

Interventional

Interventional radiology in Carney triad

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Received: 21 January 2004 **Accepted:** 2 February 2004

Abstract Interventional therapeutic methods are presented in Carney triad, which is a syndrome defined as the simultaneous presence of gastric leiomyosarcoma, extra-adrenal paraganglioma, and pulmonary chondroma. The paragangliomas in the carotid bifurcation and the mediastinum were successfully treated via transarterial embolization with particles. Three intrapulmonary chondromas were ablated using MRI-guided laser-induced thermotherapy (LITT) after previous devascularization via transvenous pulmonary particle embolization. In summary, interventional techniques could be a therapeutic option in patients suffering from Carney triad.

Keywords Carney triad - LITT - Transarterial embolization

Introduction

The association of gastrointestinal stromal tumor (GIST) [in the past referred to as gastric (epithelioid) leiomyosarcoma], extra-adrenal paraganglioma, and pulmonary chondroma was first reported in 1977. Since then, 79 patients with at least two of the three tumors constituting this syndrome (Carney triad) have been identified. Seventeen of the 79 patients (22%) had all three tumors. The remainder had two of the three tumors (partial expression of the triad), usually the gastric and pulmonary tumors.

Characteristic clinical features of this rare disorder are: occurrence at a young age (7–48 years, mean 20.2 years), high incidence (85%) in females, multicentricity of the tumors, and absence of paucity of local symptoms caused by the tumors, particularly the gastric sarcoma.

The repeated recurrences of one or more of the three tumors comprising the syndrome have resulted in a significant morbidity of some patients. To improve their long-term outcome and decrease postsurgical morbidity, the development of nonsurgical methods of therapy that could permit

repeated local treatment and follow-up of the tumors would be of great value.

Herein, we present the case of a patient with the complete triad whose paragangliomas and recurrent pulmonary chondromas were treated by transarterial chemoembolization and magnetic resonance imaging (MRI)-guided laser-induced interstitial thermotherapy (LITT).

Case report

The patient, a 33-year-old woman, presented at our institute in March 2003, 15 years after the diagnosis of Carney triad had been made. In 1987, at the age of 17 years, she had attended another hospital complaining of anemia, fatigue, and intermittent gastric pain. Gastroscopy revealed that the anemia was caused by bleeding from several polypoid, partially ulcerated gastric lesions. At laparotomy, performed in 1988, a submucosal, partially ulcerated mass consisting of several tumors located in the antrum and the lesser curvature was found. Subtotal gastrectomy was performed. Histologically, the tumors were classified as epithelioid leiomyosarcomas.

Three months later, a chest radiograph revealed a single nodule in the right upper lobe of the lung. It was suspected that the tumor was a metastasis from the gastric sarcoma. Four cycles of a polychemotherapy were administered but there was no decrease in the tumor size. Due to uncertainty regarding the nature of the lesion, thoracotomy was performed and the lesion was removed by wedge resection (segment 3). The tumor was histologically classified as a chondroma.

From 1991 to 1995, five more nodules appeared in the left lung. In 1995, following episodes of hemoptysis, a third thoracotomy was performed with resections of segments 1, 2, 3, and a wedge resection of segment 6 and the lingual of the left lung. All five tumors were partially calcified chondromas. Over the next 7 years, serial CT revealed two and three more tumors in the right and left lungs, respectively.

In 2002, fluorine 18 Fluoro-2-deoxyglucose positron emission tomography (F^{18} FDG-PET) revealed a large mass located in the middle mediastinum. CT and further localization tests [131 I-metaiodobenzylguanidine (MIBG) scan and 111 In octreotide scintigraphy] confirmed these findings, showing a highly vascular tumor in the mediastinum and a mass in the right and left carotid body. The findings were consistent with nonfunctioning extra-adrenal paragangliomas.

When examined at our institution, the patient had six pulmonary lesions, four in the left lung and two in the right lung, ranging from 0.4 to 3.8 cm in diameter, respectively, and an aortopulmonary mass of 2.9 cm in diameter (Fig. 1a).

