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ORIGINAL PAPER

Osteochondroma of the Hyoid Bone: A Previously Unrecognized Location and Review of the Literature

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Abstract Osteochondroma is a benign cartilaginous neoplasm and the most common benign tumor of bone. Osteochondromas occur primarily in the axial skeleton with a predilection for the distal femur, and relatively few cases occur in the head and neck region. The majority of cases of osteochondromas in the head and neck region affect the mandibular condyle, with fewer cases reported in the skull base and the neck. To our knowledge, there is no reported case of osteochondroma of the hyoid bone documented in the English literature. We thus report the first case of a hyoid bone osteochondroma, presenting as an asymptomatic mass in a young woman.

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Introduction

Osteochondromas are the most common benign bone tumor, comprising more than a third of all benign bone tumors. Osteochondromas typically occur in young patients, with most cases presenting before the age of 30 [1–3]. These appear to be more common in males, although the exact reported male to female ratio varies by series and ranges from about 3:1 to 1:1 [3, 4]. Although osteochondromas can occur in virtually any bone that undergoes enchondral bone formation, they are most commonly found in or around the distal femur.

Osteochondromas occurring in the head and neck region are relatively rare, especially in contrast to their occurrence in the axial skeleton. The majority of cases in this region affect the mandibular areas such as the condylar and coronoid processes [5, 6]. We describe a case of osteochondroma arising from the hyoid bone, its surgical management, and the differential diagnosis with a brief review of literature. To our knowledge, this is the first reported case of an osteochondroma involving the hyoid bone in the English literature.

Case Report

A 31-year-old female with an otherwise insignificant past medical history was referred to the otolaryngology clinic for evaluation of an asymptomatic left neck mass that she first noted 2 years ago. The mass had been closely followed and evaluated by multiple primary care physicians and

otolaryngologists, with multiple physical exams and radiological scans. The mass had not substantially grown in size over this time and did not cause the patient pain, paresthesia, or dysphagia. However, the patient and her family did harbor a substantial amount of associated anxiety as the location and nature of the mass precluded evaluation by fine needle aspiration and thus definitive characterization. The systemic review was not contributory.

A head and neck exam was performed. There was a palpable firm, non-tender, left neck mass that moved well with deglutition. It was approximately 1–2 cm in size and just inferomedial to the submandibular gland. Laryngoscopy demonstrated normal vocal fold mobility. Laboratory studies were unremarkable.

Real-time ultrasound examination of the palpable area in the submandibular region noted a lobular bony nodule with overlying hypoechoic well-marginated cartilaginous cap extending from the lateral aspect of the hyoid bone on the left. There was no infiltration into the adjacent soft tissues. No blood flow was demonstrated in the cartilaginous cap. The nodule measured 2.3 × 1.9 × 1.5 cm on imaging. These findings correlated with the MR and computed tomography (CT) scan findings (Fig. 1). The oral pharynx, larynx, parotid and submaxillary glands demonstrated no abnormalities. The MR demonstrated somewhat prominent lymph nodes in both submandibular regions and along level IIB bilaterally, which were thought to be most likely reactive.

Given the lack of a definite diagnosis, the patient's anxiety and the need for continual follow-up of this lesion, the patient decided to undergo surgical excision. At surgery, the mass was completely separate from the left submandibular gland. The hyoid bone was fully skeletonized, and the large mass was exposed extending from the midportion to the lesser cornu of the hyoid bone. The tumor was excised en bloc. Grossly, the specimen consisted of a 2.3 × 0.9 × 0.7 cm excision of tan-brown firm bone with an attached 1.6 × 1.4 × 1.3 cm tan-brown ovoid calcified nodule. The mass was serially sectioned to reveal tan-brown rubbery cut surfaces. On microscopic examination, the lesion demonstrated three layers, consisting of a fibrous perichondrium outer layer overlying a hyaline cartilage cap and underlying mature lamellar bone with marrow elements (Fig. 2). The fibrous outer layer was continuous with the periosteum of the underlying bone. The clinical, radiologic, and histologic findings were consistent with a benign osteochondroma.

Discussion

Osteochondromas are benign neoplasms consisting of a cartilage-capped bony projection from the external surface of bone. They are the most common benign bone tumor, with a reported incidence of about 35 % of all benign bone tumors and 8 % of all bone tumors overall [7]. The true frequency of these tumors may be higher, due to the fact that the majority are asymptomatic and may never be detected or excised. They may also spontaneously undergo regression [8].

