The study aimed to retrospectively review sonographic features of extra-articular giant cell tumor of the tendon sheath (GCTTS), and developed a grouping system for facilitating differential diagnosis. From January 2005 to December 2009, 15 pathologically proven extra-articular GCTTS in 15 patients were encountered in our hospital. According to the tumor sites and their sonographic features, we categorized the sonographic findings into three types: superficial type (tumor attaching to the tendon but no complete encasement of it), encasing type (tumor completely encasing the tendon) and juxta-fascial type (tumor without attachment to the tendon). The demographic data, clinical presentation symptom, sonographic feature and color or power Doppler flow in each type of the patients were documented. Histopathologically, the localized or diffuse form of GCTTS was also recorded.

The incidences of sonographic presentation in the superficial, encasing and juxta-fascial types of GCTTS were 46.7% (n=7), 33.3% (n=5) and 20.0% (n=3), respectively. The masses in the encasing type manifested with largest average size. The most common location of GCTTS in the superficial and encasing types was the hand. The 3 juxta-fascial type GCTTS were located in the subcutis of the hand and buttock, and the subfascial region of the forearm. On sonography, all GCTTS presented as hypoechoic masses with homogeneous or heterogeneous echogenicity. The tumors in the superficial and encasing types were eccentrically located to the related tendon and their superficial components were always disproportionately predominant. Bony erosion was found in three masses. No dermal attachment, decreased or increased sound through transmission, calcified or cystic component were noted in all masses. Only 26.7% of GCTTS demonstrated hypervascularity within the tumors. The two largest tumors were in encasing type and reported to be diffuse form microscopically. Two patients underwent recurrence, one with mass in superficial type and another in encasing type.

We concluded that extra-articular GCTTS typically appears as a hypoechoic mass with heterogeneous or homogeneous echogenicity and intimate contact with the abutting tendon or fascia. The diffuse form GCTTS should be considered if a characteristic mass presented with larger size, lobulated or irregular contour, complete encasement of the related tendon and hypervascularity. Besides, differential diagnosis of a well-defined and fascia-attached mass should include juxta-fascial type GCTTS.

Giant cell tumor of the tendon sheath (GCTTS) and pigmented villonodular synovitis (PVNS) represent the family of proliferative synovial disorders that may affect the joint, bursa, and tendon sheath (1-3). Extra-articular GCTTS, also referred to as nodular tenosynovitis or pigmented villonodular tenosynovitis (PVNTS), is a slowly-growing benign soft-tissue tumor and accounts for approximately 1.6
Giant cell tumor of tendon sheath: sonographic features

It occurs most frequently in third to fifth decades and presents with a slight female preponderance [1-4]. The most common location of GCTTS is the hand, followed by the foot and ankle [1, 3-7]. Clinically, the patients usually manifest with palpable soft-tissue mass or pain [1-2]. The recurrence rate after surgical resection is about 7 to 27% for localized form and 40 to 50% for diffuse form [1-2, 6]. Multifocal presentation of GCTTS is occasionally encountered and malignant GCTTS is extremely rare [1-3, 8].

On radiographs, 50 to 70% of GCTTS typically appear as a soft-tissue mass [1]. Other radiographic findings, such as bony erosion, calcification, periostal reaction, and intraosseous invasion are infrequently encountered [1-3, 9-10]. As a consequence of hemosiderin deposition and variable amount of collagenous contents in most GCTTS, the tumor is characterized by a mass with low signal on T1- and T2-weighted MR imaging, and shows “blooming” artifact on gradient echo sequences [1, 11-12]. Nonetheless, sonographic features of extra-articular GCTTS were rarely reported in the English literature [8, 13-15]. The objective of the study was to retrospectively review sonographic findings of extra-articular GCTTS, and developed a grouping system for facilitating differential diagnosis.

