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Idiopathic Multifocal Fibrosclerosis: Mediastinal and Retroperitoneal about A Case

N. Zaghba[1], Jalloul Wafaa[1*], F.Z. Haouassia[1], H. Benjelloun[1], K. Chaanoun[1], N. Yassine[1]

[1]Department of Respiratory Diseases, CHU Ibn Rochd, Casablanca, Morocco

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***Corresponding author:** Jalloul Wafaa
Department of Respiratory Diseases, CHU Ibn Rochd, Casablanca, Morocco

Abstract **Case Report**

Multifocal fibrosclerosis is a rare entity of unknown cause, characterized by systemic fibrosis. We report the observation of a 40 year old patient, without any particular pathological history, who presents a combination of mediastinal and retroperitoneal fibrosis of idiopathic origin.

Keywords: Fibrosis, mediastinum, retroperitoneum, Idiopathic, corticotherapy.

INTRODUCTION

Multifocal fibrosclerosis is a rare condition characterized by multi-organ fibrosis. It can be primary or secondary [1]. In our observation, we illustrate a new case of idiopathic mediastinal and retroperitoneal fibrosis.

OBSERVATION

Mr R.A, aged 40 years, smoker at 5 pack-years (PA), without any particular pathological history, presented for 1 month a dry cough, hemoptysis of small abundance and intermittent low back pain evolving in a context of conservation of the general state and apyrexia. The physical examination was essentially normal. Chest X-ray (Figure 1) revealed a dense, homogeneous, left hilar-projecting opacity with a spiculated border. Chest CT scan showed a poorly bounded, hypodense, left mediastino-pulmonary tissue mass measuring 4.5 cm × 3.5 cm (Figure 2), encompassing the left stem bronchus and lingular bronchus without an endoluminal bud, with slight enhancement after contrast injection. Bronchial fibroscopy visualized a 2nd degree inflammatory state, significant thickening of the left lobar spur with spontaneous bleeding from the lingula. The bronchial biopsies were in favor of a non-specific chronic inflammation and the bacteriological work-up was negative. A mediastinoscopy was performed, revealing a mediastino-pulmonary process, enveloping the different structures of the mediastinum, in particular the left main bronchus and the lingula. The anatomopathological study of the tumoral process showed a thick hyalinized fibrous tissue associated with lymphocytic cellular infiltrates without visualization of a tumoral proliferation or epithelioid granuloma thus concluding to a mediastinal fibrosis. Abdominal and pelvic CT showed a hypodense retroperitoneal process, slightly enhanced after injection of contrast medium, encompassing the aorta, the inferior vena cava and the distal portion of the right lumbar ureter complicated by a ureterohydronephrosis, making the appearance of retroperitoneal fibrosis. The biological work- up showed an inflammatory anemia with an accelerated sedimentation rate and an increased CRP. The etiological investigation was negative: no similar case in the family, no thoracic trauma, no medication and no radiotherapy. The immunological (anti-nuclear antibodies, rheumatoid factor, anti-CCP antibodies, ANCA) and thyroid tests were negative. Thyroid ultrasound showed no evidence of thyroiditis. Serum IgG4 assay was normal. The diagnosis of idiopathic mediastinal and retroperitoneal fibrosis was retained, resulting in a picture of idiopathic multifocal fibrosclerosis. Treatment with oral corticosteroids based on prednisone (1 mg/kg/day for 4 weeks and then progressive degression) was started. After 12 months of treatment, the evolution was marked by the disappearance of hemoptysis and low back pain with a normalization of the inflammatory syndrome and a clear regression of the radiological lesions, in particular retroperitoneal fibrosis (Figure 4).

