Comparative study on imaging and pathological features of elastofibroma dorsi

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Abstract Background and Objective: Elastofibroma dorsi has an extremely low incidence. At present, comparative study on imaging manifestations and pathologic findings of elastofibroma dorsi has not been reported in China. This study was to investigate clinical manifestations, computed tomography (CT) and magnetic resonance imaging (MRI) appearances, and pathologic features of elastofibroma dorsi and to improve preoperative imaging diagnosis of this disease. Methods: The clinical manifestations, imaging findings, and pathologic appearances of 6 cases of elastofibroma dorsi were retrospectively analyzed and related literatures were reviewed. All patients were examined with MRI, and 4 of them were examined with CT scan. Results: All patients were above 30 years old without obvious symptoms. The tumors presented as a lenticular soft-tissue mass in the deep subscapular region. The tumor’s density on plain CT scan or signal intensity on MR T₁-weighted image was approximately equal to that of muscle with some interfaced fat-like areas within mass suppressed by fat-suppression MR sequences, which corresponded to dense collagen tissue and interspersed mature adipose tissue observed microscopically. Conclusions: CT and MRI can reflect the histological features of elastofibroma dorsi. On the basis of their imaging characteristics, a correct preoperative diagnosis of elastofibroma dorsi can easily be made.

Key words: Elastofibroma, tomography, X-ray computed, magnetic resonance imaging, pathology

Elastofibroma dorsi is a rare soft tissue benign tumor, originated from mesenchymal tissue. Histologically, this tumor is composed of dense collagen fibers with some elastic fibers and mature adipose cells. Surgical excision of tumor has good therapeutic outcome, with an extremely low recurrence rate after marginal excision¹⁻². Previous reports of this tumor in China were focused on clinical and pathologic analysis, and only few studies were about imaging description³⁻⁵. Until to now, only 1 case of elastofibroma dorsi underwent magnetic resonance imaging (MRI) was reported in China⁶. We aimed to investigate clinical manifestations, CT and MRI appearances, and pathologic features of 6 patients with pathologically confirmed elastofibroma dorsi, to enhance our understanding of computed tomography (CT) and MRI features of this disease, and to improve preoperative imaging diagnosis.

Materials and Methods

Clinical data

Clinical, imaging and pathologic records of 6 patients with pathologically confirmed elastofibroma dorsi, treated in the First Affiliated Hospital of Sun Yat-sen University between January 2004 and May 2009, were analyzed. Tumor location: left side, 2 cases; right side, 3 cases; and both sides, 1 case. The patients, including 3 men and 3 women, were between 32 and 65 years old, with a median age of 45 years old. Disease courses were from 3 months to 2 years. The tumors grew slowly and caused no obvious clinical symptoms. Physical examination showed that the tumors located at the inferior angle of scapula were hard with no tenderness; 1 patient had movable mass while 5 had unmovable mass, with no fluctuation or pulsation; local skin temperature, superficial veins, and upper extremity...
function were normal. Marginal excision was performed in all 6 patients. After surgery, clinical and ultrasonic follow-up were performed every 6 months or one year for 7 months to 5 years.

Relevant original literatures about elastofibroma dorsi published in between 1979 and 2009 were searched through China National Knowledge Infrastructure (CNKI) using keyword: elastofibroma. The age, sex, tumor location, disease course, and other clinical features of Chinese patients were analyzed and summarized.

Methods

All 6 cases were examined with plain and enhanced MRI, and 4 of them were also examined with plain and enhanced CT. Single- or 64-slice spiral CT was used in the examination. After plain CT scanning, contrast agent iopromide (300 mg/mL) was intravenously administered. The CT images were performed with a slice thickness of 5 mm.