MATERIALS AND METHODS

The Institutional Review Board of our hospital approved this study. From January 2005 to December 2009, 15 pathologically proven extra-articular GCTTS in 15 patients were encountered in our hospital. The mean age of the patients was 43.4 years with a range of 12 to 66 years (Table 1). They were 7 male patients and 8 female patients. All patients presented with palpable masses and only two of them complained of painful sensation. The locations of the lesions were as follows: hand (n=8), wrist (n=1), foot (n=3), ankle (n=1), forearm (n=1) and buttoc (n=1). Eight masses were located in the flexor sides and seven in the extensor sides. The tumor sizes ranged from 1.0 to 4.1 cm with a mean of 2.2 cm. The mean follow-up period was 24.6 months and recurrent rate was 13.3%. All sonogram were performed by using a Sequoia 512 scanner (Acuson, Mountain View, CA) with a linear 15L8W-S transducer, setting at 14 MHz. All patients also underwent color or power Doppler sonographic examinations. Two radiologists jointly reviewed all the sonographic images and all interpretations were confirmed by consensus of the two radiologists.

Based on sonographic features and tumor locations, we categorized the sonographic findings into three types: superficial type (tumor attaching to the tendon but no complete encasement of it) (Fig. 1), encasing type (tumor completely encasing the tendon) (Fig. 2) and juxta-fascial type (tumor without attachment to the tendon) (Fig. 3). The demographic data, clinical presentation symptom and tumor site were recorded in each type of the patients. The sonographic features, including tumor size (maximal diameters at any plane), echogenicity (with reference

<table>
<thead>
<tr>
<th>Type</th>
<th>Superficial</th>
<th>Encasing</th>
<th>Juxta-fascial</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case number (%)</td>
<td>7 (46.7%)</td>
<td>5 (33.3%)</td>
<td>3 (20.0%)</td>
<td>15 (100%)</td>
</tr>
<tr>
<td>Age: mean ± SD years</td>
<td>49.6 ± 17.4</td>
<td>39.4 ± 18.5</td>
<td>35.7 ± 20.6</td>
<td>43.4 ± 18.1</td>
</tr>
<tr>
<td>Female/Male</td>
<td>3/4</td>
<td>3/2</td>
<td>2/1</td>
<td>8/7</td>
</tr>
<tr>
<td>Symptoms: Mass/Pain</td>
<td>7/1</td>
<td>5/1</td>
<td>3/0</td>
<td>15/2</td>
</tr>
<tr>
<td>Location</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hand:5</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Foot:2</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wrist:1</td>
<td>Hand:2</td>
<td></td>
<td></td>
<td>Hand:8</td>
</tr>
<tr>
<td>Forearm:1</td>
<td>Foot:1</td>
<td></td>
<td></td>
<td>Wrist:1</td>
</tr>
<tr>
<td>Buttock:1</td>
<td>Ankle:1</td>
<td></td>
<td></td>
<td>Foot:3</td>
</tr>
<tr>
<td>Forearm:1</td>
<td>Buttock:1</td>
<td></td>
<td></td>
<td>Ankle:1</td>
</tr>
<tr>
<td>Flexor/extensor side</td>
<td>4/3</td>
<td>3/2</td>
<td>1/2</td>
<td>8/7</td>
</tr>
<tr>
<td>Size: mean ± SD cm</td>
<td>2.0 ± 0.5</td>
<td>2.7 ± 1.0</td>
<td>1.8 ± 0.9</td>
<td>2.2 ± 0.8</td>
</tr>
</tbody>
</table>
Giant cell tumor of tendon sheath: sonographic features

To adjacent predominant tissue, margin (well-defined or ill-defined), contour (oval, lobulated or irregular), bony erosion (concave cortical defects in the surface of the abutting bone), dermal attachment, increased or decreased sound through transmission, calcification (hyperechoic areas with shadowing), and cystic change (anechoic regions within the lesion), were documented. On the basis of the color or power Doppler images, the vascularities of tumors were graded as hypovascularity, equal-to-vascularity and hypervascularity (with reference to Doppler flow of the surrounding tissue). Tumors were considered to be hypovascular or hypervascular if they were less or more Doppler flow than adjacent tissues, respectively. Histopathologically, the localized or diffuse form was recorded. The recurrence after tumor excision was also documented.

