

Naso-Sinus Chondrosarcoma: A Case Report¹

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Chondrosarcomas are malignant tumors of the cartilage that rarely involve the sinonasal region. Here, we describe a case of histologically verified naso-sinus chondrosarcoma in a 40-year-old female presenting with nasal stuffiness and anosmia. The tumor presented on computed tomography (CT) as an expanding soft tissue mass with bone destruction and pressure erosion. The magnetic resonance images (MRI) of the tumor demonstrated high signals on T2-weighted images with nodular and papillary enhancement along the periphery on T1-weighted images with contrast enhancement. The presence of these typical imaging features should be very helpful in diagnosing chondrosarcoma involving the sinonasal region.

Index words : Sarcoma

Paranasal sinuses

Nose, CT

Nose, MR

Chondrosarcomas are malignant, slow growing cartilaginous tumors that are most commonly found in long bone, but can originate in other bones or even soft tissue (1). Various reports estimate that 3% - 12% of all chondrosarcomas occur in the head and neck region (2 - 5). These accounted for approximately 0.1% of all head and neck cancers (3). The head and neck bones, such as the maxilla and mandible, are predominantly involved in these chondrosarcomas, and the sinonasal region is rarely involved (3 - 5). Although rarely presenting in the sinonasal region, the characteristic imaging findings of high water content on T2-weighted images and typical peripheral enhancement on enhanced T1-weighted im-

ages can lead to correct diagnosis. Here, we present a case of histologically verified naso-sinus chondrosarcoma with typical imaging findings.

Case Report

A 40-year-old woman presented with left nasal obstruction and anosmia for a week. The patient had left optic nerve palsy for 3 years and left superior orbital swelling for 5 months. Clinical examination revealed expansion of the uncinate process of the left middle meatus. Computed tomography (CT) and magnetic resonance imaging (MRI) of the paranasal sinuses and nasal cavity were performed. CT revealed an expansile lobular soft tissue mass involving the upper portion of the left nasal cavity, left ethmoid sinus, and sphenoid sinus. It extended laterally to the left orbit, displacing the medial rectus muscle and superior ophthalmic vein, and to the apex of the left orbit and superior orbital fissure. CT with a bone window setting showed bone destructions

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Received March 22, 2006 ; Accepted July 31, 2006

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involving the left orbit roof and lesser wing of the sphenoid bone (Fig. 1A). Some calcifications or remaining bony fragments were seen within the mass without prominent chondroid calcification, which was the typical imaging feature of the chondrosarcoma (Fig. 1A, B). MRI showed a high signal intensity mass with focal low signal intensity at the medial periphery of the mass on T2-weighted images (TR/TE, 4000/99) (Fig. 1C, D). On T1-weighted images (TR/TE, 570/14), most of the mass showed heterogeneous low signal intensity (Fig. 1E). On T1-weighted images with contrast enhancement (TR/TE, 532/11), diffuse peripheral enhancement was seen along the inside of the mass (Fig. 1F). Typically, a nodular and papillary enhancement pattern was observed. This enhancement was most prominent at the

focal low signal intensity region on T2-weighted images. Biopsy was performed at the mass in the left nasal cavity and ethmoid sinus. The histopathologist reported a cartilage matrix that formed irregularly-shaped lobules with hypercellularity and mild to moderate atypism on microscopic examination, thus diagnosing a well-differentiated chondrosarcoma (Fig. 1G, H).

Discussion

Chondrosarcomas arise in tissue known to be formed of cartilage or in bone that is preceded by a cartilaginous plate which is later replaced by bony tissue (6). An origin from the nasal septum would explain their presence in the nasal cavity and in bones that can ossify in carti-