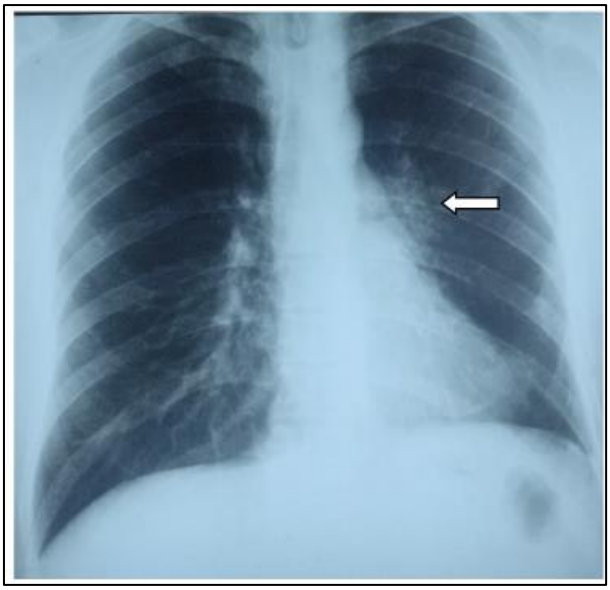

Figure 1: Frontal chest radiograph showing a left hilar-projecting opacity with ascension of the left diaphragmatic cupola

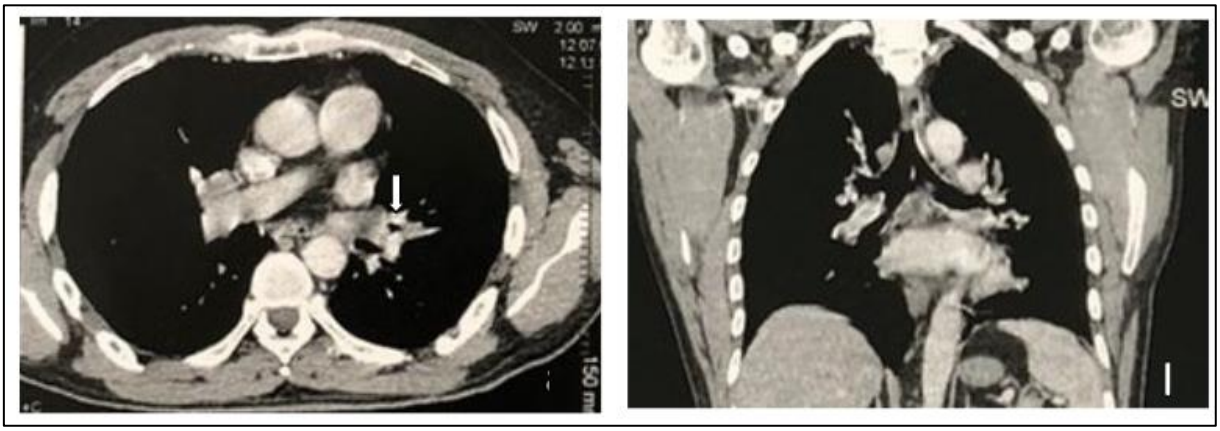

Figure 2: Thoracic CT scan in mediastinal window, axial (a) and coronal (b) sections, showing a poorly limited tissue mass with calcifications, surrounding the left main bronchus and the lingular bronchus with reduction of its caliber