Osteochondromas generally present as a long-standing, hard, painless mass near a joint on the axial skeleton, although they may also present as a painful mass associated with a local trauma. Symptoms, when they occur, depend on both the size of the lesion as well as the location. For instance, osteochondromas that affect the ribs are occasionally associated with complications including

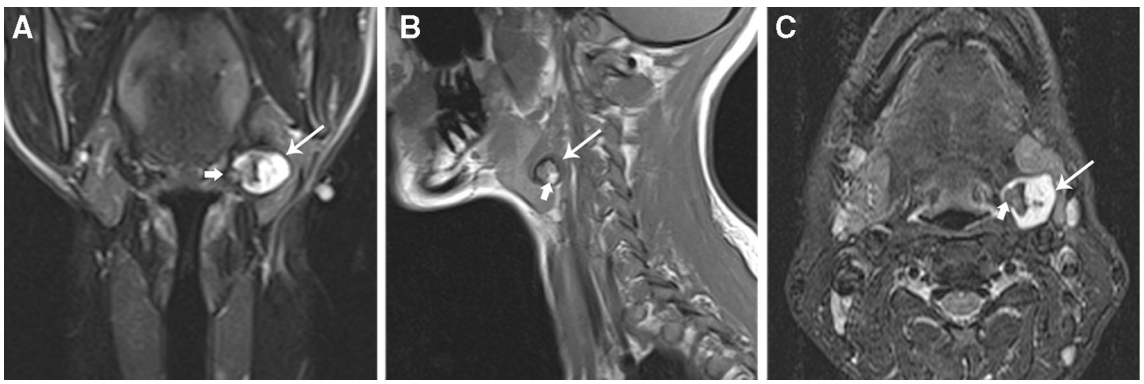

Fig. 1 MRI of hyoid bone osteochondroma. **a** Coronal fat-suppressed T2-weighted image shows a mixed signal intensity mass (*long arrow*) arising from the *left* greater horn of the hyoid bone, near its junction with the hyoid body and lesser horn (*short arrow*). **b** Sagittal T1-weighted image shows internal bright signal within the *center* of the mass (*short arrow*), reflecting fat within marrow elements contiguous with the underlying bone. The cartilage cap (*long arrow*) shows low T1 signal. **c** Axial fat-suppressed T2-weighted image shows suppression of fat signal the central portion of the mass (*short arrow*), and bright signal in the cartilaginous cap (*long arrow*). The cartilaginous cap measures 9 mm in thickness

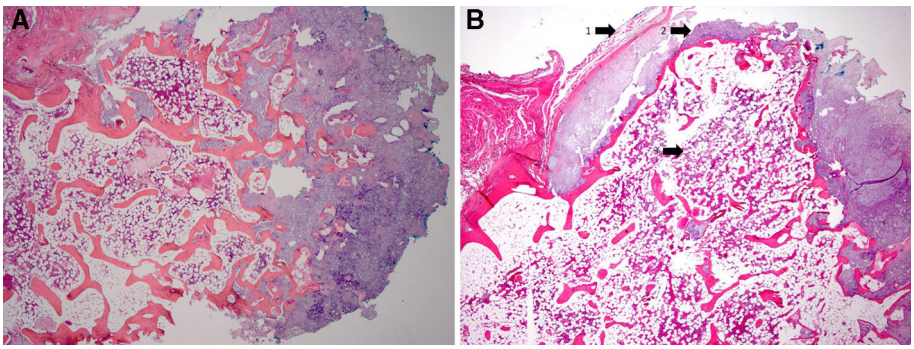

Fig. 2 **a** Histology demonstrates a cartilaginous neoplasm on high power. H&E 2×. **b** On microscopic examination, the lesion shows three layers: *1* A fibrous perichondrium outer layer *2* A hyaline cartilage cap and *3* Underlying mature lamellar bone with marrow elements

pneumothorax, hemothorax, and pericardial effusion [9–12]. Osteochondromas may also affect growth plates, leading to short stature and angular deformities, particularly in patients with hereditary multiple osteochondromas. A number of secondary complications can occur such as fracture, bursa formation, arthritis, and impingement on tendons, nerves, or vessels, including encroachment on the spinal canal [13, 14].

Osteochondromas are commonly found incidentally on radiographs that are obtained for other reasons. The radiographic appearance is very characteristic, with osteochondromas growing out in one direction opposite to the adjacent joint when located in the metaphyses of long bones. The cortex of the bony protuberance is continuous with the cortex of the underlying bone. Both CT and MRI are helpful modalities to demonstrate the bony and cartilaginous trabeculae of this lesion [15–17]. Continued lesion growth and a cartilage cap greater than 1.5 cm in thickness, after skeletal maturity, are associated with malignant transformation. Although the features are often pathognomonic, additional imaging modalities may be used, particularly when the lesion is symptomatic or in unusual locations, and include bone scintigraphy, ultrasonography (US), CT, and magnetic resonance (MR) imaging [18].

