

# Ultrasonographic imaging of calcifying fibrous tumor of cervical esophagus

## A case report

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### Abstract

**Rationale:** Calcifying fibrous tumor (CFT) is a rare benign mesenchymal tumor characterized by cytologically benign fibroblasts that produce abundant collagen with scattered lymphocytes, plasma cells, and psammomatous or dystrophic calcifications.

**Patient concerns:** A 37 years old women was initially diagnosed thyroid nodule in local hospital. She was referred to receive microwave ablation of the nodule in our hospital. The patient was in good health with occasional foreign body sensation in the neck. Routine laboratory examination was normal.

**Diagnosis:** The lesion was suspected benign and originating from the esophagus rather than the thyroid gland by conventional ultrasound and contrast-enhanced ultrasound in our hospital and was finally proved to be CFT by pathology.

**Interventions:** The lesion was surgically removed.

**Outcomes:** The patient recovered well and was followed up for 2 years without recurrence.

**Lessons:** Although imaging diagnosis of CFT is difficult, the findings of clear border, coarse calcification on conventional ultrasound and peripheral hypoenhancement without central enhancement on contrast-enhanced ultrasound (CEUS), together with real-time evaluation of the relationship between the tumor and esophagus help to distinguish CFT of cervical esophagus from other lesions in the neck.

**Abbreviations:** CFT = calcifying fibrous tumor, FNA = fine needle aspiration, CEUS = contrast-enhanced ultrasound, CT = computer tomography.

**Keywords:** calcifying fibrous tumor, contrast-enhanced ultrasound, esophagus, ultrasonographic imaging

## 1. Introduction

Calcifying fibrous tumor (CFT) is a rare benign mesenchymal tumor composed of hyalinized fibrous tissue with interspersed bland fibroblastic spindled cells, scattered psammomatous, and/

or dystrophic calcifications and variably prominent mononuclear inflammatory infiltrate.<sup>[1]</sup> CFT is often found incidentally as a gradually enlarging painless mass and shows a predilection for the abdominal cavity and soft tissue. CFT in cervical esophagus is rare. However, it may impose a differential diagnosis of more common tumor of neighboring thyroid gland. Herein we report an incidental finding of a CFT in cervical esophagus which was initially diagnosed as thyroid tumor.

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## 2. Case report

A 37 years old woman was diagnosed mass in the left lobe of thyroid at local hospital for regular check-up by ultrasonography. She was referred to have microwave ablation of the thyroid nodule in our hospital. The patient was in good health with occasional foreign body sensation in the neck. Preoperative routine laboratory examination, thyroid function, thyroid antibody test, and parathyroid hormone (PTH) were normal.

