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Radiologic Findings of Bilateral Elastofibroma Dorsi: A Case Report[1]

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Elastofibroma dorsi is a rare, slow-growing, ill defined soft tissue tumor that's typically found between the inferior scapula and chest wall. The characteristic findings on ultrasonography, MRI and CT usually allow the correct diagnosis and so prevent unnecessary surgical procedure. We experienced a case of bilateral elastofibroma dorsi in an 87-year-old man, and we report on this case along with a review of the literature.

Index words : Soft tissue, fibroma
Soft tissue, US
Soft tissue, CT
Soft tissue, MR
Soft tissue, neoplasms

Elastofibroma dorsi is a rare, non-encapsulated benign soft tissue lesion of the chest wall; it's characterized by a proliferation of fibrous tissue with elastin and it occurs most often in the infrascapular area of elderly women. This lesion usually arises unilaterally beneath the rhomboid major and latissimus dorsi muscles and subjacent to the inferior angle of the scapula (1). However, bilateral involvement has been reported in 10 to 60% of cases (2 - 4). Magnetic resonance image (MRI) and computed tomography (CT) clearly show the layered pattern of the fibrous tissue and fatty tissue, as well as the highly characteristic location of the mass (1, 3, 5, 6). Ultrasonography (US) can also contribute to the diagnosis (7).

We report here on a case of an 87-year-old man with bilateral elastofibroma dorsi, and we describe the radiologic and pathologic findings.

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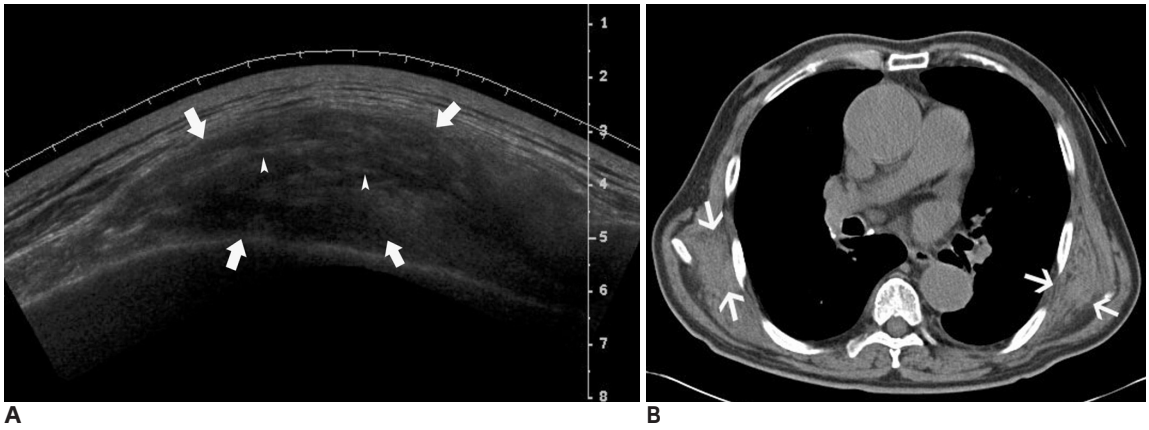
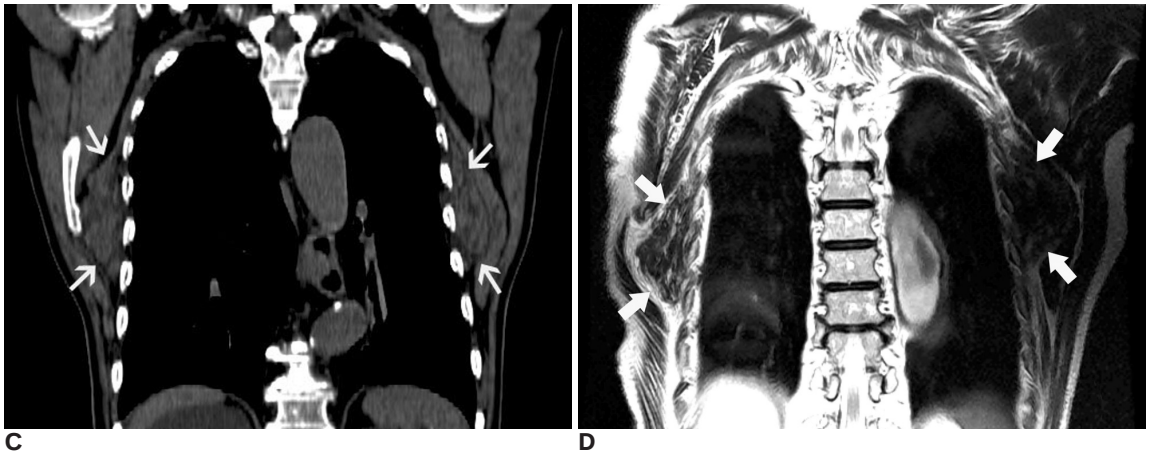
Case Report

