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RESEARCH ARTICLE

PULMONARY SARCOIDOSIS PRESENTING WITH CANNONBALL PATTERN MIMICKING LUNG METASTASES

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Abstract

Sarcoidosis is a multisystem granulomatous disease that might affect many organs, thoracic involvement is common (up to 90% of cases). Radiological findings are typical and well known by all. However, atypical findings exist and could be misleading. A 63 year old male patient was admitted to our department, he has been suffering from exertional dyspnea and dry cough, a CT scan has shown bilateral mediastinal lymphadenopathy and multiple pulmonary nodules defining a cannonball pattern, the most likely diagnosis was lung metastases. The diagnosis of sarcoidosis was finally made on clinical, radiological and histological criteria after having ruled out a malignancy, an impressive improvement was noticed under corticotherapy. The aim of this article is to shed the light on the polymorphism that sarcoidosis could present with, which might sometimes mimic a serious illness such as cancer. This latter should be excluded by performing the appropriate investigations.

Introduction:-

Sarcoidosis is a multisystemic granulomatosis of unknown aetiology that can affect several organs. Mediastino-pulmonary involvement is common and affects up to 90% of patients. It is usually made up of mediastinal lymphadenopathy, most often, non-compressive bilateral hilar and / or parenchymal involvement in the form of micronodules, reticulo-micronodular opacities or fibrosis.

Atypical mediastino-pulmonary presentations exist, such as unilateral mediastinal lymphadenopathy, isolated pleurisy, parenchymal damage of the alveolar type and above all pseudo-tumor forms including the multiple nodular form producing the appearance of cannonball pattern which raises the fear of a secondary malignancy. This form is encountered in less than 5% of cases.

We report the case of a patient with cannonball pattern on the CT scan. The taking into account of clinical and para-clinical data made it possible to make the diagnosis of sarcoidosis, supported a posteriori by the good progress under treatment.

Clinical Case:

We report the case of a 63-year-old patient, a professional cook, without toxic habits and without any medical or surgical history, who consults for exertional dyspnea classified as stage I according to the mMRC and a dry cough

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that has progressed for a year, without other associated respiratory or extra-respiratory symptoms, with preservation of its general condition.

The clinical examination on admission (taking vital signs, palpation of lymph node areas, abdominal palpation, examination of the external genitalia) was completely normal.

A frontal chest X-ray showed bilateral hilar opacities as well as multiple rounded nodular opacities at the level of the upper 2/3 of the two thoracic hemi-fields producing the appearance of cannonball pattern, initially suggesting a secondary neoplastic pathology **(Fig.1)**, without omitting other etiologies such as tuberculosis, multiple pulmonary hydatidosis or sarcoidosis.

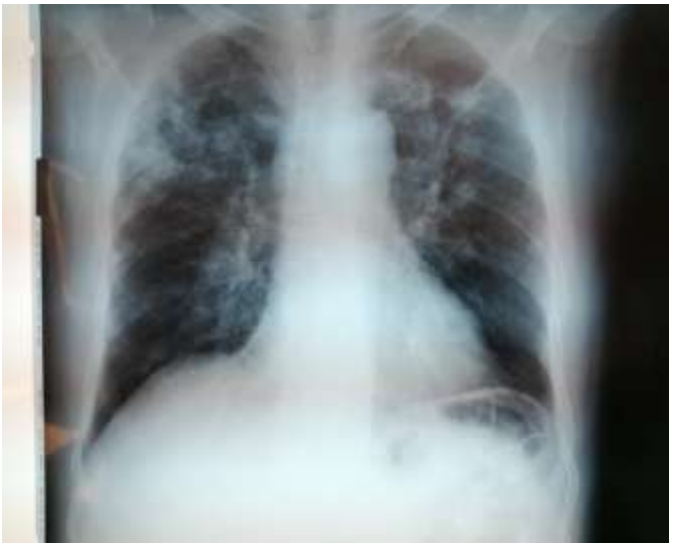

Fig 1:- Chest X-ray showing bilateral hilar opacities and multiple rounded nodular opacities at the upper 2/3 of the two thoracic hemi-fields.