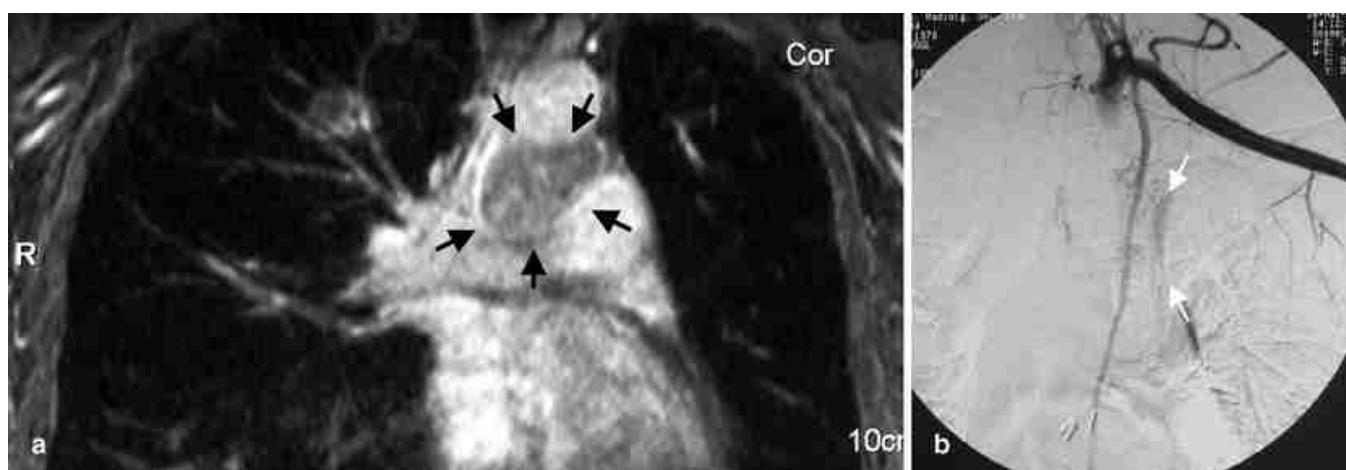


Fig. 1 **a** Coronal contrast-enhanced T2-weighted image (TR/TE=4.5/2.1) displays a mediastinal paraganglioma (arrows) with a maximum diameter of 3.8 cm. **b** Angiography before embolization. Note the

Transarterial embolization (TE) was performed in order to achieve growth cessation of the mediastinal paraganglioma (Fig. 1b) and the carotid gangliomas (Fig. 2). Via a 5-French arterial inguinal access, a pigtail catheter was introduced, and an angiographic survey revealed the hypervascular nature of the lesions. Via selective catheterization the tumor-supplying arteries were superselectively approached using a headhunter 5-F catheter and a turbo tracker system (Boston, Frankfurt, Germany). Using contour particles, the supplying arteries from the internal mammary artery and the cervicothoracic branches were catheterized and embolized (250–350 μm) in two sessions (Fig. 1b). In a third session the directly feeding arteries of the carotid paragangliomas were embolized using the identical protocol. No side effects or complications from the procedures were noted. Three-month interval follow-up studies revealed complete growth cessation of both lesions.