An 87-year-old man presented with slowly enlarging painless masses that he had experienced for several months, and these masses were in the bilateral subscapular spaces. He had undergone endoscopic mucosal resection for gastric cancer 2 months previously. Physical examination revealed the nontender, firm bilateral masses in the subscapular area, and the mass were symmetric and measured about 5×5 cm. His laboratory data was within the normal limits.

Ultrasonography was performed using a HDI 5000 scanner (ATL, Bothell, WA, U.S.A.) equipped with a 5-12 MHz linear array transducer. During the sonographic examination, the patient was placed in the prone position with his arms elevated and adducted. We clearly noticed the prominent bilateral chest wall masses that were deep to the inferior pole of the scapula. Ultrasonography showed poorly defined heterogeneous hypoechoic soft tissue masses with interspersed linear echoes in the bilateral infrascapular regions (Fig. 1A).

Both infrascapular regions were scanned by CT





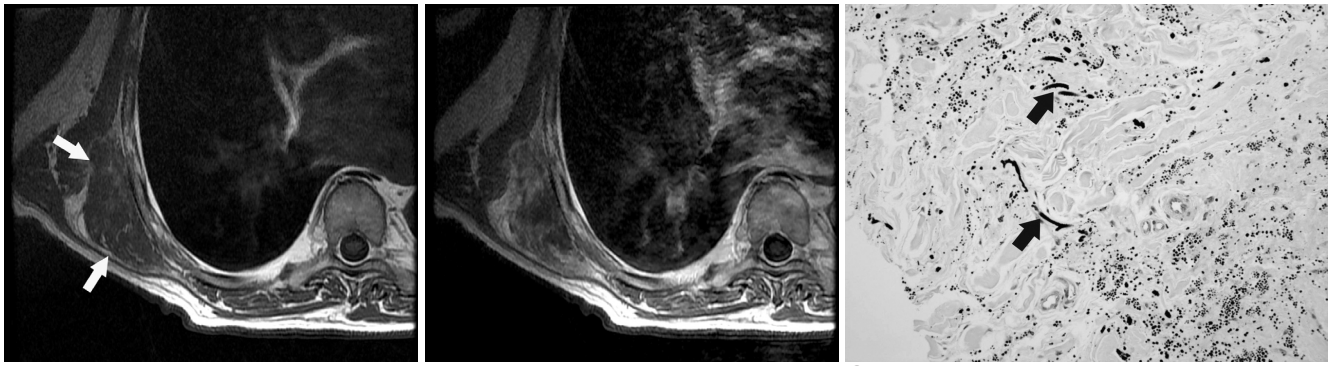


Fig. 1. A. Sonogram shows a poorly defined right back mass (arrows) at the inferior angle of the scapula, and the mass has interspersed linear echoes (arrowheads).

B. Axial noncontrast CT scan shows bilateral lentiform masses (arrows), with attenuation values similar to those of the neighboring muscles, and linear interspersed hypodense areas that are suggestive of fatty streaks within the mass.