A chest CT scan showed the existence of bilateral mediastinal hilar lymphadenopathy and bilateral nodular parenchymal infiltration predominantly in the apico-dorsal segments **(Fig 2).**

Fig 2:- Thoracic CT (parenchymal window) showing mediastinal lymphadenopathy bilateral hilars and bilateral nodular parenchymal infiltration predominant in the apico-dorsal segments.

A biological assessment carried out showed leukopenia (GB = 3000 / mm) and lymphopenia (lymphocytes = 1050 / mm), the dosage of tumor markers (ACE, AFP, PSA) was negative, the search for mycobacterium tuberculosis in the induced sputum was negative, negative aspergillus and hydatid serologies, negative viral serologies (HVB, HVC, HIV).

An immunologic workup including rheumatoid factor (RF), antinuclear antibodies (ANA) and anti-neutrophil cytoplasm antibodies (ANCA) was negative. The dosage of angiotensin converting enzyme and serum calcium were normal and a normal 24h proteinuria.

An abdomino-pelvic ultrasound showed a liver and a spleen of normal morphology and there were no intra-abdominal lymphadenopathy. Cervical ultrasound as well as a vesico-prostatic ultrasound were normal.

The eso-gastro-duodenal fibroscopy as well as the colonoscopy were normal.

The bone scanning returned without abnormalities.

Faced with the negativity of the neoplastic assessment, the preservation of the general condition, the non-compressive aspect of the lymphadenopathy, the location of the nodules in the upper and middle parts of the lungs, sarcoidosis was strongly suspected and the assessment supplemented by:

A bronchial fibroscopy showed a normal endoscopic appearance on the right and on the left except for a thickening of the inter culmino-lingular spur, staged bronchial biopsies found an epithelio-gigantocellular granuloma without caseous necrosis **(Fig 3)**

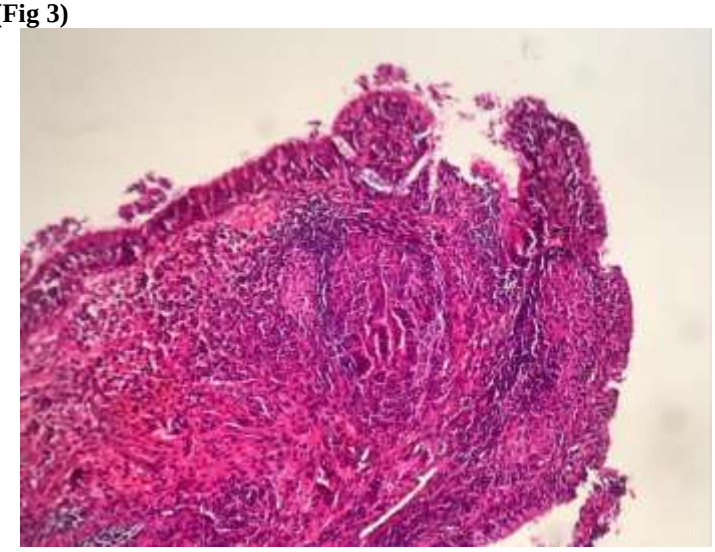

Fig 3:- Bronchial biopsy showing epithelio-gigantocellular granuloma without necrosis caseous.

Bronchial aspiration with direct examination and GeneXpert MTB / rif for mycobacterium tuberculosis, parasitic (echinococcus granulosis) and mycological (aspergillus fumigatus) studies were negative.
The biopsy of the accessory salivary glands was normal.
The plethysmography objectified a mild restrictive ventilatory disorder (CPT = 4.55 l = 70%), the measurement of The DLCO = 80%, the walking tolerance test did not show exercise hypoxemia.
The echocardiogram was normal.
The ophthalmologic examination which returned normal, there was notably no uveitis.

We retained the diagnosis of sarcoidosis based on clinical, radiological and histological arguments and put the patient on oral corticosteroids at a dosage of 0.5 mg / kg / day in the face of exertional dyspnea and restrictive ventilatory disorder. Evolution after two months of treatment was marked by dyspnea improvement and radiological cleaning on the control chest CT **(Fig 4)**.

Fig 4:- Chest CT (parenchymal window) showing radiological cleaning with almost complete regression of parenchymal nodules (after 2 months of treatment).

Discussion:-

Cannonballs pattern sarcoidosis is a rare presentation of mediastino-pulmonary sarcoidosis. Its frequency is between 1 and 5% [1 - 3].