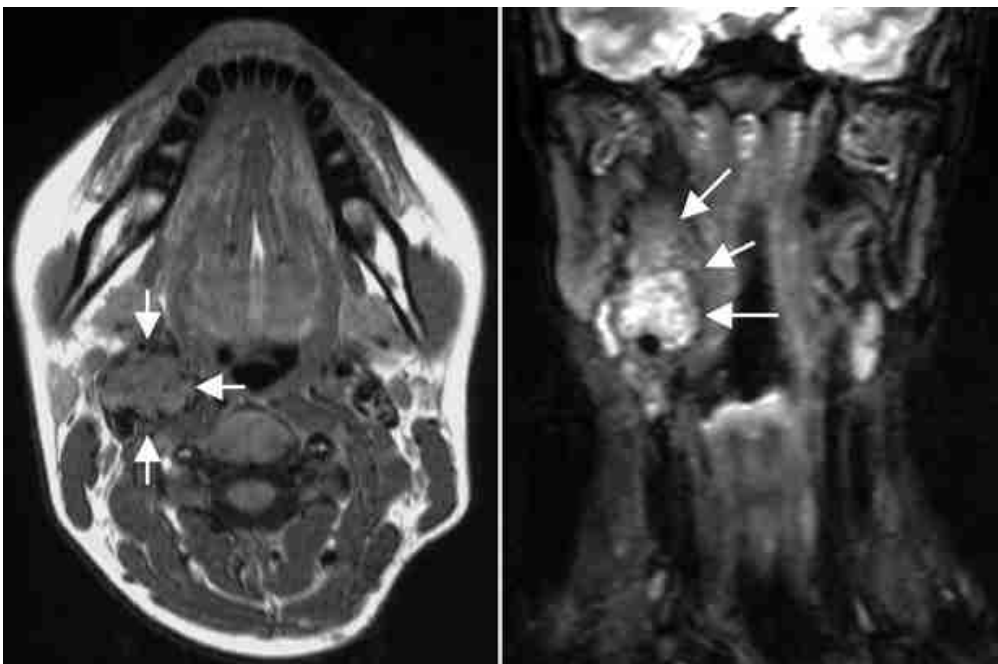
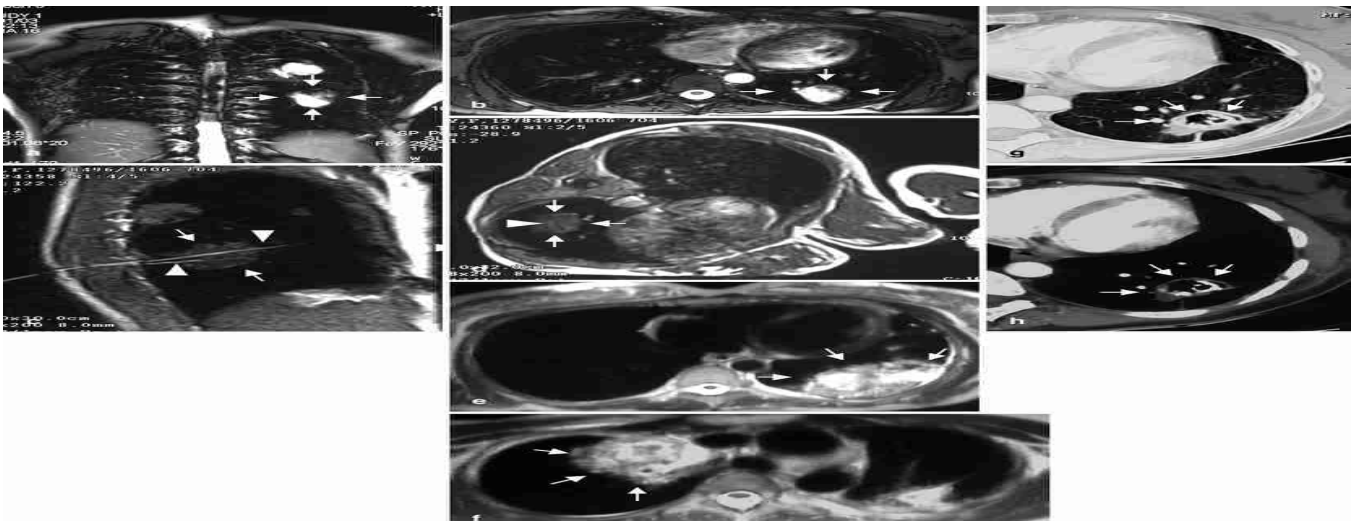


Fig. 2 T1-weighted images (TR/TE=663/15 and 6,290/22) show the carotid paraganglioma in the right paracarotid space (arrows)

The pulmonary chondromas were thermally ablated via MR-guided LITT (Fig. 3a,b). After localization with CT, a specially notified one-step LITT system (Somatex, Berlin, Germany) was introduced under continuous fluoroscopic CT imaging (Care vision, Siemens, Erlangen, Germany).