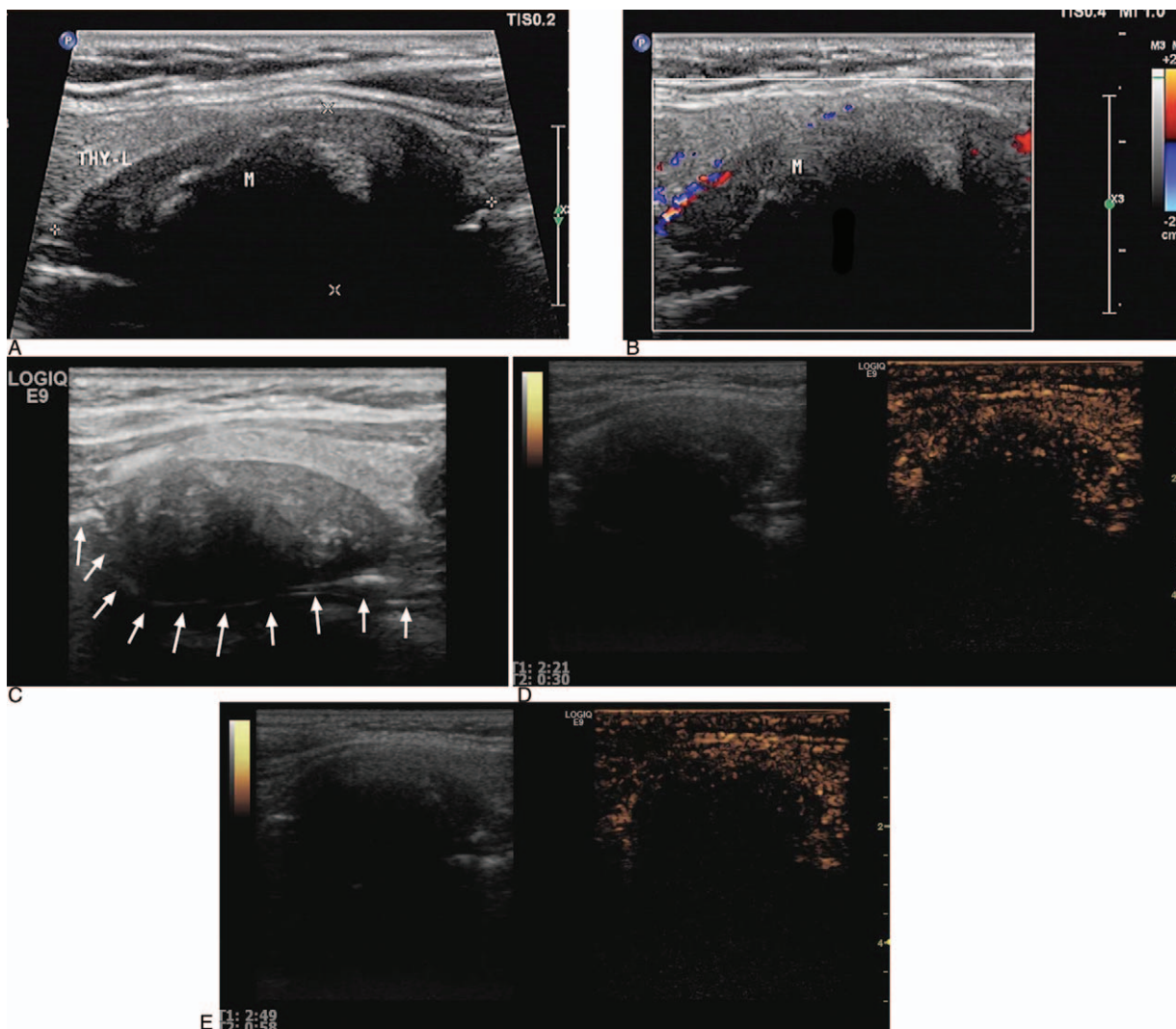
The patient was arranged ultrasound guided thyroid nodule fine needle aspiration (FNA) before microwave ablation of the nodule. However, the lesion was suspected benign and originating from the esophagus rather than the thyroid gland based on the findings of high-frequency conventional ultrasound and contrast-enhanced ultrasound (CEUS) before FNA. Conventional ultrasound was performed with Affinity 50 (Philips Medical System, Netherlands). A heterogeneous nodule measuring 36 × 15 × 28 mm was shown locating posterior inferiorly to the left lobe of the thyroid gland by gray scale ultrasound. The nodule was close

to the lower pole of the thyroid gland and adjacent to the anterior wall of the esophagus. The nodule was hypoechoic in the periphery and hyperechoic in most of the internal area with multiple coarse calcifications. The nodule was ellipse in shape with well-defined margin, accompanied by wide posterior shadows (Fig. 1A). Dotty blood flow signals were demonstrated in the periphery by color Doppler flow imaging (Fig. 1B). To determine the relationship between the nodule and the esophagus, the patient was asked to drink water while scanning. Fluid can be seen flowing through the lesion when swallowing (Fig. 1C) by real-time ultrasound imaging. Further, CEUS was performed by LOGIQ E9 (GE Healthcare) ultrasound system with 4 to 9 MHz linear array probe. The contrast agent used in this study was SonoVue (Bracco, Milan, Italy), a sulfur hexafluoride-filled microbubble. A dose of 2.0 mL SonoVue was injected into the antecubital vein in a bolus fashion through a 20-gauge intravenous cannula, followed by a flush of 10 mL of 0.9% sodium chloride solution. CEUS showed peripheral hypo-

