Original article

Increased Uptake of Tc-99m MDP in Pulmonary Osseous Chondrosarcoma in a Patient with Carney's Syndrome

Kikuo MACHIDA, Norinari HONDA, Makoto HOSONO, Takeo TAKAHASHI, Akio KASHIIMADA
Yuji SHIMIZU, Hisato OSADA, Masahide OHMICHI, Wataru WATANABE, Takemichi OKADA
Nobutaka USUI, Nobuyuki OHTAWA, Mikinito HONDO, Keiichiro NISHIMURA and Shinji ITOYAMA*

Department of Radiological Science and Department of Pathology*
Saitama Medical Center, Saitama Medical School

Abstract

The authors describe a 33 year-old female with Carney's syndrome. Her chest roentgenogram and X-ray CT images revealed multiple calcified round shadows and bone scintigram showed multiple hot areas corresponding to the round calcified areas. She also had multiple submucosal tumors in the stomach. Pathology showed nodular stony hard mass, pulmonary chondro-osseous hamartoma, containing mature and immature, cartilage and bony tissue. Mesenchymal and fatty tissues are also observed with epithelial clefts attached with bronchial tissue. Pathological diagnosis was chondro-osseous hamartoma of the lung. In her resected stomach that caused severe anemia secondary to gastrointestinal bleeding, multiple gastrointestinal stromal tumors were found.

Neither X-ray CT nor I-123 MIBG scintigram was positive for the diagnosis of paraganglioma.

Keywords:Tc-99m MDP, hamartoma, gastrointestinal stromal tumor, Carney's triad, pigmented nodular adrenocortical disease (PPNAD)

Introduction

In bone scintigram pulmonary uptake is seen in any cause of metastatic calcification, fibrothorax, metastatic osteogenic sarcoma, pleural fluid, lung cancer, radiation therapy, alveolar microlithiasis, idiopathic pulmonary ossification, mitral stenosis, inappopriate radiopharmaceutical preparation and sarcoidosis [1]. In this article we report a 33 year-old female with hamartoma of the lung presenting with calcified nodules, and bone scintigram revealed multiple hot areas of the lung. Furthermore, as in this patient gastric stromal tumors were found later, although extraadrenal paraganglioma was not found. Carney' triad was diagnosed.

Case Report

A 33 year-old female was found multiple calcified pulmonary nodules on chest roentgenogram **(Figure 1)**, CT revealed multiple

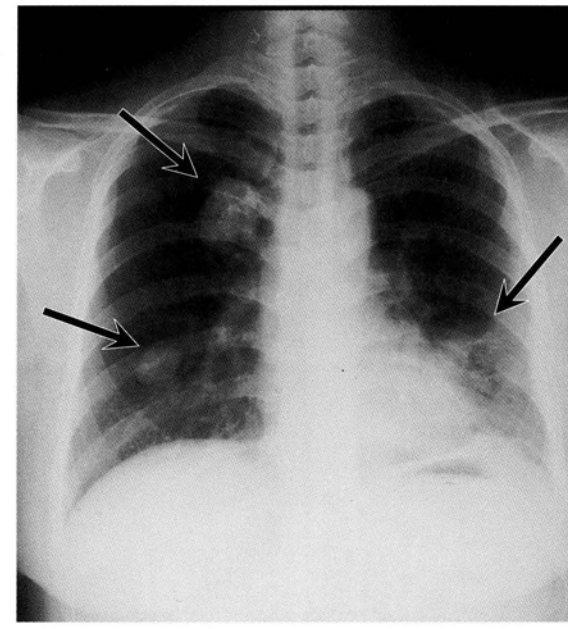
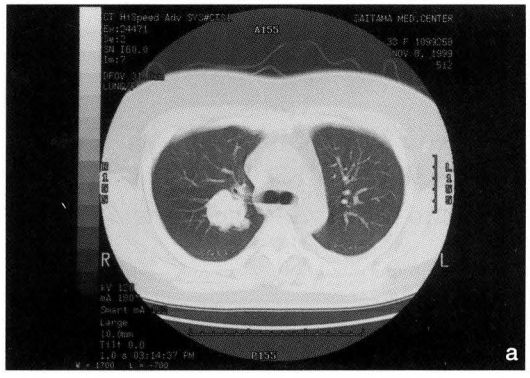


Figure 1
Chest roentogen 0 gram shows multiple calcified medicales (arrows).

