CASE REPORT

Chondrosarcoma of the hyoid bone: an atypical site of a sarcoma of the head and the neck

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SUMMARY

We describe a case of a 73-year-old man with a chondrosarcoma of the hyoid bone. The patient presented with a painless palpable lump in the upper anterior cervical region that had been growing for over 10 months. Fine-needle aspiration cytology suggested pleomorphic adenoma but further imaging investigation with CT revealed an exophytic tumour originating from the body of the hyoid bone with prominent chondroid-like calcifications typical of a chondrosarcoma. Histopathology of the surgical specimen confirmed the diagnosis of a moderately differentiated chondrosarcoma. Chondrosarcomas account for 11% of all bone cancers, of which up to 12% are in the head and neck. Primary sites of the head and the neck include the skull base, the nasal cavity, the maxilla, the mandible and the larynx. Chondrosarcomas of the hyoid bone are exceptionally rare, with only 20 such cases previously reported in the English literature.

BACKGROUND

Chondrosarcomas of the hyoid bone are a rare occurrence with only 20 cases reported in the English literature since the early 1970s. Cytopathology of such suprahyoid neck masses without imaging guidance may lead to misdiagnoses with other more common cartilage-containing lesions such as pleomorphic adenoma originating from the submandibular or sublingual glands, as occurred in this case. Therefore, cross-sectional imaging is crucial in the preoperative diagnosis of such lesions. Moreover, in contrast to other more common locations in the head and neck, hyoid bone chondrosarcomas tend to present as painless, often slow-growing palpable masses with no other associated symptoms such as hoarseness, dysphagia, breathing difficulty or enlarged cervical lymph nodes. The paucity of symptoms and signs, and non-aggressive behaviour, may suggest a benign mass, which may lead to a more conservative surgical approach. However, the recommended treatment is wide local surgical excision with total hyoidectomy, to decrease the chance of recurrence and histological dedifferentiation.

CASE PRESENTATION

A 73-year-old man with a painless palpable mass that had grown in the right submandibular/submental region for over 10 months was referred to our institution. On physical examination, a 5 cm, ovoid-shaped submental mass was detected. Other physical and laboratory assessments were unremarkable.

INVESTIGATIONS

A neck CT was performed, revealing a large and relatively well-defined, multilobulated lesion in the submental and prehyoid regions, measuring approximately 50×40×30 mm (figures 1–3). The lesion had a hypodense soft tissue component and exuberant calcifications, with a coalescent ring-and-arc pattern, suggestive of chondroid matrix (figures 1–3). The lesion extended into the prelaryngeal space, pushed the floor of the mouth upwards, and was clearly separated from the sublingual gland and from the submandibular glands. The pre-epiglottic space was also preserved.

The lesion was in close contact with the right paramedian region of the body of the hyoid bone where insufflation and rupture of its outer cortical margin could be seen, although tumour growth was predominantly exophytic into the prehyoid region (figures 1 and 2). The remainder of the hyoid bone was normal in size and morphology. Also noted was the anterior displacement of the strap muscles confining the lesion anteriorly.

No cervical lymphadenopathy was seen.

These imaging findings were highly suggestive of a chondrosarcoma of the hyoid bone. In spite of the prior cytological diagnosis of a pleomorphic adenoma, no relationship was found between the tumour and the surrounding major salivary glands. Pleomorphic adenoma of minor salivary glands, although exceedingly rare in this location, could be included in the differential diagnosis.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of a calcified mass in the hyoid/prehyoid region should include:

- ▶ Ectopic thyroid gland tissue/neoplasms, namely papillary carcinomas or psammomatous calcifications in association with thyroglossal cysts or with the pyramidal lobe of the thyroid;
- ▶ Tumours of salivary gland origin, particularly pleomorphic adenoma with chondroid differentiation originating from minor salivary glands or from the adjacent sublingual or submandibular glands;
- ▶ Dermoids;
- ▶ Calcified haematomas (particularly in patients, such as haemophiliacs, with blood dyscrasias);
- ▶ Pseudotumoural calcifications in patients with chronic renal insufficiency;
- ▶ Synovial sarcomas arising from the retrohyoid bursa;
- ▶ Bone/cartilaginous tumours such as chondrosarcomas and osteosarcomas on the malignant side,

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Figure 1 Chondrosarcoma of the hyoid bone. Axial contrast-enhanced neck CT scan at the soft tissue window reveals a relatively well-defined, multilobulated lesion in the submental and prehyoid regions. The lesion has a hypodense soft tissue component and exuberant calcifications with a coalescent ring-and-arc pattern, suggestive of chondroid matrix (arrows). A focal area of contiguity with the right hemibody of the hyoid bone is seen with insufflation and rupture of the outer cortical margin (dashed arrow).

and osteomas, osteblastomas and chondroblastomas in the benign category.