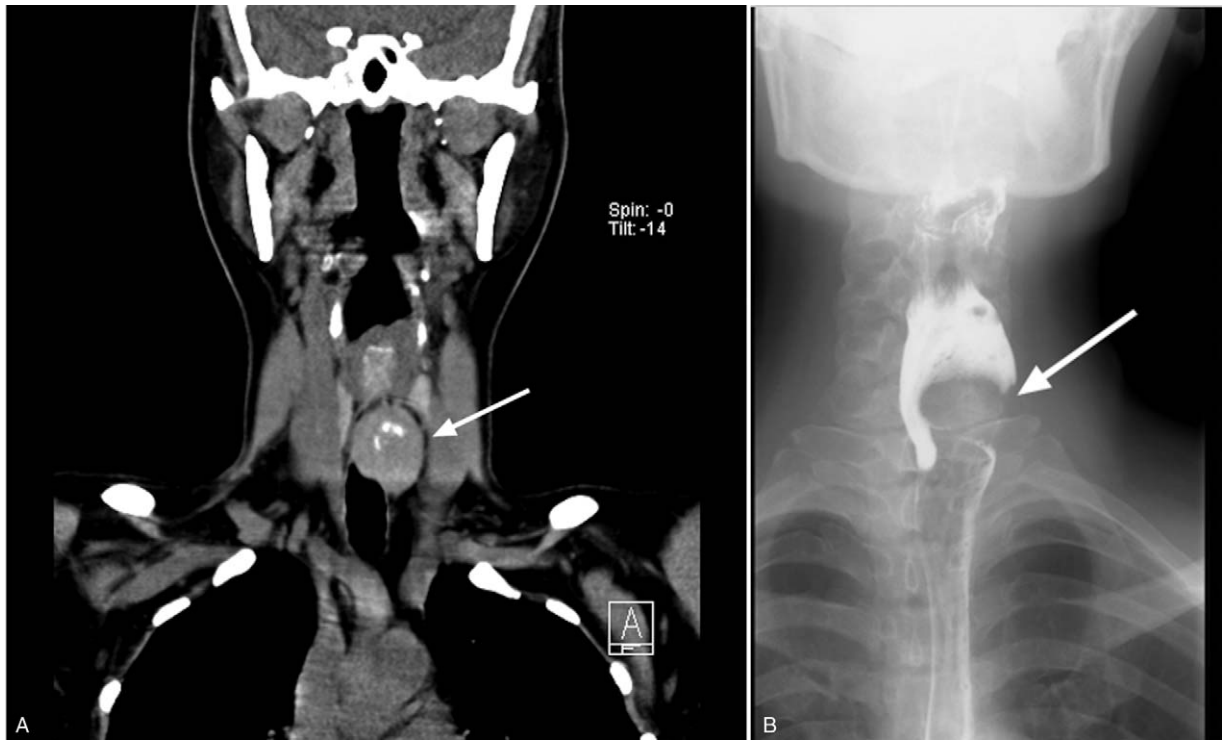
enhancement in arterial phase and no enhancement in the central area (Fig. 1D). The lesion was nearly absent of enhancement in the venous phase (Fig. 1E).

After multidisciplinary panel discussion, microwave ablation was canceled due to ultrasound findings. Esophageal origin of the lesion was further confirmed by subsequent computer tomography (CT) (Fig. 2A) of the neck and gastrointestinal barium meal examination (Fig. 2B). Finally, the lesion was surgically removed. During the surgery, a tumor in the esophageal muscular layer measuring  $30 \times 30 \times 30$  mm with clear border was seen posterior to the lower pole of left thyroid lobe.

A gray solid mass by  $33 \times 30 \times 20$  mm in size, hard in nature with intact capsule was reported by postoperative pathology. Tumor cells were arranged in bundles with a high percentage of dense collagen fibers. Focal calcification and lymphocytes infiltration were seen in the interstitium. Positive for tumor cells vim and smooth muscle actin, negative for Desmin, CD34, CD99, and STAT6 was demonstrated by immunohistochemical staining.