Contact address of the principal author :
981 Kamoda, Kawagoe City, Saitama Pref. 350-8550 Kikuo MACHIDA
TEL. 0492-28-3512 FAX. 0492-26-5284



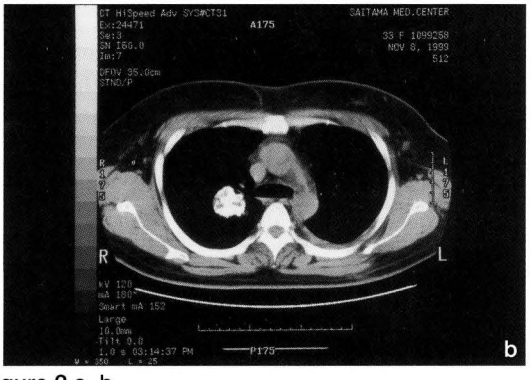
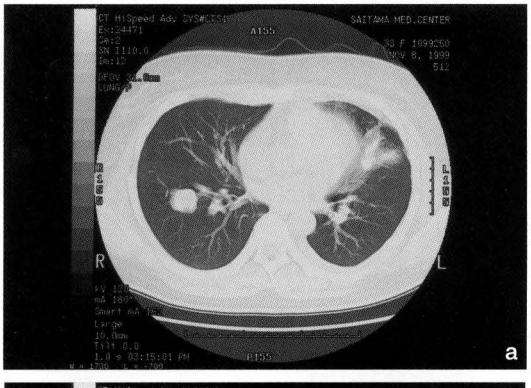


Figure 2 a, b
CT images show a calcified abnormal shadow in the right lung and left pleural thickening.



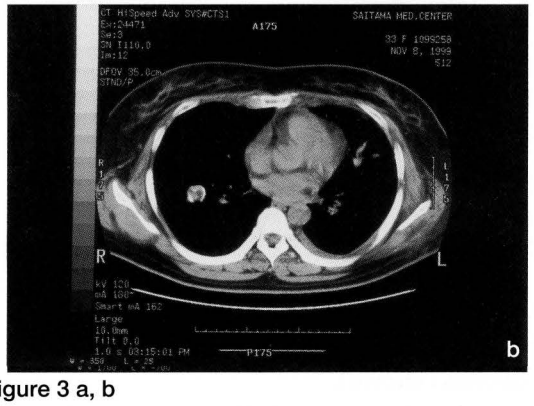


Figure 3 a, b
CT images show a calcified shadow in both lungs and pleural thickening in the left.

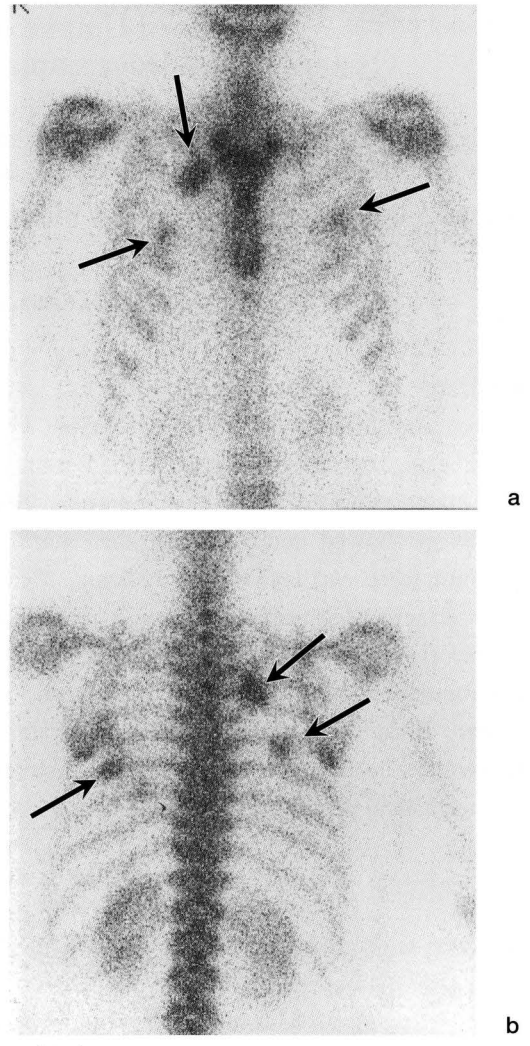


Figure 4 a, b
Bone scintigrams reveal multiple hot areas (arrows) both (a) anterior and (b) posterior view.

calcified nodules more clearly **(Figure 2, 3)**.

After intravenous administration of 740 MBq of Tc-99m MDP, bone scintigram was performed for the detection of bone diseases and revealed multiple hot areas in the lungs **(Figure 4)**.

I-131 MIBG scintigram showed no abnormal uptake in the patient.