C. Coronal reformation CT image shows bilateral soft tissue masses (arrows) in the characteristic location between the rib cage and the overlying serratus anterior and latissimus dorsi muscles.

D. Coronal fast spin echo T2-weighted MR image shows the bilateral masses (arrows) in the subscapular lesions with low signal intensity and high signal intensity linear strands.

E. Axial spin echo T1-weighted MR image shows a intermediate signal intensity mass lesion (arrows) in the right chest wall with interspersed lines of high signal intensity; this is consistent with fat.

F. Moderate enhancement of the mass was detected after injecting gadolinium.

G. The histologic section shows numerous globular and linear elastic fibers (arrows) within a collagenous matrix (Elastic stain, × 200).

(Somatom Sensation 64; Siemens, Forchheim, Germany). The CT examination without IV contrast material showed bilateral poorly circumscribed, heterogeneous soft tissue masses, with attenuation values that were similar to those of the neighboring skeletal muscles, and linear low density fatty strands (Fig. 1B, C).

Magnetic resonance (MR) imaging of the upper thorax was performed with a 1.5 Tesla magnet MR unit (Signa Exite; GE Medical System, Milwaukee, Wisconsin, U.S.A.). The axial, sagittal and coronal spin echo T1-weighted images (TR/TE: 500/12 ms) and the fast spin echo T2-weighted images (TR/TE: 4000/123 ms) were obtained. Then, after IV gadolinium administration, the T1-weighted images were also obtained. MR examination revealed lenticular, poorly circumscribed, unencapsulated masses that had signal intensity similar to the adjacent skeletal muscle on both the T1 and T2-weighted images. The strands of fatty tissue were seen as high signal intensity on the T1-weighted sequences and as intermediate to high signal intensity on the T2-weighted images (Fig. 1D, E). The masses appeared moderately enhanced on the postgadolinium administrated MR images (Fig. 1F).

Although the diagnosis of elastofibroma was considered because of the clinical and imaging findings, the patient underwent percutaneous needle biopsy to exclude such malignant tumors as metastasis or soft tissue sarcomas. Histologically, the tumors exhibited the characteristic structure of elastofibroma, in which streaks of fatty tissue alternate with strands of hyalinized collagen admixed with scattered fibroblasts. The presence of deeply stained enlarged, hypereosinophilic, refractile elastic fibrils was confirmed by elastic staining (Fig. 1G). The final pathologic diagnosis was elastofibroma dorsi.

Discussion

Elastofibroma dorsi is a rare, benign, soft tissue tumor of the periscapular area. This disease entity was first reported in 1961 by Järvi and Saxęn (8).

The greater majority of elastofibromas occur in the infrascapular or subscapular region beneath the rhomboid major and latissimus dorsi muscles (1). The other reported locations are the hand, foot, greater trochanter, ischial tuberosity, the infraolecranon area, the cervical epidural space, sclera, rectum and stomach (2).

Elastofibroma mainly occurs in older individuals with a mean age of 70 years (2). The lesion occurs more frequently in women with a ratio as high as 13:1 (1). The tumor is usually unilateral; however, bilateral involvement has been reported in 10 to 60% of cases (2, 4). Elastofibroma dorsi is asymptomatic in over 50% of the cases. When symptoms are present, they are usually mild, consisting of swelling, joint stiffness, a clicking sensation or pain when moving. If the patient has a non-palpable small tumor, then an elastofibroma cannot be detected when the patient stands in a normal position because the scapula may overlie the mass and mask it. The patient positioned with the arms elevated forward and adducted allows much better depiction of the mass (7, 9).

Elastofibroma dorsi is a slow growing tumor of the connective tissue. It has been suggested that elastofibroma is a chronic reactive process between the chest wall and scapula that's caused by repeated mechanical friction. In fact, many patients have a past occupational history of heavy manual labor. Yet recent studies have suggested that abnormal neoelastogenesis rather than degeneration of the preexisting elastic fibers is the main pathogenetic factor (10).

