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Lung: Case Report

Extra-axial Chordoma of the Sternoclavicular Joint

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Chordomas are rare, slowly growing, aggressive primary bone tumors that originate from notochord remnants and occur almost exclusively in the axial skeleton. Here, we describe a patient with an enlarging right-sided neck mass that was later diagnosed as a sternoclavicular joint chondroid chordoma. En bloc surgical resection was accomplished with negative margins. The patient continues to do well 15 months after resection without radiotherapy. This case highlights the importance of considering a wide differential in managing sternoclavicular joint tumors.

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Chordomas are rare, slowly growing, aggressive primary bone tumors that originate from notochord remnants and almost always occur in the axial skeleton.[1,2] The most common sites for axial chordomas are the dorsal sacrum (50%), skull base (35%), and vertebral bodies (15%).[2] Very few cases of extra-axial chordomas exist, and none to our knowledge have been reported in the sternoclavicular joint.

In this case, we describe a 69-year-old man with a 48 pack-year smoking history and chronic obstructive pulmonary disease who presented to his primary care physician with an enlarging right-sided neck mass. The patient first noticed the mass 3 weeks before presentation. Review of systems was positive for dysphagia, mild dyspnea, and unintentional 20-pound weight loss during the previous 3 months. Physical examination was remarkable for a hard, nontender, right-sided neck mass with no overlying skin erythema.

Computed tomography imaging of the chest demonstrated a 4 × 3.7-cm mass centered in the superior mediastinum, posterior to the right sternoclavicular joint. The mass had heterotopic calcifications and appeared separate from the right sternocleidomastoid muscle and right thyroid lobe. Initial imaging results were concerning for a cartilaginous tumor with differential diagnoses including osteochondroma and low-grade chondrosarcoma. The patient underwent ultrasound-guided fine-needle aspiration, and pathologic analysis was significant for tumor cells positive for cytokeratin AE1/3, EMA, S100, brachyury, and INI-1, favoring a diagnosis of chordoma (Figure 1).

Magnetic resonance imaging of the spine and head was performed to rule out a primary lesion in the axial skeleton, and no other lesions were found. No mass or lymphadenopathy elsewhere in the neck was noted (Figure 2).

Because of the aggressive nature of chordomas, an en bloc surgical resection was planned, with the goal of an R0 resection. During the procedure, the mass was found to be well demarcated, adjacent to the trachea and right internal jugular vein, but without any invasion to surrounding structures. The medial third of the right clavicle, the first rib, and a wedge of the manubrium were resected en bloc. The superior margin of the pectoralis major muscle was sutured to the sternocleidomastoid muscle. Final pathologic analysis was consistent with chondroid chordoma, and all histologic margins were negative.

Given the R0 resection, the patient's age, and functional status, adjuvant radiotherapy was not offered to the patient. Instead, plans were made to observe the patient with imaging every 6 months for 3 years and then yearly thereafter. However, should there be any recurrence of disease, radiotherapy will be considered. The patient remains free of any recurrence after 15 months of follow-up.

COMMENT

We present an exceptional case of an extra-axial chordoma noted in the sternoclavicular joint. The patient in this case underwent R0 resection of the tumor, without adjuvant radiotherapy, and continues to remain recurrence free after 15 months of follow-up.

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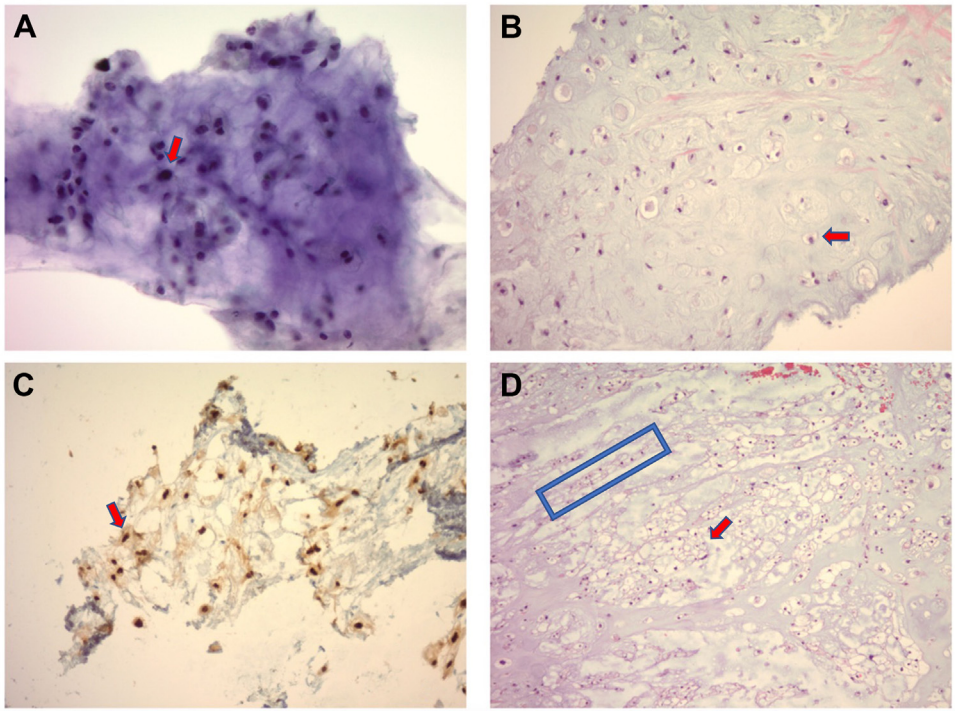