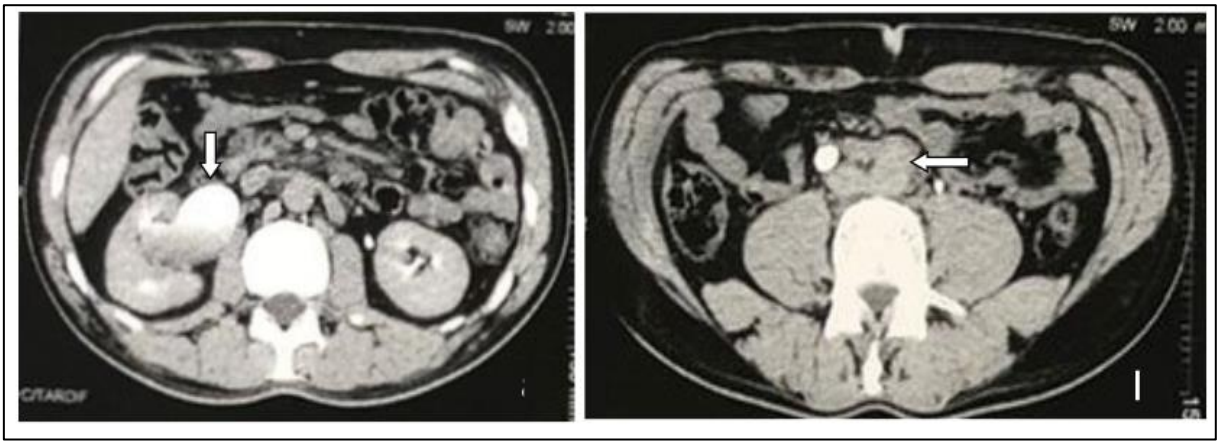

Figure 3: Uroscanner axial sections (a+b) show retroperitoneal fibrosis encompassing the right lumbar aorta and ureter, complicated by moderate ureterohydronephrosis

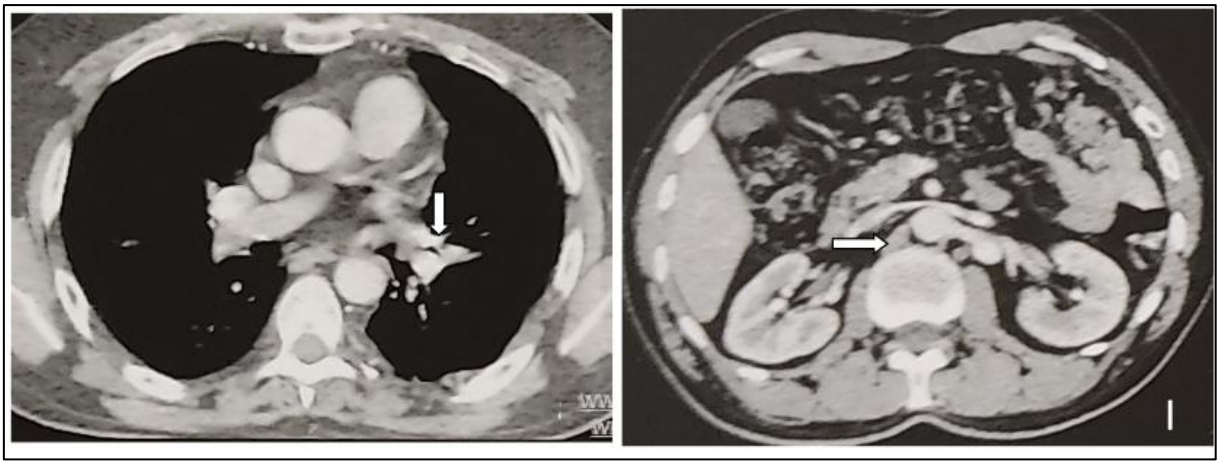

Figure 4: Thoracic (a) and abdominal (b) CT scans in axial section showing stabilization of mediastinal fibrosis and clear regression of retroperitoneal fibrosis with disappearance of the right ureterohydronephrosis

DISCUSSION

Multifocal fibrosclerosis first described in 1967 by DE. Comings *et al.*, is a syndrome of unknown etiology, characterized by multi-organ fibrosis [2]. These are mainly retroperitoneal fibrosis, mediastinal fibrosis, cervico-cephalic fibrosis, pseudotumor of the orbit, Riedel's thyroiditis, sclerosing cholangitis, and sclerosing pancreatitis, which may be combined [3]. Many cases have been reported showing two or more of these entities. However, presentation of mediastinal and retroperitoneal fibrosis is rare. In a series of 491 patients, this association was found in only 3.3% of cases [3].

Multifocal fibrosclerosis is often idiopathic as in our case, but it can be secondary to chronic infections of mycotic origin (histoplasmosis, cryptococcosis, blastomycosis, aspergillosis), tuberculosis, mycobacteriosis or bacterial infections (nocardiosis, actinomycosis) or non-infectious causes such as autoimmune diseases (lupus, rheumatoid arthritis, vasculitis, IgG4 disease), Behçet's disease, Hodgkin's disease, sarcoidosis, silicosis, radiotherapy, trauma. The origin can also be drug-induced (methysergide, practolol, bromocriptine...) [1, 4]. In our patient, the primary character was retained mainly on the negativity of the bacteriological and immunological workup and of sarcoidosis, the diffuse character of the process, and the favorable evolution under corticosteroids. Multifocal fibrosclerosis occurs mainly in the fourth decade of life and affects both sexes with a male predominance [4].