Histologically, elastofibroma exhibit a characteristic structure in which streaks of fatty tissue alternate with strands of fibrous tissue. The histologic analysis of the core biopsy shows a mixture of eosinophilic collagen, elastic fibers and adipose tissue (2, 10). The elastic stain shows dark brown to black elastic fibers with an elongated or globular appearance and serrated margins (2, 10).

The imaging features of elastofibroma dorsi have been recently characterized. Plain radiographs are usually of little value in diagnosing elastofibroma dorsi, but they occasionally show a soft tissue mass displacing the scapula. The sonographic appearance of elastofibroma dorsi consists of arrays of interspersed linear or curvilinear hypoechoic strands against an echogenic background (5, 7). Theses findings reflect the alternating pattern of fibroelastic tissues and fatty streak, and this pattern can also be detected with other imaging technique.

On CT examination, Elastofibroma dorsi is typically seem as a well or poorly defined, non-encapsulated, heterogeneous soft tissue mass with attenuation similar to that of skeletal muscle, and the mass contains linear steaks of fat attenuation (1, 3, 5, 6). MRI is the imaging modality of choice, as it clearly shows the characteristic layered pattern of fibrous tissue and fatty tissue (6). The dense fibrous connective tissue shows intermediate signal intensity on the T1-weighted images and relatively low signal intensity on the T2-weighted images, which

is nearly identical to that generated by skeletal muscle. The foci of interspersed fatty tissue show high signal intensity on T1-weighted images and intermediate signal intensity on T2-weighted images. The MR imaging after gadolinium administration showed a variable enhancement pattern that ranges from subtle to marked enhancement (3, 6).

The radiographic features of elastofibroma dorsi are usually diagnostic, but the differential diagnosis of a periscapular soft tissue mass lesion that displays decreased to intermediate attenuation and signal intensity, which is similar to that of skeletal muscle on the CT and MR imaging, includes fibrous tissue tumors (extraabdominal desmoid and fibroma) as well as malignancies such as metastases and sarcomas. Similarly, other differential diagnostic options that have increased signal intensity on T1-weighted MR images include lipoma, liposarcoma, hemangioma and hematoma. In this case, although the imaging findings were relatively typical, biopsy was performed to rule out such malignant tumors as metastasis because the patient presented with a previous history of stomach cancer.

Earlier studies have recommended complete resection for the diagnosis and treatment. Surgical excision is recommended when elastofibroma dorsi causes functional disability, symptoms of compression or pain, or when the tumor dimensions exceed 5cm in diameter (4). Only a few cases of local recurrence have been reported after incomplete resection (2). The current studies suggests that a definitive pathologic diagnosis of this entity can be made, based on the presence of aberrant elastic fibers, by fine needle aspiration biopsy (FNAB). Mojica and Kuntzman reported that fine-needle aspiration represents the simplest and quickest method of obtaining a definite diagnosis of elastofibroma dorsi (10).

In conclusion, elastofibroma dorsi is a slow growing, benign soft tissue tumor that is typically located in the periscapular area. The typical radiologic appearance of elastofibroma dorsi is a poorly circumscribed soft tissue mass with attenuation values similar to those of the neighboring muscles to, and the mass is interspersed with linear strands of fat in a characteristic location. Although its incidence is very low, the physician's awareness of this tumor's characteristic appearance and location will decrease the misdiagnosis of these lesions as malignancies and so avoid unnecessary aggressive surgical procedures.

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흉벽에 발생한 양측성 탄력섬유종의 방사선학적 소견: 1예 보고[1]

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견갑골하 탄력섬유종은 서서히 자라고, 경계가 좋지 않은 드문 양성종양으로, 견갑골 하부와 흉벽 사이에서 전형적으로 발견된다. 초음파, 자기공명영상 및 컴퓨터단층촬영에서 특징적인 소견들이 탄력섬유종의 정확한 진단을 가능하게 하고, 불필요한 수술적 치료를 방지할 수 있다. 저자들은 87세 된 남자환자의 양측성 탄력섬유종을 경험하였기에 문헌고찰과 함께 보고 한다.