RESULTS

The incidences of sonographic presentation in the superficial, encasing and juxta-fascial types of GCTTS were 46.7% (n=7), 33.3% (n=5) and 20.0% (n=3), respectively (Table 1). The average patient’s age was oldest in the superficial type (49.6±17.4 years) and youngest in the juxta-fascial type (35.7±20.6 years). Slight female predominance was noted in the encasing (female/male=3/2) and the juxta-fascial (female/male=2/1) types. Two patients complained of painful sensation, one in the superficial type and another in the encasing type. The most common location of GCTTS in the superficial and encasing types was the hand. The three GCTTS in the juxta-fascial type were located in the subcutis of the hand and buttock, and the subfascial region.

Figure 1. A superficial type of giant cell tumor of the tendon sheath in the flexor side of the 1st toe of a 12-year-old boy. Sonographic study along the longitudinal axis of the toe reveals a heterogeneously hypoechoic mass (arrows) attaching to the underlying flexor tendon (arrowheads). The deeper aspect of tendon is not encased by the tumor.

Figure 2. An encasing type of giant cell tumor of the tendon sheath in the ankle of a 17-year-old man. Transverse scan of sonographic study exhibits a lobulated well-defined and heterogeneously hypodense mass (arrows) which totally encases the posterior tibialis tendon (arrowheads). The tumor is not contact with the dermis (open arrows).

Figure 3. A juxta-fascial type of giant cell tumor of the tendon sheath in the extensor side of the forearm of a 50-year-old woman. On transverse scan of sonography, an oval well-defined and heterogeneously hypoechoic mass (arrows) is located in between the subcutis, extensor carpi ulnaris muscle (right star) and ulna (left star). During surgical intervention, the mass is found to adhere to the fascia of the muscle, reflecting probable origin of the tumor from the fascia.
of the forearm. The three tumors were found to attach to the fasciae of the palm, gluteal maximus, and extensor carpi ulnaris muscles on sonography respectively and confirmed during surgical intervention. The tumors in the superficial and encasing types were located slightly more common in the flexor sides (n=4 and 3 respectively) than in the extensor sides (n=3 and 2 respectively). The average size of the tumors in the encasing type was largest (2.7 cm).

On sonography, all GCTTS presented as hypoechoic masses. Slight preponderance of homogeneous echogenicity was only noted in the masses of the superficial type. The masses in the superficial type were eccentrically located to the tendon and always in the superficial aspect (Fig. 1). The masses in the encasing type were also eccentric location to the related tendon and their superficial proportions were always disproportionally larger than deep proportions (Fig. 2). Sonographic appearance of two masses manifested as partially ill-defined border. Oval contour was noted in all masses of the superficial type. Two masses in the encasing type and one mass in the juxta-fascial type appeared as lobulated or irregular contour. There were one mass in the superficial type and two masses in the encasing type presenting with bony erosion (Fig. 4a). No dermal attachment, decreased or increased sound through transmission, calcified or cystic component were noted in all masses. Color or power Doppler sonographic study revealed 53.3% of masses with vascularity equal to the abutting soft tissue. Only 26.7% of masses demonstrated hypervascularity within the tumors (Fig. 4b). The two largest masses were confirmed to be diffuse form of GCTTS and both of them belonged to the encasing type (Fig. 2, 4a, 4b). They were located in the foot and ankle, and aged 32 and 17 years respectively. Color or power Doppler study also exhibited hypervascularity in both of them. Two patients underwent recurrence, one with mass in the superficial type and another in the encasing type.

**DISCUSSION**

Histopathologically, GCTTS contains variable amounts of synovial cells, inflammatory cells, multinucleated giant cells, histiocytes, macrophages, and xanthoma cells [2]. The presence of collagenous stroma and hemosiderin deposition in GCTTS are also characteristic microscopic findings. Jaffe et al firstly proposed a unified classification, including PVNS, pigmented villonodular bursitis (PVNB), and PVNTS, to represent the proliferative diseases affecting the synovial tissues such as joint, bursa and tendon sheath [16]. Subsequently, PVNS is now commonly used to represent the diffuse intra-articular form, whereas the localized extra-articular, localized intra-articular, and diffuse extra-articular forms are included in GCTTS [2, 17]. In the classification of bone and soft-tissue tumors of World Health Organization system, all these disease entities are now categorized as “fibrocystic tumor” [18].