**Figure 1.** Conventional and contrast enhanced ultrasound of a mass in the neck of a 37-year-old woman. (A) On conventional gray scale ultrasonography, a heterogeneous nodule measuring  $36 \times 15 \times 28$  mm with well defined margin was revealed closing to thyroid gland and adjacent to esophagus. The nodule was hypoechoic in the periphery and hyperechoic inside with coarse calcifications and wide posterior shadows. (B) Color Doppler flow imaging demonstrated dotty blood flow signals in the periphery. (C) When the patient was asked to drink water, fluid was observed flowing through the lesion. Air line in the esophagus was marked by arrows. (D) In the arterial phase of contrast enhanced ultrasound, hypoenhancement in the periphery was observed. No enhancement was seen in the center. (E) The lesion was almost non-enhanced in venous phase of contrast enhanced ultrasound.



**Figure 2.** Computed tomography and barium meal findings of the lesion. (A) Computed tomography of the neck demonstrated a soft tissue mass (arrow) measuring 27 × 25 mm posterior to the lower pole of the left thyroid gland with coarse calcifications; (B) A space occupying lesion (arrow) with smooth margin was shown by barium meal of the esophagus.

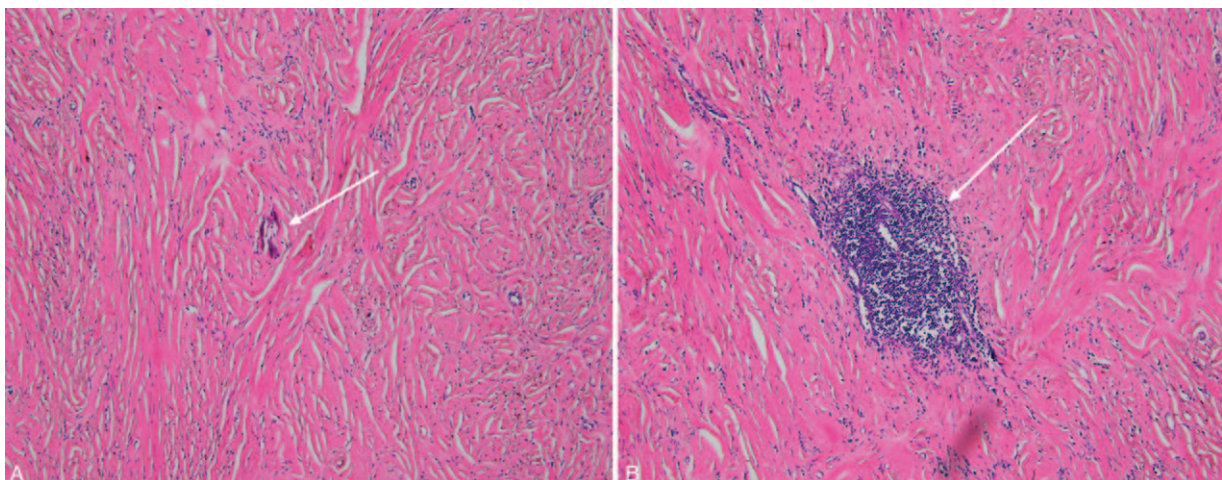
Calcified fibrous tumor was diagnosed according to the morphology and immunohistochemical results (Fig. 3). The patient recovered well after surgery, and no recurrence was found in the ultrasound follow-up for 2 years.