FIGURE 1 Fine-needle aspiration biopsy, core biopsy, and resection specimens of chordoma from right sternoclavicular joint. (A) Fine-needle aspiration biopsy specimen with Papanicolaou stain shows cells with dark nuclei and abundant cytoplasm with occasional vacuolation (arrow), embedded in magenta stroma. (B) Core biopsy specimen with hematoxylin and eosin stain shows cells located in lacunar-like spaces (arrow), surrounded by hyalinized cartilaginous matrix. (C) Immunohistochemical staining of the core biopsy specimen shows strong positive nuclear brachyury stain (arrow). (D) Resected specimen with hematoxylin and eosin stain shows areas with corded arrangement of tumor cells (box), representing conventional chordoma. Physaliphorous cells are also seen (arrow).

Chordomas are exceedingly rare, and this patient's tumor location is unique because chordomas almost exclusively affect the axial skeleton.[2] In a case series of 131 chordomas, only 3 (2.3%) were extra-axial, and none involved the sternoclavicular joint.[3] Another report that presented 20 cases of extra-axial chordomas did not describe any chordomas affecting the sternoclavicular joint.[4]

Chordomas provide a diagnostic challenge because they are rare and difficult to differentiate from more common primary bone malignant neoplasms. On imaging, chordomas can be similar to chondrosarcomas because both types may appear as soft tissue masses with variable amounts of calcifications.[5] Therefore, histologic evaluation is crucial for proper diagnosis, and brachyury can serve as a strong diagnostic tumor marker.[6]

Proper diagnosis is crucial because chordomas have a greater rate of recurrence and worse 5-year survival than chondrosarcomas. Thus, proper management of chordomas requires more aggressive surgical resection, often followed by adjuvant radiotherapy. The frequent need for radiotherapy may be due to the fact that more than half of patients with chordomas have some degree of residual tumor on imaging studies.[7] In comparison, management of chondrosarcomas typically involves surgical resection alone. However, if an R0 resection is achieved, radiotherapy may not be necessary for chordomas, as was seen in the case we present.[8]

This case offers several important lessons regarding management of bone masses of the sternoclavicular joint. First, it demonstrates the importance of considering a wide differential diagnosis for bone tumors, which should now include chordomas. In addition, tissue diagnosis with immunohistochemistry should be obtained, specifically with brachyury. Diagnosis of chordomas before an operation is important because these tumors require more aggressive surgical resection than other tumor types, and adjuvant radiotherapy should be considered if an R0 resection is not accomplished.

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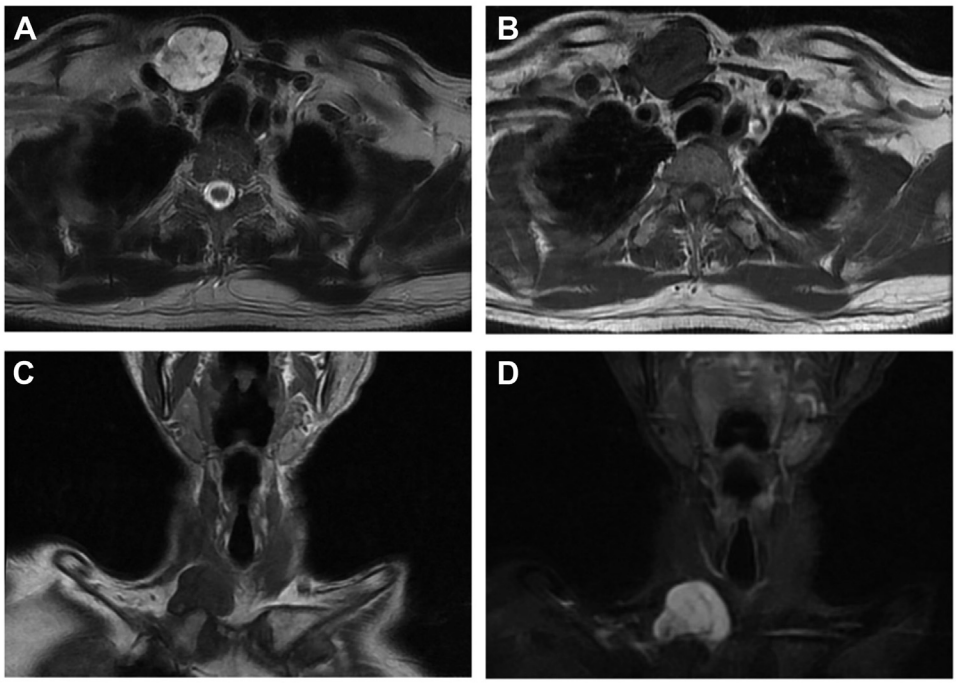

FIGURE 2 Magnetic resonance imaging of neck soft tissue. A mass lesion in medial aspect of right lower neck, along superior margin of medial clavicle and sternoclavicular articulation, measuring 3.3 × 4 × 3 cm (right-left × anterior-posterior × superior-inferior). The lesion is thinly marginated, mildly hypointense to muscles on T1-weighted images, and hyperintense on T2-weighted images and shows heterogeneous enhancement after administration of contrast material. (A) Axial T2 weighted. (B) Axial T1 weighted. (C) Coronal T1 weighted. (D) Coronal T2 weighted.

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