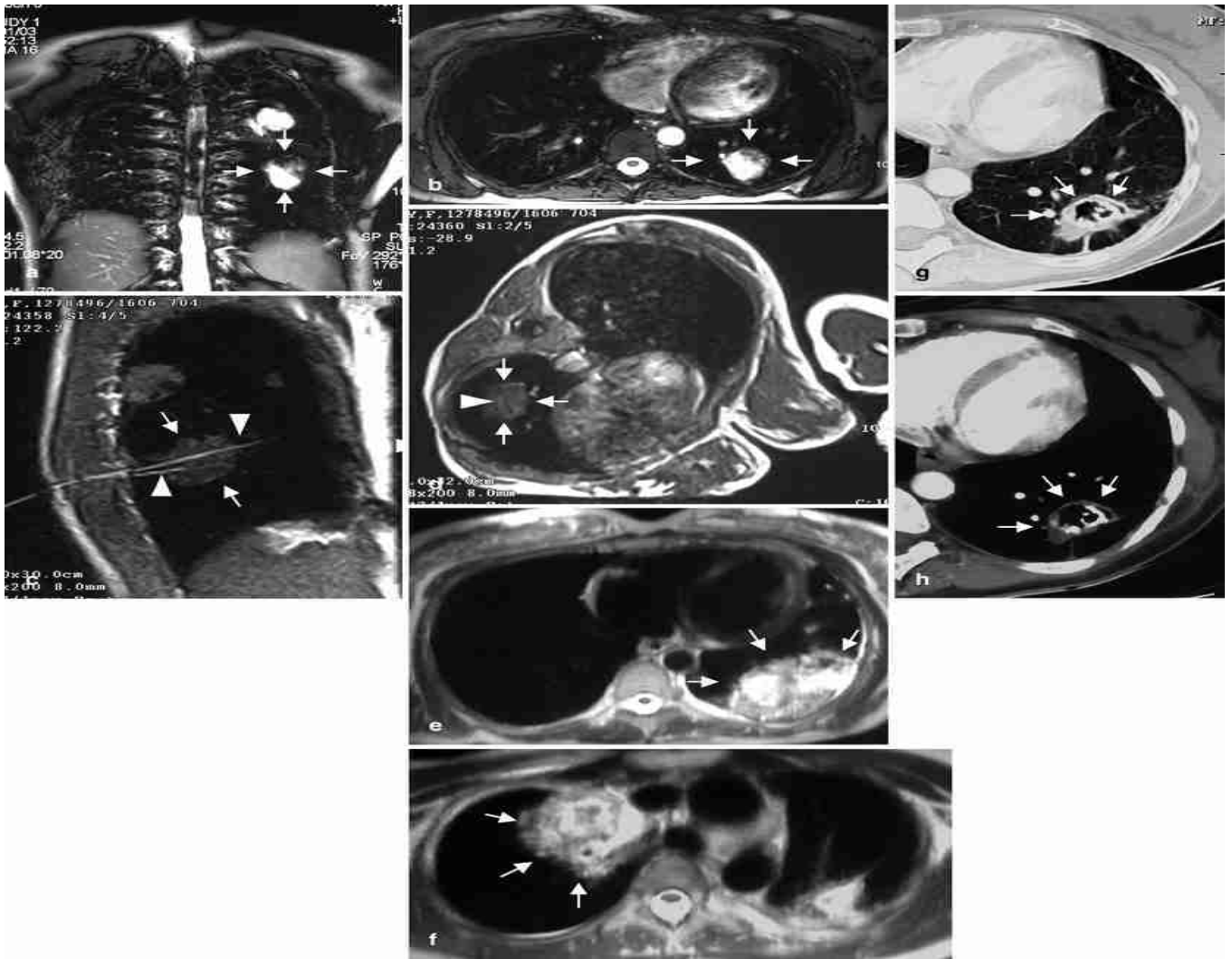


Fig. 3 **a, b** T2-weighted images (TR/TE=4.5/2.2) obtained 2 weeks before laser treatment show the pulmonary chondroma in the left lung (*arrows*). **c, d** Unenhanced T1-weighted images (TR/TE=140/12) immediately before starting the LITT treatment show the pulmonary chondroma (*arrows*) and the positioned laser fibers (*arrow head*). **e** T2-weighted images (TR/TE=1,000/57) obtained 24 h after laser treatment show the induced coagulation area (*arrows*) with some reactive perilesional tissue charring. **f** Unenhanced T2-weighted images (TR/TE=1,000/57) show the induced coagulation area (*arrows*) of the laser-treated pulmonary chondroma in the right lung 24 h after laser treatment. **g, h** CT images show a complete ablation of the chondroma 6 months after laser treatment (*arrows*)

After exact demonstration of the position of the catheter system in the center of the pulmonary lesions, the metallic stylet was removed and the patient transferred to a 0.5-T MRI system (Elscent, GE, Frankfurt, Germany) (Fig. 3c,d) where MR thermometric imaging was performed during the laser application (Nd-YAG laser 1,064 nm, Dornier, Germany). The LITT application was performed in three sessions, two times in the left and one time in the right lung.

MRI imaging showed an induced coagulation area (Fig. 3e,f) 24 h after laser treatment. In the course of regular follow-ups, complete ablation of the treated chondromas (Fig. 3g,h) and growth cessation of the extra-adrenal paragangliomas could be documented.

Discussion

Carney triad of gastric stromal sarcoma, pulmonary chondroma, and extra-adrenal paraganglioma is a rare syndrome of unknown cause primarily affecting young women. The clinical picture is chronic, persistent, and indolent. The disorder may be familial.

To date, the therapy for gastric and paraganglionic tumors has been surgical resection of the primary and recurrent tumors. Some recurrent and inoperable GISTs have been treated with STI-571 (Glivec). Unresectable paragangliomas have been treated by radiation or chemotherapy. Many of the pulmonary chondromas have been resected for diagnostic purposes; others have been left in situ when their likely benign cartilaginous nature was realized based on X-ray appearances in young patients with gastric sarcomas, particularly women.

Minimal invasive methods in the treatment of Carney triad and its various components allow a decrease of the postsurgical morbidity and offer a repeatable local treatment in order to control the paraganglionic tumors and pulmonary chondromas along with their recurrence.