Case report

Magnetic resonance imaging of neck chondrosarcoma: A case report

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Abstract

Chondrosarcoma is a malignancy rarely encountered in the head and neck. The reported prevalence of chondrosarcoma involving the head and neck region has a range of 1–12%. In our case report, we demonstrate a chondrosarcoma of the left lower neck region by means of magnetic resonance imaging. We were not able to show its exact origin and considered the possibility of a rare extraosseous origin. It presented as a slowly growing, painless mass and showed characteristic calcification and high signal intensity on long TR, long TE sequences. It caused left jugular vein compression and left subclavian artery kinking due to mass effect. Histopathologic examination revealed a low-grade mesenchymal myxoid chondrosarcoma. In this study, we review the clinical presentation, imaging features, management, and treatment procedures of these rare tumors.

Keywords: Chondrosarcoma; Head and neck; Magnetic resonance imaging; National cancer data base; Tumor

1. Introduction

Chondrosarcomas are a heterogeneous group of malignant tumors of cartilaginous origin. Chondrosarcoma constitutes approximately 11% of all primary malignant bone tumors and, after osteosarcoma, is the second most common sarcoma arising in bone [1,2]. Although chondrosarcoma is the second most common primary spindle cell tumor of bone, it is uncommon in the head and neck region. The head and neck region has been reported as the site of origin of chondrosarcomas in a range from 1 to 12% [3–5]. In our case report, we present a chondrosarcoma case which originated in the neck region and particularly emphasize the magnetic resonance imaging (MRI) features of these tumors.

2. Case report

A 56-year-old man was admitted to our hospital complaining of a painless mass in his neck region on the left side which had increased in size during the past 6 months. He was previously in good health and except for this mass his physical examination was unremarkable. His personal history is below:

Age, 56; sex, male; race, Caucasian; height, 1.75 cm; weight, 88 kg; social status, married; alcohol habit, social drinker; smoking status, heavy smoker.

The patient was referred to the our MRI division and we performed axial spin-echo T 1, axial fast spin-echo fat-suppressed T 2, coronal fast spin-echo IR (inversion recovery) weighted images. Following 40 cm³ gadolinium DTPA administration, we also obtained FSPGR/80 weighted and 3D MR angiographic images through the neck region by using a 1.5 T superconducting magnet (GE, Signa, Milwaukie, Wisconsin, USA.)

The images revealed a 9×7 cm mass in the left lower neck region. It showed isointense signal intensity on T 1 and strong high-signal intensity on T 2 and IR images (Fig. 1). Heterogeneous calcification was present within the mass. There was no definitive origin of the mass and it did not seem as if there was a connection between the mass and the cervical vertebrae as a primary site of the tumor.

The left jugular vein was compressed by the mass, but the carotid arteries were patent, and were not involved. The tumor showed a patchy enhancement (Fig. 2).
The left subclavian artery appeared to make a sharp kink in its proximal portion which was most likely due to compression by the mass. But no definitive stenosis or occlusion of the left subclavian artery was seen (Fig. 3).

In the differential diagnosis, because of the patient’s age, clinical presentation, and MRI features of the mass (i.e. calcification, very high T2 signal) we initially thought of chondrosarcoma. An incisional biopsy under local anesthesia was performed and histopathologic analysis revealed grade 1 (low grade, well-differentiated) mesenchymal myxoid chondrosarcoma as the final diagnosis.

The patient was referred to the ENT (Ear–Nose–Throat) division where a radical tumor excision was performed without complication. The patient is being followed regularly both clinically and radiologically by means of MRI every 6 months.

3. Discussion

Chondrosarcoma is a malignant tumor in which the tumor cells form chondroid (cartilage) but no osteoid bone [6]. Osteosarcoma, which may also produce cartilage, is discriminated from chondrosarcoma by the presence of osteoid as well [7].

Chondrosarcomas are slow-growing, but locally aggressive neoplasms with a propensity for local recurrence. Approximately two-thirds of the patients are men [8] and the most common presentation is a slowly growing mass in an adult male, older than 40 years. In our case, the patient was a 56-year-old man with a painless mass in the lower neck which increased in size over months.

