Lung Fibrosis in Carcinoid Syndrome

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Picture 1. Diffuse interstitial fibrosis of the lungs and multiple pulmonary nodules compatible with metastasis were demonstrated by computed tomography scan.

A 59-year-old woman presented with progressive severe bronchial constriction, cyanosis, telangiectasias, erythematous to purplish flushing, abdominal cramping, watery diarrhea, and metabolic acidosis. The patient had never complained of respiratory symptoms prior to that time. She was affected by a multi-metastatic mid-gut carcinoid tumor, histologically proven by percutaneous biopsy of the liver lesion. Serotonin plasma concentration and 24-hour urinary excretion were 76 ng/ml (normal values: 5-12) and 4,390 mg (normal values: 30-180), respectively. In addition to the primary neoplasia and metastatic lesions at the liver, computed tomography showed diffuse interstitial fibrosis of the lungs and multiple pulmonary small nodules compatible with metastasis (Fig. 1). Octreoscan showed an intense focal uptake of the radio-labeled tracer by numerous neoplastic abdominal lesions. Diffuse octreotide uptake at lungs and scant signs of focal enhancement at the right pulmonary hilum were evident as well (Fig. 2). Pulmonary fibrosis secondary to carcinoid syndrome was diagnosed. Other possible respiratory diseases were clinically excluded. Despite inten-

Picture 2. Octreoscan findings showed an intense focal uptake of radio-labeled octreotide by liver lesions and a nodular medial area in the abdomen. A diffuse pulmonary uptake of the tracer with scant signs of focal enhancement at the right hilum was also evident.

sive treatment with octreotide, interferon- α , corticosteroids, ondansetron, and cyproheptadine (1), bronchostenosis and severe impairment of gas exchange with respiratory acidosis (pH 7.238, pCO₂ 102 mmHg, pO₂ 55 mmHg) persisted. Respiratory failure progressively worsened and the patient died. Serotonin as well as other factors secreted by carcinoid tumors, like tachykinins and TGF- β family peptides, are able to enhance fibroblast activation and fibrosis process of the heart and, more rarely, the lung (2, 3), potentially leading to pulmonary hypertension and respiratory failure. In this carcinoid syndrome case, lung fibrosis was particularly relevant. Octreoscan highlighted the abdominal sites of the neuroendocrine tumor and suggested the presence, at the level of the

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lungs, of activated fibroblasts and infiltrated lymphocytes, the cell surface and are involved in the process of interstitial which express somatostatin receptors (sst2 and/or sst5) at fibrosis (4).

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