Recently, the evidences of autonomous growing
ability and cytogenetic studies of GCTTS and PVNS strongly supported that the histogenesis of these masses is a neoplastic process [1,19]. Besides, the implication of GCTTS and fibroma of the tendon sheath remained debated [20-21]. Some authors addressed that consequent or progressive vascular impairment may lead to sclerosing of GCTTS and final formation of fibroma of the tendon sheath [20-21]. In our study, one GCTTS revealed regressing change microscopically and no Doppler flow was found in it. We proposed the different evolution stages of GCTTS might be a major contributory factor which resulted in diverse sonographic echogenicity and variable vascularity in the tumors.

In the study, we demonstrated the superficial type was the most common sonographic presentation. Given that the tumor were always located in the superficial aspect of the related tendon, we name the sonographic pattern as “superficial” type. The differential diagnosis of superficial type GCTTS may include ruptured ganglion cyst, fibroma of the tendon sheath, calcifying aponeurotic fibroma, fibromatosis, epidermal cyst and sarcoma. The absence of fluid in the ruptured ganglion cyst can help to exclude GCTTS. To our knowledge, sonographic findings of fibroma of the tendon sheath have not been elucidated in the literature. However, apparent low signal in hemosiderin-containing GCTTS on long TR or gradient echo MR imaging is a favorable sign to differentiate GCTTS from fibroma [22]. Unlike GCTTS, ill-defined margin and calcification are frequently encountered in calcifying aponeurotic fibroma [23-24]. Fibromatosis is usually infiltrative, not obviously contact with the tendon and occurs in the deeper location [8, 24]. On sonography, epidermal cyst is characterized by a mass with dermal attachment, increased through transmission, hyperechoic reflector and hypoechoic linear cleft, which are unseen within GCTTS [25, 26]. In addition to rapid growing history and ill-defined border frequently noted in sarcoma, the related tendon, which always remains intact in GCTTS, may be invaded by sarcoma [13].

The masses in the encasing type occurred second most common in the study. The two largest GCTTS were in encasing type and their histopathological appearances were diffuse form. Diffuse form of GCTTS occurs less commonly than localized form and accounts for only 12% of total GCTTS [4, 6, 27]. It has been addressed that diffuse form tends to occur in the younger patient and presents with larger and multinodular or irregular mass [4, 6, 27]. Given aggressive behavior and higher recurrent rate of diffuse form GCTTS, meticulous sonographic identification of the characteristics of diffuse form GCTTS is imperative. On sonography, the encasing type GCTTS should be distinguished from tenosynovitis and sarcoma. The presence of fluid in most tenosynovitis is a helpful sign to distinguish the disorder from GCTTS [28]. Nonetheless, chronic or granulomatous tenosynovitis may appear as a tumor-like lesion with thickened synovium and absence of fluid [28-29]. In this situation, disproportionally larger superficial component in encasing type

| Table 2. Summary of sonographic features, diffuse or localized histopathological form and recurrence of 15 pathologically proven giant cell tumors of the tendon sheath. |
|---|---|---|---|---|
| **Type** | **Superficial** | **Encasing** | **Juxta-fascial** | **Total** |
| Sonographic features | | | | |
| Homo-/Heterogeneously hypoechoic | 4/3 | 2/3 | 0/3 | 6/9 |
| Well-defined/ill-defined margin | 6/1 | 4/1 | 3/0 | 13/2 |
| Oval/Lobulated or irregular contour | 7/0 | 3/2 | 2/1 | 12/3 |
| Bony erosion | 6/1 | 3/2 | 3/0 | 12/3 |
| Dermal attachment | 0 | 0 | 0 | 0 |
| Through transmission | 0 | 0 | 0 | 0 |
| Calcification or cystic changes | 0 | 0 | 0 | 0 |
| Hypo-/equal-to-/hypervascularity | 1/4/2 | 2/1/2 | 0/3/0 | 3/8/4 |
| Localized/Diffuse form | 7/0 | 3/2 | 3/0 | 13/2 |
| Recurrence | 1 | 1 | 0 | 2 |
GCTTS may be helpful. The key differential diagnostic points of sarcoma from encasing type GCTT were still relied on rapid growth history, ill-defined border and potential invasion to the related tendon in sarcoma.