Most chondrosarcomas arise from bone, but they can also develop from extrasosseous tissues. We could not demonstrate any relationship between the tumor and neighbouring cervical vertebrae within MRI limits so there was a possibility of extrasosseous origin. Literature shows that chondrosarcoma in an extrasosseous location without associated bony destruction is exceedingly rare [9].
Chondrosarcoma can develop in any bone formed in cartilage, the most common sites being the pelvis, femur and humerus, followed by the tibia, ribs, scapula, skull, vertebrae, and sternum. The primary sites in the head and neck are the maxilla, cervical vertebrae, and mandible, with skull, sphenoid and ethmoid sinuses, frontal sinus, nasal septum, and orbits involved less frequently.

Malignant cartilaginous tumors arise de novo in bone or are superimposed on benign cartilaginous tumors, such as osteochondromas or enchondromas. Secondary chondrosarcomas rarely develop in patients with solitary benign cartilaginous lesions but more commonly develop in patients with multiple hereditary exostosis or multiple enchondromatosis [10]. Chondrosarcoma also occurs in association with malignant conditions, including melanoma, fibrosarcoma [11] and leukemia [12].

In fact, sarcomas in general, occur uncommonly in the head and neck, comprising less than 1% of all head and neck malignancies [13]. In a recent review of 229 head and neck sarcomas seen at the University of California, Los Angeles, Medical Center between 1955 and 1988, 18 (8%) were chondrosarcomas, ranking fourth in incidence behind rhabdomyosarcoma (20%), fibrosarcoma (13%), and angiosarcoma (12%) [14].

The typical radiographical appearance of chondrosarcoma is that of a predominantly lytic lesion with sclerotic margins, endosteal scalloping, and associated stippled or popcorn calcification. Cortical destruction occurs late in the disease and periosteal new bone formation is rare [15].

MRI appearances of chondrosarcoma have not been well documented. These tumors often return heterogeneous signals because of the mixture of soft tissue, cartilage, calcification, and even hemorrhage [8]. Chondrosarcomas usually have a bright signal intensity on T2 weighted images. Areas of calcification within the tumor give rise to a decreased signal intensity on all sequences [8]. Such calcification does not enhance following administration of intravenous paramagnetic contrast agents, leading to underestimation of actual tumor size [16].

Optimal management in these rare tumors remains controversial. Adequate surgical resection is the gold standard. Due to the locally invasive nature of chondrosarcomas and the inherent anatomic restrictions in the head and neck limiting adequacy of surgical margins, local recurrence after surgery alone is a frequent problem [17,18]. In the largest series to date (representing 151 cases of chondrosarcoma of the head and the neck) Weiss and Bennett [19] noted that “metastasis is not common and generally occurs late, usually following multiple surgical manipulations.”

The overall low incidence of regional (5.6%) metastases in the NCDDB (National Cancer Data Base) series suggests that radical neck dissection is not indicated [20]. This NCDB analysis demonstrated that decreased survival was statistically associated with advanced stage, higher grade, and the histological subtypes, myxoid or mesenchymal [20].

The efficacy of adjuvant radiotherapy is unclear because it was used in fewer than 25% of patients and was used primarily for tumors with poor prognostic variables, such as high grade, myxoid or mesenchymal subtype, and positive surgical margins. The role of chemotherapy for head and neck chondrosarcoma is even less clear, with no consistent indications that are supported scientifically.

The disease-specific survival period of 5 (87.2%) and 10 years (70.6%) for chondrosarcoma of the head and the neck, as calculated from the NCDB data, is higher than reported by most other publications [4,21].

4. Summary

Although chondrosarcoma is a rarely encountered tumor in the head and neck region, it must always be considered in the differential diagnosis, especially when presenting with a slowly growing mass in the middle-aged group of male patients. It characteristically shows high T2 signal intensity on MR images and optimal treatment is surgery.

References


Ahmet Mesrur Halefoglu was born on 06/06/1963 in Antakya, Hatay, Turkey. In 1986, he served as the Istanbul University Cerrahpasa Medical Faculty. Residency: Sisli Etfal Training and Research Hospital, 1987. He is currently a Specialist at Radiology, Sisli Etfal Training and Research Hospital.