The Siemens Magnetom Trio 1.5T or 3.0T MRI system was used in chest cross section scanning for all patients. Additional coronal section scanning was performed on 4 patients. Spin-echo (SE) $T_1$ weighted imaging ($T_1$WI), fast SE $T_2$ weighted imaging ($T_2$WI), and fat suppression $T_2$WI with short time inversion recovery (STIR) sequences as well as SE enhanced $T_1$WI scan were performed on all patients. Additional SE fat suppression $T_1$WI enhanced scan was performed on 3 patients. Contrast agent gadolinium-diethylenetriamine pentaacetic acid (Gd-DTPA) was used at a dose of 0.1 mmol/kg. Image parameters were TR 400–600 ms and TE 9–40 ms for $T_1$WI, TR 3000–4000 ms and TE 80–100 ms for $T_2$WI; slice thickness, 4 mm; slice space, 0.3 mm; field of view (FOV), 22–36 cm; matrix, 256 x 256; number of excitation (NEX), 2.

All patients underwent tumor resection. Tumor samples were fixed, paraffin-embedded, and sectioned at a thickness of 4–6 μm for pathologic examination. Tumor slices were stained with hematoxylin-eosin (HE) and aldehyde-fuchsin for microscopy.

Results

Images of the 6 cases of elastofibroma dorsi showed flat hummocky soft-tissue mass at inferior angle of the scapula, closely attached to the bony thoracic cage, the inner margins of tumor contacted with the ribs, and the outer margins contacted with the serratus anterior muscle, latissimus dorsi muscle and lower part of the scapula. Tumor sizes were about 1.5 cm × 8.5 cm × 5.5 cm to 5.0 cm × 10.0 cm × 12.0 cm. On plain CT images, all tumors had almost equal density as muscle, which was heterogeneous with a few strap-shaped densities signals and slightly heterogeneous enhancement on enhanced CT scanning images (Figures 1 and 2). On $T_1$WI images, compared with muscle, all tumors showed isointense signals with a few strap-shaped high intensity signals; on $T_2$WI images, the tumors showed mixed heterogeneous high intensity signals; and on fat suppression MR scanning

Figure 1 CT images of the elastofibroma (arrow) in the right subscapular area
A, on plain CT scan, the tumor shows a soft-tissue mass with heterogeneous density, and clear margins; B, on enhanced CT scan, the tumor shows slight heterogeneous enhancement.
images, the tumors showed slightly high intensity signals, containing a few strap-shaped low intensity signals corresponded to high intensity signals on T₁WI images. On enhanced MRI scan, slightly or moderately heterogeneous enhancement was observed (Figures 3). However, on fat suppression scan, tumor enhancement was relatively obvious (Figure 4). The margins of tumors were clear in 5 cases. In 2 of them, tumors were surrounded by a few adipose tissues, and adjacent muscle was compressed, but its density and signals were normal; in 1 case, the tumor had ambiguous margin, surrounded the scapula, and invaded into surrounding muscles (Figure 5), but the ribs and scapula were normal.

During operation, the tumors were found to locate at the deep side of the serratus anterior muscle, latissimus dorsi muscle, and rhomboid muscle and adhered to the ribs.
Figure 4 MRI images of the elastofibroma (the same case in Figure 2) in the bilateral subscapular areas (arrows)
A, on T₁WI, the tumors show iso-intensity signals with a few hyper-intensity signals; B, on T₂WI, the tumors show mixed iso- and hyper-intensity signals; C, on STIR T₁WI, the high signals can be suppressed by STIR sequence; D, on enhanced STIR T₁WI, the tumors show obvious enhancement.

Figure 5 MRI images of the elastofibroma in the left subscapular area (arrow)
A, on T₁WI, the tumor shows an irregular soft-tissue mass with unclear margins and signals similar to that of muscle; B, on enhanced T₁WI, the tumor shows moderate enhancement.
and ligamenta intercostalia. Tumors had no capsule, were flat, irregular, and hard. Pathologic examination with HE staining showed lots of collagen fibers stained in red, spindle cells with no heterocaryon or mitosis, a few strap, string beads, and globular shaped elastic fibers with deep staining, and a few interspersed mature adipose tissue, but no inflammatory cells (Figure 6). Elastic fiber stained with aldehyde-fuchsin was positive and appeared black (Figure 7). The 6 patients were followed up for 7 months to 5 years after marginal excision, and no recurrence occurred.