Osteochondromas can be sessile or pedunculated, with smaller ones tending to be sessile and large ones tending to be pedunculated. The average greatest dimension is about 4 cm, although they may reach more than 10 cm in size. Grossly, osteochondromas have a characteristic cartilaginous cap that is covered by a thin fibrous membrane. The cartilaginous cap has an average thickness of about 0.6 cm, and rarely exceeds 1 cm. It is continuous with the periosteum of the adjacent bone. Microscopic examination demonstrates mature bony trabeculae containing normal bone marrow and an overlying cartilaginous cap composed of cells resembling those of normal hyaline cartilage. Between these two areas are commonly areas of enchondral ossification. Eosinophilic, periodic acid-Schiff stain positive inclusions can be seen in the cytoplasm of these cells [19, 20].

Although osteochondroma can occur in virtually any bone, they show a predilection for the metaphyseal region of the distal femur, the upper humerus, the proximal tibia, and the fibula [8]. Rarely, osteochondroma can occur in the head and neck. When found in this region, the majority of cases affect the mandibular areas such as the condylar and coronoid processes, although cases affecting the cranial base, jaw, maxillary sinus, ramus, body, and symphyseal mandibular region have also been noted [5, 6]. The incidence of osteochondroma in the jaw bone only accounts for 0.6 % of the whole body incidence [21]. Symptoms in this region may include painless or painful swelling and facial asymmetry.

The exact pathogenesis of these neoplasms is not fully understood. Studies have suggested osteochondroma pathogenesis involves herniation of the epiphyseal cartilage as a result of a defect in the periosteal cuff of bone. However, genetic karyotyping has revealed reproducible genetic abnormalities that would support a neoplastic nature to these benign lesions. The majority of both hereditary and sporadic osteochondromas demonstrate biallelic inactivation of the EXT1 and EXT2 tumor suppressor genes, although spontaneous mutations can also occur [1, 22]. The EXT gene products are involved in heparan sulfate biosynthesis, with mutated cells being heparan sulfate deficient. The cartilage cap of osteochondromas is made of a combination of both wildtype and mutated cells [23–25], and it is thought that loss of heparan sulfate may give the affected chondrocyte a proliferative advantage [26, 27]. Additionally, heparan sulfate is also involved in the hedgehog signaling pathway, and thus may affect the formation of the bony collar [28].

Osteochondromas clinically may mimic a number of lesions, and the differential diagnosis varies from relatively benign to malignant lesions. Benign conditions in the differential diagnosis include bony spur formation from osteoarthritis, condylar hyperplasia chondroma and osteoma, while malignant neoplasms that may mimic osteochondromas include chondrosarcoma, osteosarcoma, fibrosarcoma, giant-cell tumor, and metastatic tumors [5, 29–31]. Bizarre parosteal osteochondromatous proliferation (also known as Nora's lesion) rarely may enter the clinical differential diagnosis [32–34]. This surface based bony lesion

also is continuous with the bony cortex, but occurs most commonly on the hands and feet [3]. On histology, Nora's lesion usually is easily distinguished from osteochondroma as it shows disorganized bone, bizarre fibroblasts, and highly cellular cartilage. A combination of clinical, radiological, and histological criteria should help distinguish osteochondroma from other entities.

Asymptomatic osteochondromas do not require any further treatment, and can be monitored initially with radiographs and subsequently by clinical examination. Further investigation and possible surgical intervention may be implemented if the lesion is painful or becomes painful, or if there is an increase in size of a preexisting lesion. Increasing pain may be a manifestation of malignant transformation into a secondary chondrosarcoma, or may be a mechanical symptom. Other features that may be concern for malignant transformation include increasing size after skeletal maturity, cartilaginous cap with a thickness equal to or greater than 2 cm, growth disturbance, and new onset of symptoms [1, 35–38]. Osteochondromas arising from the spine, scapula, pelvis, or proximal femur are especially prone to malignant transformation. Definitive treatment is complete surgical excision.

In conclusion, we report the first case in the English literature of an osteochondroma arising from the hyoid bone. Although osteochondroma is the most common benign bony neoplasm, they occur infrequently in the head and neck region. Osteochondroma have characteristic clinical, radiological, and histological features no matter where they occur in the body, and thus should remain in the differential of head and neck lesions that fit these criteria.

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