In the case presented, the radiological findings were diagnostic and highly suggestive of a tumour of bone or cartilaginous origin since insufflation and rupture of the hyoid bone's cortical margin could be clearly seen.

Osteomas markedly differ from the tumour we present since they tend to be homogeneous, very radiodense lesions or appear as radiodense lesions demonstrating a central marrow.

Osteoblastomas are associated with cortical expansion, although only rarely with cortical destruction. However, osteosarcomas tend to be highly destructive bony lesions. Osteoid matrix calcifications that are present in these tumours tend to be fluffy, ill-defined and cloud-like, features not present in our case.

The pattern of the tumoural matrix calcification (ring-and-arc and popcorn calcifications) was strongly in keeping with a tumour of a chondroid nature, such as a chondroblastoma or a chondrosarcoma.

A chondroblastoma was less probable since they are benign tumours that tend to present in young patients (<20 years of age) as well-defined lucent lesions sometimes delineated by a sclerotic rim. To the best of our knowledge, only one chondroblastoma has previously been described in the hyoid bone.¹

Any cartilaginous tumour of which the long axis measures more than 2 cm tends to be a low-grade chondrosarcoma and not a benign tumour.²

Moreover the tumour displayed malignant radiological features such as cortical erosion. Therefore, the first diagnosis to consider was a chondrosarcoma.

TREATMENT

A total hyoidectomy and a right submandibulectomy were performed.

The resected specimen contained a well-defined, lobulated, firm whitish and nacreous tumour, with the longest axis measuring 55 mm.

The histopathological diagnosis was of a moderately differentiated, G2, conventional chondrosarcoma of the hyoid bone (figure 4). The right submandibular gland was unremarkable.

Since the distance to the surgical margin was <0.1 mm, adjuvant treatment options were discussed in the multidisciplinary head and neck oncological group and the patient underwent adjuvant radiation therapy. The radiation dose used was of 64 Gy.

OUTCOME AND FOLLOW-UP

Thus far the patient has undergone 1 year of follow-up with no major complications and with no evidence of tumour relapse on clinical examination or on follow-up MRI.

DISCUSSION

Chondrosarcomas are malignant tumours that mainly arise from cartilaginous and bony structures. They account for approximately 11% of all bone neoplasms with the head and the neck being involved in 1–12% of these cases.^{3 4}

Common chondrosarcoma sites of the head and neck are the skull base, the nasal cavity, the maxilla, the mandible and the larynx.

Hyoid chondrosarcomas are exceedingly rare tumours, with only approximately 20 such cases previously reported in the English literature. In contrast to their laryngeal counterparts, which are often associated with breathing symptoms, hoarseness and dysphagia, hyoid chondrosarcomas tend to present as painless, asymptomatic, palpable masses.

They are more likely in men (1.9:1) and the average age of presentation has been reported as 48.5 years.^{4 5}

Figure 2 Chondrosarcoma of the hyoid bone. Axial contrast-enhanced neck CT scan at the bone window depicts the characteristic calcification pattern of chondroid matrix—coalescent rings-and-arcs and 'popcorn-like' calcifications (arrows). Note the relatively preserved morphology of the hyoid bone (dashed arrow).

Figure 3 Chondrosarcoma of the hyoid bone. Sagittal contrast-enhanced neck CT scan at the bone window (A) and at the soft tissue window (B). Note the extension of the tumour into the prelaryngeal space (arrow) and how it pushes the floor of the mouth upward (dashed arrow) and the strap muscles anteriorly (short arrows). The lesion is clearly separated from the sublingual and submandibular glands.