Laboratory data included the following: leukocyte count, 8,500/mm^3; RBC counts, 5,210,000/mm^3, hemoglobin concentration, 14.5g/dl; platelet count, 503,000/mm^3; TP 7.5g/dl; Albumin, 4.3g/dl, A/G 1 78, GOT 17 IU/l; GPT 16 IU/l; LDH 233 IU/l;

gamma GTP, 16 IU/l; T.Bilirubin, 0.2mg/dl; Na, 144mEq/l; K, 4.3mEq; Cl, 107mEq; BUN, 13mg/dl; Creatinine, 0.62mgldl; Ca, 9.7mg/dl; Triglyceride, 146mg/dl; T-cholesterol, 183mg/dl; CRP, 0.3mg/dl, fasting blood glucose, 119mg/dl; CEA, 3.7ng/ml (normal 1.0-6.7); serum noradrenaline, 1,700pg/ml (normal 90-420); urine metanephrine 0.67 mg/day (normal 0.5-0.40); urine cathecolamine, 0.433ng/day (normal 0.025-0.171).

At the first surgery, nine hard tumors were resected from the left lung, and about a month later three hard tumors were resected from the right lung. Pathological finding showed stony hard masses which contained mature and immature cartilage tissue, immature and mature osseous tissue with mesenchymal and fatty tissue. Pathological diagnosis was multiple chondro-osseous hamartoma of the lung.

Four months after the first thoracotomy, she experienced tarry stool and anemia. Multiple submucosal tumors were found with esophagus gastric fiberscopy. A partial gastrectomy and gastrojejunostomy (Billroth I) was performed. Pathology revealed 7 tumors in the gastric wall and tumors contained dense spindle or ellipsoid cells, which had fine granular nucleus with nucleolus and acidophilic endoplasmic reticulum with some non-uniform size. These cells were supported by fine vascular network. Immuno-hisotology showed negative CD34, negative SMA, negative S100 protein, weak positive NSE and partially positive CD56. Mitotic activity varied from zero to 4/50 HPF. The diagnosis was multiple gastrointestinal stromal tumors of the stomach.

The postoperative course has been uneventful, and at this time the patient is doing well and lives a normal life.

Discussion

This patient illustrates the usual presentation of Carney's triad, namely a young female with pulmonary chondro-osseous hamartoma of the lung and multiple gastric stromal tumors, caused anemia secondary to gastrointestinal bleeding. Pulmonary metastases were ruled out by pathological findings of resected tumors. In 1994, Acha et al. were aware of 49 cases from literature [6], 45 female and 4 male. Their case was one of 11 patients who had the complete triad. The gastric and pulmonary involvement in the disorder is more common than that of the extra-adrenal paraganglioma system.

Pulmonary chondro-osseous hamartoma is a rare, benign tumor [3], generally asymptomatic, which is most frequently encountered while screening the patient for metastasis from leiomyosarcoma (multiple gastrointestinal stromal tumors of the stomach) or paraganglioma [6]. In our patient, she was detected her abnormal pulmonary shadow by the regular mass screening examination performed regularly by the company.

Coexistence of pulmonary metastases from gastric stromal tumors with benign pulmonary chondro-osseous hamartoma has been described [7].

Multiple gastro-intestinal stromal tumors (epitheloid leiomyosarcoma or malignant leiomyoblastoma) herald the disorder, presenting as acute or chronic anemia. Our patient showed acute anemia from tarry stool. Stromal tumor is rare which appears to be one of the most indolent tumors known [4,8-11], although the fact that it metastasize leaves no doubt that it is malignant [6]. Its behavior differs from sporadic gastric epitheloid leiomyosarcoma, which is a highly aggressive neoplasm [12,13]. Surgical excision is the treatment of choice [4,14]. No chemotherapeutic regimes have been used in these patients, due to the indolent progression of metastatic leiomyosarcoma in Carney's triad [6].

Paraganglioma is generally extra-adrenal in Carney's triad [2,4], although some cases of adrenal origin have been reported [8,15]. Described originally as functioning, several nonfunctioning cases have been reported [6,1617,18]. It is the least frequent component of the triad . I-131 MIBG scintigram was negative in this patient.

Signs and symptoms suggesting pigmented nodular adrenocortical disease (PPNAD) were not found in this case.

In conclusion we reported a Carney's syndrome, and according to Acha [6] data about hormonal findings, chromosomal abnormalities and exposure to teratogens or environmental mutagenes during prenatal life or childhood, should be gathered to investigate the pathogenesis. Association to some hereditary condition, in the patient or within the family, might give the clue to a genetic origin.

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