise from undersurface of skull base (arrows, B and C).

ported cases of osteosarcoma and chondrosarcoma in the craniofacial region; however, the cases described arose directly from bone without underlying exostosis. The lesions we present all arose from a bone stalk. Therefore, multiplanar CT and MR images aided in the demonstration of a characteristic benign-appearing osseous attachment, the appreciation of which suggested the origin of the lesion (and diagnosis) in all three cases.

Once the diagnosis of an osteocartilaginous lesion is made, it is then important to distinguish between benign and malignant tumor. Several imaging considerations may be of diagnostic help in this respect. The two malignant lesions (cases 1 and 2) had sizable associated soft-tissue components capping the underlying exostosis, and the benign osteochondroma (case 3) did not. Others have noted the relationship between cartilage cap thickness and malignancy. Hudson et al. [9] found that CT failed to detect cartilage caps that were less than 2.5 cm in diameter, and when CT failed to show such a soft-tissue cap the lesion was typically benign. In another series of 16 benign exostoses, CT failed to show a cap in 14. Exact size criteria for differentiation of grade based on cap thickness do not, however, exist [10].

Although the chondroid tumor matrix may have a distinctive MR appearance (homogeneous high signal intensity on T2-weighted images corresponding to areas of hyaline cartilage [11]), cartilage-origin lesions with a predominantly cellular matrix (as in our case) may not have this striking appearance, and identification of cartilage may not be possible on the basis of long TR spin-echo MR intensity alone. In case 3, a cartilage cap could not be identified on MR (or CT) despite its histologic presence, a disparity that is common according to previously reported experience [9]. In tumors of alternative origin, lesional calcifications are unusual in this location. All our cases showed calcification on CT, and in case 3 (osteochondroma) the MR scan demonstrated areas of hypointensity corresponding to these areas of mineralization.

Osteochondromas comprise 40–45% of all benign bone tumors [12]. They may arise from any bone preformed in

cartilage, but most often they begin near the metaphysis in flat and long bones [13–15]. Eighty-nine percent are solitary [12]. Three to five percent of cases involve the vertebral column [16], and these cases typically arise from the neural arch [13, 17]. The cervical spine is least frequently affected (three of 22 vertebral lesions in two series) [12, 18]. One prior case has been reported of an osteochondroma arising from a cervical vertebral body and that case presented as a neck mass with symptoms of hoarseness due to supraglottic edema related to repeated laryngeal impaction upon the mass. Other reported clinical findings in vertebral osteochondroma include dysphagia, vascular compromise, and spinal cord compression [14, 17, 19–21]. Radiographs typically show a projection of medullary bone and cortex, continuous at its base with the parent bone [15]. The attaching stalk of bone may be sessile or pedunculated; there is no associated bone erosion [14]. Calcification may be seen in a cartilaginous cap. Extensive irregular calcifications indicate malignant degeneration [12], although this is unusual (1–5% of cases), [12, 17]. Degeneration is more common in cases with multiple exostoses (10–25%) [10, 22]. Since benign tumors stop growing during adolescence, growth in adult life suggests malignancy [14].

When no cartilaginous cap is visible on CT studies, the lesion is typically a benign exostosis [9]. In our case 3, no cap or soft-tissue mass was detected. Although the cartilage cap may be more easily detected by MR [11], none could be detected in case 3. In cases 1 and 2, the presence of an associated soft-tissue mass supported the diagnosis of malignancy despite the coincident presence of a benign-appearing osseous stalk in each case.

Chondrosarcomas may arise from osteocartilaginous exostoses [10], and differentiation from benign exostosis by CT features may be difficult. The size of an associated soft-tissue mass is most important in this respect. Kenney et al. [15] noted a thin cartilage cap (less than 3 cm, average thickness of 0.6 cm), which was largely mineral in six of six cases of osteochondroma, and a cartilage cap over 3-cm thick in six



Fig. 5.—Case 3: Axial and coronal MR images show low-signal intensity in region of mineralization shown by CT. A, Axial SE 500/30/2 image shows ventral displacement of pterygoid muscle (solid arrow) and medial deviation of parapharyngeal fat pad (open arrow) by bone mass. B and C, Proton-density-weighted SE 2000/50/2 coronal MR images show lack of associated soft-tissue mass. Note site of mandibular attachment (arrow, B) and the skull base hyperostotic spur (arrow, C) as regions of signal void.

of six chondrosarcomas, some of which had extensive lobulated soft-tissue masses and a variable amount of mottled disorganized calcification.

Craniofacial chondrosarcomas tend to grow slowly and are typically large and multilobulated at the time of diagnosis [6, 7]. They have a propensity to arise from the wall of the sphenoid sinus; from the junction of the sphenoid, ethmoid, and vomer; and from the undersurface of the sphenoid bone. Three of 15 cases reported by Lee and Van Tassel [6] arose from the undersurface of the greater sphenoid wing near the pterygoid plates, without extending intracranially. Extension into the infratemporal fossa and parapharyngeal space was noted in three cases and illustrated in one. All cases had associated skull-base destruction. No such destruction was present in our case, since the malignant tumor arose at the distal end of the stalk, away from the site of attachment at the skull base. There is no previous report of a chondrosarcoma in this region arising from a benign exostosis (case 2).

Osteosarcomas typically occur in older children and young adults between the ages of 10–25 years. Common sites are the metaphysis of the distal femur and proximal tibia. Histologic subtypes depend on the dominant differentiated tumor cell type (osteoblastic, chondroblastic, or fibroblastic) [18]. Most have a calcified matrix, either cloudlike (osteoid) or stippled and ringlike (chondroid) [23].

The craniofacial bones are affected in 8.6% of cases [18]. They tend to occur one decade later than lesions arising in long bones (average age, 34 years) [18]. In a large series of 46 cases, Lee et al. [8] reported 32 de novo craniofacial osteosarcomas (14 others arose in radiated fields). Eleven arose from the maxillary alveolar ridge, 13 from the mandible, and eight from the skull and skull base. The tumors were mostly lytic, although sclerosis was often observed in the mandibular lesion. Soft-tissue masses were seen in all cases and calcification was present in over 50%. None was described as arising from an osteocartilaginous stalk.

Conclusions

Osteochondral tumors projecting into the parapharyngeal space are rare. The demonstration of a bone-stalk attachment in the three cases in our series suggested the diagnosis of a tumor of osteocartilaginous origin. CT or MR demonstration of an associated soft-tissue mass, calcification, or a chondroid tumor matrix may further aid in differentiating benign exostosis from malignant degeneration. The absence of an associated soft-tissue mass typically indicates the presence of a

benign exostosis. Ultimately, the aggressiveness of the tumor must be determined by histologic analysis.

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