Conventional chondrosarcomas tend to be lobulated masses that enlarge and alter the morphology of the primary site. They are characterised by the formation of hyaline cartilage and their cut surface is hard with grainy calcifications, whereas myxoid variants have a more gelatinous and soft appearance.²

The majority of chondrosarcomas are of low or intermediate grade with a low metastatic rate. Only about 5% are more aggressive high-grade tumours with high metastatic potential, namely to lungs and skeleton.^{2 4 6 7} In contrast to the low and intermediate tumours, these tend to exhibit marked hypercellularity, pleomorphism and mitotic figures.⁷

Radiographically, hyoid chondrosarcomas usually present as a lobulated lytic lesion, with characteristic ring-and-arc or popcorn calcifications.^{4 8}

CT imaging shows a lobulated, destructive, heterogeneously enhancing soft tissue mass, which, in 75% of cases, is associated with matrix calcification.⁵

CT is the best imaging method to depict the characteristic calcification pattern as well as the bony destruction or cortical erosion generally associated with these types of tumours.

MR is helpful in determining the soft tissue extent of the lesion and therefore in planning the surgical procedure.^{5 9}

Figure 4 Chondrosarcoma of the hyoid bone (H&E staining, $\times 40$). Hyoid bone chondrosarcoma characterised by hypercellularity, with cells of various sizes and shapes embedded in the myxochondroid matrix. Focal areas of calcification are also observed.

Chondrosarcomas display a characteristically high signal on T2-weighted images and a low-to-intermediate signal on T1-weighted images. Calcifications appear as hypointense foci best appreciated in gradient echo sequences. Gadolinium-enhanced images may show a diffuse, moderate to avid uptake or a predominantly septal, honeycomb-like and peripheral enhancement.

Fine-needle aspiration and core biopsy must be accompanied by radiological examination of the lesion, as clearly demonstrated in our case. They may sample non-malignant foci or miss zones of higher dedifferentiation. Moreover, when performed without imaging guidance, these tumours may be misdiagnosed as sublingual or submandibular neoplasms, most commonly as pleomorphic adenomas with chondroid differentiation, which are much more common lesions in these particular locations.

Wide surgical excision is the treatment of choice for hyoid chondrosarcomas. Surgery is associated with important side effects such as impaired swallowing, and speech and breathing difficulties.^{4 10} Injury of the submandibular gland has also been reported in a hyoid chondrosarcoma that involved the greater cornua of the hyoid.¹¹ Careful surgical excision is required on such tumours as they may seed during surgical manipulation.

Adjuvant radiotherapy is recommended in high-grade tumours, in partially resected tumours and for palliative treatment. To the best of our knowledge, this is only the fifth time where adjuvant radiotherapy was used to treat hyoid bone chondrosarcoma.⁴

Interstitial photodynamic therapy was performed on a low-grade tumour where complete tumour excision required the sacrifice of the larynx, base of tongue and hyoid bone.⁶ Tumour volume decreased after 5 months, and regrowth was noted on the patient's follow-up, which was successfully treated with a second round of interstitial photodynamic therapy.

Chondrosarcomas are considered to be 'resistant' to chemotherapy.

Adequacy of surgical excision, tumour location and tumour histological grade are the most important prognostic factors.

All patients with hyoid chondrosarcomas that recurred had non-negative surgical margins.⁴ Therefore a positive surgical margin is a strong predictor of recurrence.⁴

Although distant metastasis has been related to higher histological grade tumours, low-grade and moderate-grade hyoid

Reminder of important clinical lesson

bone chondrosarcomas have shown to metastasise to the lungs. Therefore, chest imaging is recommended at the time of diagnosis and during follow-up.⁴

Learning points

- ▶ Chondrosarcomas of the head and the neck represent up to 12% of all chondrosarcomas, with the hyoid bone being an extremely rare site of occurrence.
- ▶ A hyoid bone chondrosarcoma tends to present as a non-specific painless palpable mass in the submental region.
- ▶ Doctors often assume a more common cause and misdiagnose these type of tumours with other more common lesions such as salivary gland neoplasms.
- ▶ CT is the best imaging method to depict bony destruction and cortical erosion as well as the characteristic calcifications of the chondroid matrix, whereas MR is useful in the characterisation of the soft tissue extent of the lesion, helping surgical planning.
- ▶ Wide surgical excision is the treatment of choice, since inadequate excision is strongly associated with relapse, particularly in higher histological grades.

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