Uremic lung: The “calcified cauliflower” sign in the end stage renal disease

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A B S T R A C T
Metastatic pulmonary calcification (MPC) is a rare pathological condition consisting of lung calcium salt deposits which commonly occurs in patients affected by chronic kidney disease probably for some abnormalities in calcium and phosphate metabolism. CT represents the technique of choice for detecting MPC findings including ground glass opacities and partially calcified nodules or consolidations. We present a case of MPC in a patient affected by hepato-renal autosomic-dominant polycystic disease; chest CT revealed extensive lobar-segmental parenchymal calcification with a peculiar cauliflower shape which we called “calcified cauliflower” sign. The “calcified cauliflower” sign can be reported as a new CT pattern of uremic lung that needs to be identified for a correct diagnosis and patient management.

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1. Introduction
Metastatic pulmonary calcification (MPC) is a rare lesion, characterized by calcium salt depositing in normal lung tissue and can be caused by a large spectrum of benign and malignant conditions. In chronic kidney disease (CKD) and in patients undergoing dialytic treatment, MPC is detected at autopsy in 60–80% of cases, but it represents a rare finding at imaging studies, often occasional during studies performed for detecting other thoracic conditions [1–4].

Computed tomography (CT) is the modality of choice in demonstrating pulmonary calcifications [5,6]. Classic CT pattern of MPC in CKD was described by Hartman et al. and consisted in alveolar calcifications, calcifications in the tracheobronchial tree and in vessels of the thoracic wall [5]. During last decades, other patterns have been described, but, to our knowledge, completely calcified lobar and segmental consolidations, located in the lower lobes and with air bronchograms, have never been reported in the medical literature. We report the case of a patient affected by CKD with this new CT pattern of uremic lung that we called “calcified cauliflower” sign.

2. Presentation of case
A 71 year-old male patient affected by hepato-renal autosomic-dominant polycystic disease, with IV stage CKD and undergoing hemodialytic treatment, underwent chest CT scan. Five months before a scintigraphic diagnosis of pulmonary thromboembolism was performed and the patient was waiting for a combined hepatorenal transplant. CT examination was performed by using a 320-row device (Aquilion One, Toshiba Medical Systems, Tochigi, Japan) from the lung apices to the diaphragmatic domes without intravenous injection of contrast material in order to detect any cardiopulmonary contraindication to the transplant itself. No CT sign of interstitial edema or pleural effusion or pulmonary fibrosis was detected while an extensive calcified cauliflower-like lung consolidation involving the lingula and the left lower lobe was found, in segmental distribution and with air bronchograms (Fig. 1). Some patchy consolidations, completely calcified, were also detected in the upper lobes, but of minor extension (Fig. 2). The patient had no symptoms suggestive for pneumonia and respiratory function was normal. Calcifications within the tracheobronchial walls and in the arterial vessels of the thoracic wall, consisting with MPC in CKD, were found.

As collateral finding, CT scans passing through the upper abdomen demonstrated multiple hepato-renal simple cysts (Fig. 3).
Patient’s history for past imaging or previous episodes of extensive pneumonia revealed that the described CT lung pattern was unmodified in a previous CT examination performed two years before. The patient had no history of silica or asbestos expositions, nor barium ingestion, and no recent history of pneumonia.

The MPC pattern reported in this case and never described before consisted in an extensive lobar-segmental calcification with air bronchograms and a massive alveolar involvement, prevalent in one lung and in its lower lobe and sparing the subpleural airspaces, that resembled a cauliflower shape. That’s why it was called as “calcified cauliflower” sign.

3. Discussion

Two million people worldwide suffer from ESRD and the number of patients diagnosed with the disease continues to increase at a rate of 5–7% per year. Taiwan, Japan, Mexico, the United States, and Belgium currently have the highest prevalence of ESRD [7].