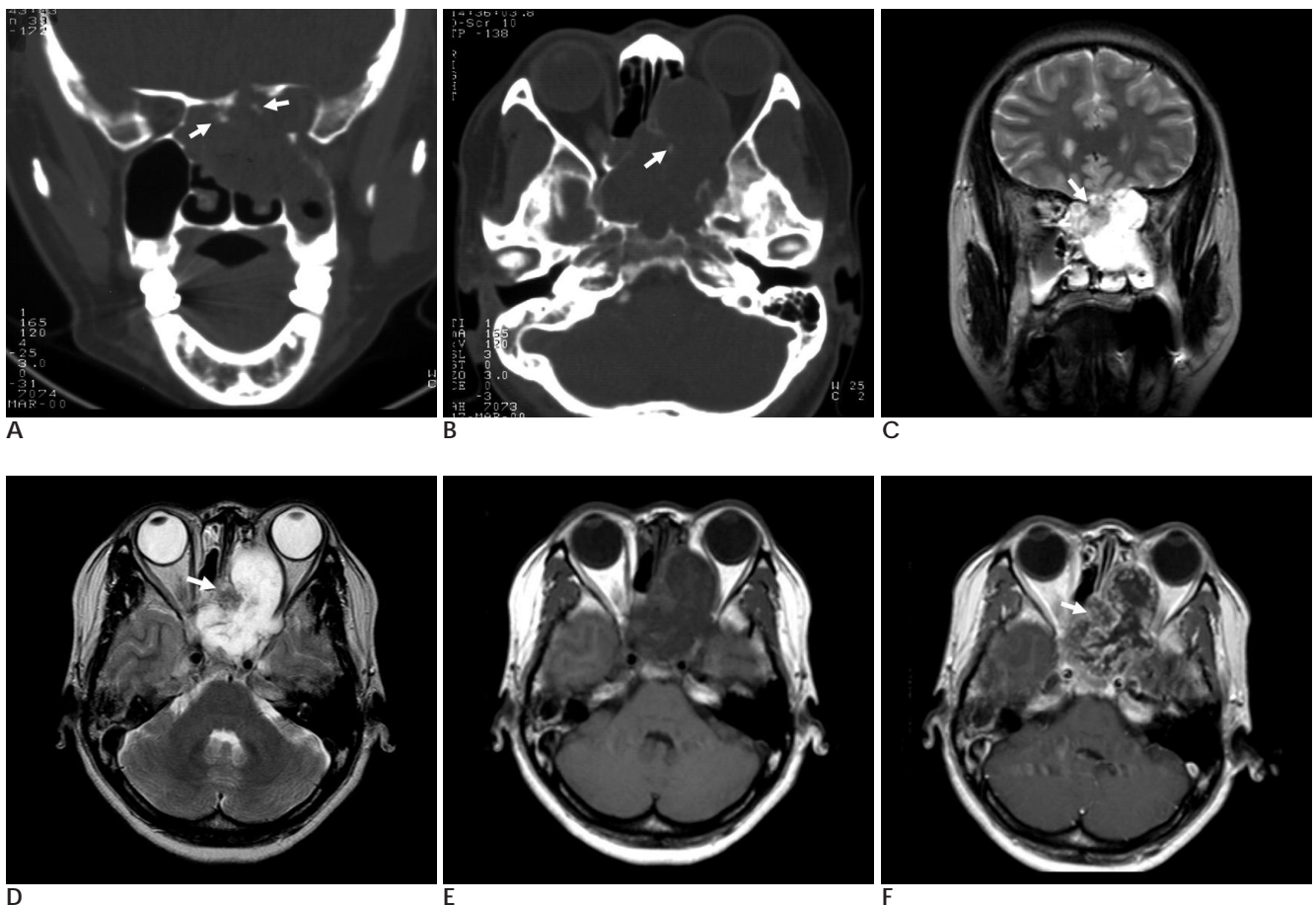


Fig. 1. A 40-year-old female with chondrosarcoma in the sinonasal region.

A, B. Coronal (Fig. 1A) and axial (Fig. 1B) CT scans with bone window setting reveal an expansile soft tissue mass involving the upper portion of the left nasal cavity, left ethmoid sinus, and sphenoid sinus. Bone destructions and erosive changes involving the lesser wing of the sphenoid bone and left orbital roof are shown. Some calcifications or remaining bony fragments (white arrows) are noted within the mass.

C, D. Coronal (Fig. 1C) and axial (Fig. 1D) T2-weighted MRI demonstrate a lobulated, contoured, bright signal intensity mass with focal low signal intensity (white arrows) at the medial periphery of the mass.

E. On axial T1-weighted MRI, this mass shows heterogeneous low signal intensity.

F. On contrast-enhanced axial T1-weighted MRI, nodular and papillary enhancement along the inside of the peripheral wall is demonstrated. This enhancement is most prominent at the focal low signal intensity region on T2-weighted images (white arrow).