It remains asymptomatic for a long time and is often discovered by chance, but clinical manifestations may be seen, reflecting obstruction of the affected organs [3, 4]. Mediastinal involvement is most often manifested by superior vena cava syndrome, dyspnea, cough, chest pain, and rarely by hemoptysis as in our patient's case [4]. While retroperitoneal fibrosis is revealed by nonspecific signs such as abdominal or lumbar pain, which is the mode of revelation of the disease in our observation, renal colic or edema of the lower limbs have also been described in the literature. Less common symptoms such as fever and altered general condition may be present [5].

Imaging plays an important role in the diagnostic orientation. Thus, CT with contrast injection is the gold standard. It allows to visualize the fibrous plaque as a more or less voluminous tissue mass of heterogeneous density in the periphery sometimes associated with calcifications, to specify its location and its extension to the surrounding structures, and to follow its evolution under treatment [4]. At the mediastinal level, the right paratracheal, subcarinal and hilar regions are the most frequently affected [7]. MRI can also be used to evaluate the extension to neighboring organs, while PET scans can be used to identify other inflammatory sites [6, 7].

The diagnosis of certainty is based on the anatomopathological study of the biopsy specimen obtained by scannoguided puncture or by surgical biopsy; this confirms the diagnosis by showing a fibrocollagenous stroma without necrosis or vascular abnormalities and eliminates other infectious and neoplastic pathologies [4]. However, biopsy is not always essential, especially if the radiological characteristics are in favor of fibrosis, except in the case of a strong suspicion of an underlying malignant tumor, as in the case of our patient, or poor response to the initial treatment [6].

The treatment of multifocal fibrosclerosis can be medical and/or surgical. Medical treatment is currently based on corticosteroid therapy, which is the first-line treatment, thanks to its ability to inhibit fibroblast proliferation and collagen synthesis. The dosage of corticosteroids varies from 0.5 to 1 mg/kg/day for a minimum of 6 months, which can be extended to two years [1, 4, 8]. In refractory or corticosteroid-dependent cases, other immunosuppressive drugs are used, such as tamoxifen,

azathioprine, cyclophosphamide, and methotrexate [1, 4]. A study published in 2011 comparing tamoxifen to prednisone in two groups of patients (n=18 per group) with idiopathic retroperitoneal fibrosis, the first treated with prednisone, and the second with tamoxifen, showed that prednisone was better than tamoxifen in maintaining remission in these patients, and induced greater shrinkage of the retroperitoneal mass [9]. Surgery has become an adjuvant treatment since the use of corticosteroids. Indeed, it allows the management of obstructive complications, in particular urological complications, most often by endoluminal (double J catheterization) or percutaneous (nephrostomy) means, and vascular complications by arterial or venous prosthesis, although the latter does not seem to bring any real benefit [4, 6]. In our patient, corticosteroid therapy alone prolonged for 12 months allowed the disappearance of the symptoms and the clear regression of the radiological lesions.

The prognosis is generally favorable with early and adequate treatment. Indeed, regression or stabilization under corticosteroid therapy has been reported in several cases in the literature [4]. Recurrence is also frequent, especially in the first 5 years, hence the interest of close monitoring [1, 3]. Our patient is still under corticosteroid therapy with a good clinical and radiological improvement.

CONCLUSION

Idiopathic multifocal fibrosclerosis or idiopathic systemic fibrosis is a rare condition, usually benign [1, 4]. Its diagnosis is based on the suggestive scannographic aspect and confirmed by the anatomopathological study of the biopsied samples and the elimination of secondary forms, notably infectious and malignant [5]. The first-line treatment is based on prolonged systemic corticosteroid therapy to avoid recurrence [8].

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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