CKD causes a large spectrum of lung and thoracic alterations, usually represented by interstitial edema, pleuritis, pericarditis, cardiomegaly, pneumonia and various vascular and alveolar

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**Fig. 1.** Transverse CT scans (A-B Lung window; C-D Mediastinum window) showing extensive calcified cauliflower-like lung consolidations involving the lingula and the left lower lobe, in segmental distribution and with air bronchograms.

**Fig. 2.** Multi Planar CT Reconstruction on the coronal plane (A Mediastinum window; B Lung window) showing some patchy consolidations, completely calcified, in the upper lobes (arrows), but of minor extension as compared with the contralateral lower lobe.

**Fig. 3.** Transverse CT scan passing through the upper abdomen demonstrating extensive hepato-renal simple cysts.
Damage, due to the increased vascular permeability, channel dysregulation, increased level of some cytokines/chemokynes, increased leucocyte trafficking, decreased cellular and humoral immunity and macrophages activity.

Imaging findings in ESRD consist in atelectasis, pleural effusion, vascular congestion, parenchymal consolidation and pulmonary scarring fibrosis [3,8,9].

Metastatic calcification in extra-renal tissues, including the thorax and in particular the lung parenchyma, is another aspect of CKD, caused by abnormalities in calcium and phosphate metabolism [2,3,10,11], but no correlation between the severity and the extension of calcifications and serum levels of calcium, phosphate and parathormone or with the duration of hemodialytic treatment has been demonstrated [2,3,12,13].

MPC, although frequently (60–80% of cases) demonstrated at autopsy, is rarely diagnosed intra vitam, because often asymptomatic, and because only some patients with ESRD undergo a chest CT examination. In fact, plain chest X-ray has low sensitivity (10%) in detecting MPC, often showing only alveolar nodular opacities, mainly in the upper lungs, but calcification is often underdiagnosed or not demonstrated [2,3,12,14–16].

CT is very sensitive in detecting calcified structures, and represents the imaging modality of choice for diagnosing MPC [5,6]. The classical pattern of MPC was described by Hartman et al. as the association of pulmonary airspace opacities, with or without macroscopic calcification and with calcification of the vessels of the thoracic wall [5]. Airspace opacities are often bilateral and symmetrical [3]. The predilection for the upper lung zones is probably due to the relative hypercapnia and the consequent tissutal alkalinity [2,17,18]. Also infections and pulmonary edema could increase calcium depositing, adding a dystrophic component to the metastatic calcium salt deposition.

Other patterns of pulmonary calcification in ESRD have been described in the medical literature: pulmonary nodules with “ring calcification” by Lingam et al. [18]; ground glass opacities in the upper lobes, central and peripheral, not segmental and centrilobular by G. Gavelli et al. [3]; ill-defined fluffy nodules, areas of calcification, dense not calcific patchy or lobar consolidations, ground glass opacities, prevalent in the upper zones and with sparing of the costophrenic angles and subpleural space by Madhusudhan et al. [2].

The last one pattern is the most similar to the one we describe, having in common the sparing of the costophrenic angles and the subpleural space, but it differs for the absence of a clear air bronchogram, for the asymmetry of lesions (in our case mainly in the left lung) and their distribution (involving lingular and the left lower lobe).

Symptomatic patients with worsening dyspnea and restrictive dysfunction can undergo a conservative treatment (oxygen, vitamin D, inhaled steroids) or surgical parathyroidectomy in order to eliminate secondary hyperparathyroidism [14].

Our patient had no respiratory symptoms, and no treatment was given. Our hypothesis for that unusual distribution is an undiagnosed segmental pneumonia involving the lingula and part of the left lower lobe, which created the conditions for dystrophic calcium depositing in those lung segments with the atypical CT pattern described in this paper.

In conclusion, Metastatic pulmonary calcification on an end-stage-renal-disease background represents an occasional finding. CT is the imaging modality of choice for detecting and characterizing pulmonary calcification being able to detect both typical or atypical patterns. The calcified cauliflower sign can be reported as a new CT pattern of uremic lung in this field that needs to be identified for a correct diagnosis and patient management.

Conflict of interest

None.

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