Rarely, extra-articular GCTTS can arise from the subcutaneous or intramuscular regions [5-6, 30]. Somerhausen and Fletcher reported that 6% of GCTTS occurred in the subcutaneous areas [5]. For the three masses of juxta-fascial type GCTTS in our study and another one reported by Sanghvi DA et al, all these tumors were away from the tendons but attached to the abutting fasciae. Hence, we proposed the fasciae were another potential origin for the rarest type of extra-articular GCTTS. Owing to its location and non-specific sonographic findings of juxta-fascial type GCTTS, the differential diagnosis becomes more challenging. Common tumors, such as schwannoma, neurofibroma, lipoma, epidermal cyst, nodular fasciitis, fibromatosis or sarcoma, may be included in the differential diagnosis. Juxta-fascial type GCTTS should be considered if a well-circumscribed hypoechoic mass with close attachment to the fascia and no cystic or calcified component was encountered.

In our study, some limitations might exist. First, the case number was small and no statistic data was analyzed. As in other retrospective study, the selection bias may be present because all the masses in the study were selected from the patients who received the surgical intervention and then confirmed by histopathological examination.

In summary, extra-articular GCTTS typically appeared as a hypoechoic mass with heterogeneous or homogeneous echogenicity and close contact with the abutting tendon or fascia. The diffuse form GCTTS should be considered if a characteristic mass presented with larger size, lobulated or irregular contour, completely encasing the tendon and hypervascularity. Besides, the differential diagnosis of a well-defined and fascia-attached mass should include juxta-fascial type GCTTS.

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Giant cell tumor of tendon sheath: sonographic features

關節外腱鞘巨細胞瘤之超音波表徵

林理涵¹  黃宗正¹  高常發¹  黃玄嬴²  王俊聞³  鄭汝汾¹  李子瑜¹

長庚紀念醫院高雄醫學中心  長庚大學醫學系 放射線診斷科¹  病理科²  骨科³

此研究目的為回溯性評估關節外腱鞘巨細胞瘤之超音波表徵，並嘗試發展一分類方法以利於鑑別診斷。

從民國 95 年 1 月至 98 年 12 月間，我們共收集了 15 個經病理報告證實為關節外腱鞘巨細胞瘤。根據腫瘤位置及其超音波表徵，我們將超音波的表現分為三類：表淺型（腫瘤附著於肌腱但並未將其完全包圍），包圍型（腫瘤完全包圍肌腱），筋膜旁型（腫瘤附著於肌腱）。各型腫瘤的臨床症狀及超音波表徵均為收集項目。

超音波表現為表淺型，包圍型和筋膜旁型發生率各為 46.7%，33.3% 和 20%。包圍型的腫瘤表現出最大的平均尺寸。表淺型及包圍型腫瘤最常發生於手。所有的腱鞘巨細胞瘤均呈現為均質或非均質的低回音性腫塊。表淺型及包圍型的腫塊偏離中心地附著於相關肌腱上，且腫塊的表淺部份總是為格外顯著。兩個最大的腫塊為包圍型且屬於潰漫型腱鞘巨細胞瘤。

此研究的結論為典型的關節外腱鞘巨細胞瘤之超音波表徵為一均質或非均質的低回音性腫塊且與相鄰的肌腱或筋膜緊密的接觸。當一典型的腫塊表現出大尺寸，分葉狀或不規則的輪廓，完全包圍相關的肌腱和高血管性時，應考慮潰漫型腱鞘巨細胞瘤。此外，一個界限明確且附著於筋膜之腫塊的鑑別診斷應包含筋膜旁型腱鞘巨細胞瘤。