### 3. Discussion

Calcifying fibrous tumor is a rare soft tissue neoplasm originally described by Rosenthal and Abdul-Karim in 1988, while they reported 2 cases of childhood fibrous tumor with psammoma

bodies.<sup>[2]</sup> Subsequently, Fetsch et al introduced the term CFT following an analysis of 10 additional cases, renaming it as calcifying fibrous pseudotumor and pointing out the probable reactive nature of the lesion.<sup>[3]</sup> Nowadays, CFT is considered to represent a true neoplasm, because of the tendency of local recurrence which is occasionally reported. It is now named calcifying fibrous tumor according to recent World Health Organization (WHO) classification.<sup>[4]</sup>

CFT is an uncommon lesion characterized by cytologically benign fibroblasts that produce abundant collagen with scattered



**Figure 3.** Microscopic finding of the lesion. (A) The lesion was composed of dense, hyalinized collagenous tissue and focal calcification (arrow) (H&E stain ×10). (B) Numerous lymphocytes (arrow) infiltration was identified in the lesion (H&E stain ×20). H&E=hematoxylin and eosin.

lymphocytes, plasma cells, and psammomatous or dystrophic calcifications.<sup>[2–4]</sup> CFT occurs in many locations including the chest wall, pleura, mediastinum, adrenal gland, stomach wall, mesentery, testis, retroperitoneal, axilla, and extremities.<sup>[5–10]</sup> Imaging plays an important role in the diagnosis of CFT. However, ultrasound findings of esophageal cases are rarely reported. A previously reported case was imaged by endoscopic ultrasound (EUS). It was shown as an isoechoic tumor over the deep mucosa and submucosal layers, with hyperechoic components inside.<sup>[11]</sup> Our case shared similar gray scale ultrasound features with the previously reported case. Moreover, we further demonstrated the lesion being lack of blood flow by color Doppler flow imaging (CDFI) and CEUS due to predominance of fibrous component on pathology. Delayed enhancement on contrast-enhanced computed tomography (CECT) and contrast-enhanced magnetic resonance imaging (CEMRI) was the major enhancing feature due to the high percentage of fibrous component of CFT.<sup>[12,13]</sup> Comparatively, SonoVue is a pure blood pool contrast agent, which will not leak into the interstitium. Therefore, only low intensity of enhancement was illustrated in arterial phase, without delayed enhancement on CEUS.

Imaging diagnosis of CFT is challenging. In this case, the lesion was initially suspected thyroid nodule with calcification which is much more common for soft tissue mass in the neck. Other lesions, including diverticulum of the esophagus and parathyroid gland tumor should be put into differential diagnosis. When esophagus diverticulum was filled with dietary residue, it would be demonstrated as a hyperechogenic nodule protruding anterolaterally into the thyroid gland, simulating a thyroid nodule. Movement of contents with swallowing, changing in sonographic appearance on repeat exam, hardly palpable and the hyperechogenicity due to the nature of “air or dietary residue” instead of calcification contribute to the differential diagnosis.<sup>[14]</sup> Parathyroid gland tumor, especially parathyroid carcinoma may have undemarcated border and its relationship with the esophagus maybe further revealed when the patients were asked to drink water. Besides, blood calcium and phosphorus level, parathyroid hormone level and serum level of 25-hydroxyvitamin D are useful for the differential diagnosis.<sup>[15]</sup>

Nowadays, the pathogenetic mechanism of CFT is not completely clear. Some researchers have reported CFT was associated with immunoglobulin G4-related disease (IgG4-related disease) and suggest corticosteroid treatment, but radical tumor removal by surgery is the major treatment method, because postoperative histopathology and immunohistochemistry can obtain a definitive diagnosis.<sup>[16,17]</sup>

#### 4. Conclusion

The finding of clear border, coarse calcification on conventional ultrasonography and peripheral hypoenhancement with absence of enhancement in the center on CEUS, together with real-time evaluation of the relationship between the tumor and

esophagus when drinking water help to distinguish CFT of cervical esophagus from other lesions in the neck.

#### Author contributions

**Data curation:** Yan Liu, Xiao-li Wu.

**Funding acquisition:** Qiang Lu.

**Investigation:** Yan Liu.

**Methodology:** Qiang Lu.

**Resources:** Qiang Lu, Guo-ju Shen.

**Software:** Yan Liu.

**Supervision:** Qiang Lu.

**Visualization:** Tao Luo.

**Writing – original draft:** Yan Liu.

**Writing – review & editing:** Qiang Lu.

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