In Chinese literature, 51 original papers about elastofibroma dorsi were found. Among these reports, 45 were about clinical and pathologic manifestations, 6 were about CT presentations; only 1 case with MRI examination was reported, which located at the inferior angle of left scapula. In summary, 378 cases of elastofibroma dorsi were described in clinical details. Of them, 49 patients were men and 329 were women; 265 patients had unilateral lesion and 113 had bilateral lesions; the oldest patient was 82 years old, while only 10 were under 40 years old; disease courses varied from months to years and the longest one was 23 years.

Discussion

Elastofibroma dorsi is very rare and was first reported by Järvi et al.[7] in 1961 with four cases. In the past 5 years,
only 6 cases were treated in our hospital. With accumulative reports and enhanced understanding, it was found that elastofibroma dorsi is a specific kind of elastofibroma that locates on the back, and 93% locate at the inferior angle of scapula. Also, elastofibroma can be found at some frequently moved sites such as the greater trochanter of femur, ischial tuberosity, cardiac valves, and anterior part of thighs. In the 2002 WHO soft tissue tumor classification, elastofibroma is classified as “benign fibroblast/myofibroblast tumor”. However, most scholars do not accept it as a real tumor, but as hyperplasia, transformation and tumor-like growth of elastic tissues, which are derived from fibroblasts under chronic trauma or frequent mechanical friction.

Elastofibroma dorsi has some specific clinical features. It is mostly seen in the elderly people, with an incidence higher in women than in men. Most patients have unilateral lesion, and only a few have bilateral lesions. Patients usually have a long disease course without subjective symptom. Most patients visit hospital for growing hard mass. In 378 cases of the elastofibroma dorsi reported in Chinese literatures, 97.4% were older than 40 years, male to female ratio was 1:6.7, unilateral to bilateral ratio was 2.3:1, and the longest disease course was over 20 years.

In pathology, wavy, string beads, serrated and globular shaped elastic fibers and interspersed mature adipose cells were the diagnostic evidences for this disease, and elastic fiber staining was positive. The prognosis after surgical excision is good with an extremely low recurrence rate. No recurrence was found in our patients at 7 months to 5 years after marginal excision.

CT and MRI can clearly show the location, size, composition, and structural relation with adjacent tissues of elastofibroma dorsi. In our study, images showed that the tumors located at the deep side of the serratus anterior muscle, latissimus dorsi muscle, rhomboid muscle, and subscapularis and adhered to the thorax. These soft-tissue masses were irregular and flat, with clear margins in most cases. The vast amount of collagen fibers in the tumor determined its CT and MRI presentations, with density or signals similar to those of muscle density or signal; and interspersed adipose tissue in the tumor determined its specific CT and MRI presentations, with a few adipose tissue density or signals. Adjacent scapula and thoracic ribs were normal, surrounding soft-tissue seructure was often compressed and some tumors had a few surrounding adipose tissues, indicating benign behaviors of this disease. However, invasive growth was found in one of our patients. Tumors showed slight or moderate enhancement on CT or MRI images, suggesting blood supply was not abundant. This was consistent with the results of color Doppler flow imaging reported by Jiang et al. Clinically, elastofibroma dorsi can be distinguished from adipoma, fibroma, and angiomia by CT or MRI, but sometimes specific elastic fiber staining of samples from puncture biopsy is needed for differential diagnosis.

All together, elastofibroma dorsi usually occurs in senile people, and has specific location, imaging features, and tissue features. According to its imaging characteristics of CT or MRI, it is not difficult to make correct diagnosis of elastofibroma dorsi before surgery.

References