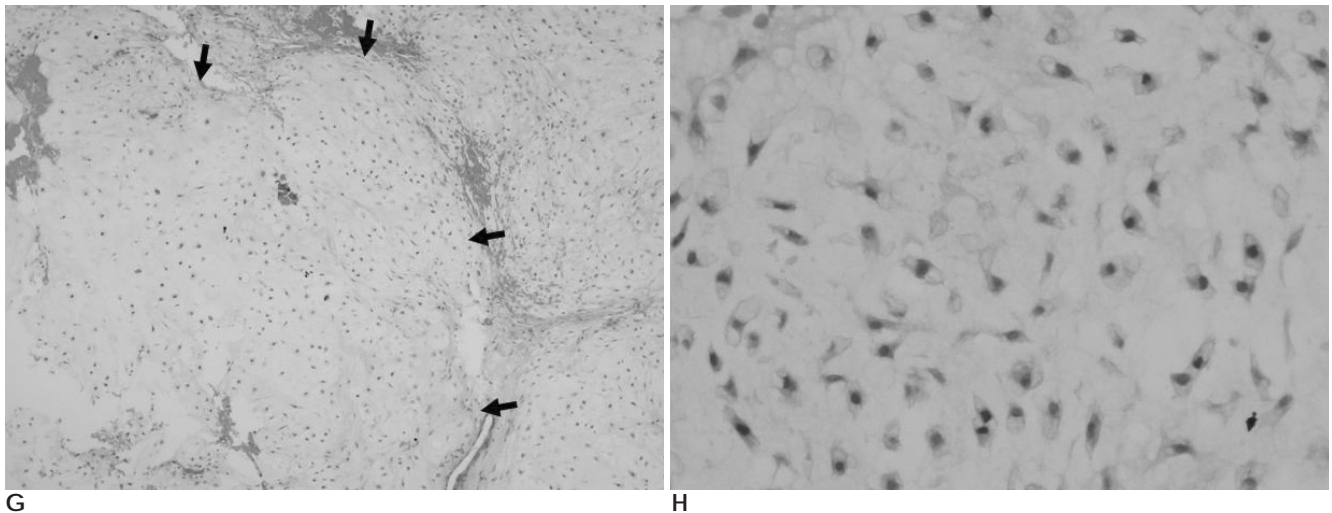


Fig. 1. G. Photomicrograph of the biopsy specimen (hematoxylin-eosin stain, original magnification $\times 40$) showing an abundant blue to grayish cartilage matrix that formed irregularly-shaped lobules (black arrows). In general, chondrosarcoma shows increased cellularity when compared to enchondroma.
H. On high magnification (original magnification $\times 200$), chondrocytes show mild to moderate atypism containing enlarged, hyperchromatic nuclei. Occasional binucleation and mitotic figures are also found.

lage such as the sphenoid.

However, it is not yet completely understood how chondrosarcomas develop in the remaining paranasal sinuses without any cartilage during their development. One suggestion is that the tumor may result from spread from neighboring cartilaginous tissue. Alternatively, it has been pointed out that the multidirectional differentiation of primitive mesenchymal cells gives rise to these tumors and accounts for their appearance in tissue that does not normally harbor cartilage (7).

There are numerous pathologic types of primary chondrosarcomas, including conventional intramedullary, clear cell, myxoid, mesenchymal, and dedifferentiated chondrosarcomas. Although the pathologic appearance varies with the specific lesion type, chondrosarcomas grow with a lobular type architecture, resulting in hyaline cartilage nodules with high water content and peripheral enchondral ossification (1).

Macroscopically, they appear lobulated, and are composed of bluish-white masses of tissue. The consistency may be hard and the tumor may show calcification or ossification, and may contain soft gelatinous areas. Microscopically, the neoplasm consists of cells showing varying degrees of malignancy. Lobularity is also a feature at the microscopic level, and high cellularity is often seen at the periphery of the lobules (8).

Appearance on imaging directly reflects the histopathology of the tumor. The distinctive imaging features of chondrosarcomas are shown optimally by

combined CT and MRI. Both CT and MRI illustrate the high water content of the lesion through low attenuation and very high signal intensity on T2-weighted images (1). CT is optimal for detecting the matrix mineralization, particularly when it is subtle or when the lesion is located in an anatomically complex area (8).

On the CT scan, the tumor presents as a lobulated, contoured hypodense mass expanding and destroying bone with nodular or plaque-like calcification and often shows the characteristic appearance of a ring and arc pattern of calcification. This characteristic chondroid calcification, causing enchondral ossification around the margins of the cartilaginous lobules, usually allows for a confident radiologic diagnosis of a cartilaginous lesion (1, 8).

MRI shows low intensity on T1-weighted images, high intensity on T2-weighted images, and inhomogeneous enhancement on enhanced T1-weighted images. The distinctive features included very low or absent enhancement centrally in the tumor mass and strong enhancement at the periphery on enhanced T1-weighted images (8).

In our case, although the typical ring and arc pattern of calcifications was not present, the peculiar peripheral enhancement pattern with nodular and frond-like papillary enhancement could be a diagnostic clue for differentiating chondrosarcoma from other malignant tumors involving the sinonasal region.

Clinical and radiographic differential diagnosis in-

cludes other naso-sinus tumors presenting as an expanding mass with calcifications such as chondroma and chordoma.

Chondroma often may be difficult to distinguish from a well-differentiated chondrosarcoma based on pathology alone. On CT scan, bony erosion is absent in chondromas, and lesions greater than 2 cm in size are considered chondrosarcomas despite a benign appearance on histology. The differential diagnosis of chondroid chordoma versus chondrosarcoma is difficult with radiographic imaging alone. Tumors involving the clivus are generally chordomas (4, 9).

The most definite treatment for chondrosarcoma is total surgical excision, but posteriorly located tumors involving the sphenoid and skull base cannot be totally resected. Local recurrence is not uncommon in high-grade tumors. Chondrosarcomas grow and invade adjacent structures slowly, and metastasize to the lung at a later time, as it is the most common site (1).

Radiation therapy may be employed for higher grade conventional chondrosarcomas (grade 2 and 3) that are incompletely removed or are in locations that are surgically inaccessible such as the skull base (1). Chemotherapy is generally given before definitive surgical resectioning. Overall survival at 5 years for all grades combined is approximately 44% to 81% (1 - 5).

Our patient presented with typical symptoms and characteristic radiographic findings indicative of chon-

drosarcoma arising from the nasal cavity and paranasal sinus. Of interest in this case is the unusual location of the tumor and the typical imaging findings of peripheral nodular and papillary frond-like enhancement. Imaging studies such as CT and MRI are essential in making the correct diagnosis before biopsy and staging.

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