It does not seem to have a predominance of sex or predilection age, however most of the cases described concern young subjects aged between 20 and 30 years [1 - 7]. This form is little described in elderly subjects such as our patient.

The most frequent symptoms are dyspnea and dry cough [3 - 8], general health is the rule with some exceptions [6], the accidental discovery of radiological abnormalities in totally asymptomatic patients is not rare [2, 4] our patient had exertional dyspnea and a dry cough with preservation of general condition, he meets the usual type of description in this presentation.

The biological assessment is often normal (CBC, ACE, calcium), an elevation of the converting enzyme has been noted in a few cases [3, 4, 8] the calcium level is constantly normal if requested [1 - 4, 8], the rest of the work-up aiming to rule out a differential diagnosis was always normal when it was done [1, 3, 7]. We carried out an exhaustive work-up including even the dosage of tumor markers so as not to under-diagnose a neoplasia and the whole was normal except for leuko-lymphopenia which was not described by the authors [1, 3, 4, 8].

Thoracic CT usually reveals mediastinal lymphadenopathy and parenchymal nodules of variable size which predominate in the upper and middle lobes [3 - 5, 9], such is the presentation in our patient.

Bronchial fibroscopy is almost always performed as the first line. It can find a normal endoscopic aspect [1, 2, 4, 6, 7, 9, 10] or an inflammatory aspect [4, 8]. The histological proof is obtained thanks to the Staged bronchial biopsies [1, 10] or more often with trans-bronchial biopsies [2, 4 - 6, 8 - 10]. Sometimes both techniques are inconclusive and another more invasive diagnostic method is necessary to confirm the diagnosis. MH Marques et al. reported three cases diagnosed by mediastinoscopy [4]. E. Onal et al. three cases retained after surgical lung biopsy [7]. Staged bronchial biopsies were conclusive in our case.

Faced with a picture of mediastinal lymphadenopathy and bilateral parenchymal nodules, the first etiology to look for is tumor involvement, whether it is, in the context of a primary or secondary solid tumor or in the context of a hemopathy. The age of our patient was a risk factor in favor of malignancy, since, sarcoidosis generally affects young subjects between the ages of 20 and 30, bearing in mind the existence of a late peak around the fifth decade. However, the absence of smoking in his history, of respiratory symptoms of seriousness (hemoptysis for example), of extra-respiratory symptoms (digestive or urinary) which may represent a call point for an extra-thoracic tumor, the absence of tumor syndrome (peripheral lymphadenopathy, hepato-splenomegaly), the preservation of general condition, the normality of the biological assessment (in particular of the tumor markers), the normality of the

radiological (ultrasound scans performed) and functional (digestive fibroscopies) and isotopic (bone scanning) explorations made it unlikely the possibility of a progressive neoplasia, even less at a metastatic stage and made it possible to retain the diagnosis of sarcoidosis, especially since the histology showed the existence of a granuloma without necrosis and the radioclinical discordance in this pathology is well known.

However, other differential diagnoses should be considered depending on the clinical picture and in particular:

- Infections: Tuberculosis, multiple pulmonary hydatidosis, or invasive aspergillosis.

1. The elements against tuberculosis in our patient were the absence of general signs, the negativity of the phthisiological assessment.
2. The absence of potentially contaminating contact, the tissue density of the nodules on CT and the negativity of the parasitic findings were against hydatidosis.
3. The absence of immunosuppression, the negativity of the mycological assessment and aspergillary serology were against invasive pulmonary aspergillosis.

- Connectivitis / vasculitis: rheumatoid arthritis and granulomatosis with polyangiitis (GPA) with parenchymal nodules, but the absence of extra-respiratory signs, the negativity of the immunological assessment and the presence of Mediastinal lymphadenopathy on CT are unusual in these two diagnoses.

In the light of these data, we retained the diagnosis of mediastinopulmonary sarcoidosis without extra-pulmonary involvement. A systemic corticosteroid therapy was indicated due to the symptoms and functional repercussions.The evolution was marked by clinical and radiological improvement after only two months of treatment.

Conclusion:-

Sarcoidosis is a relatively frequent disease and mediastino-pulmonary involvement is almost constant, on the other hand, certain radiological presentations are atypical and may pose a diagnostic problem for the practitioner.

Through this work, we wanted to shed light on one of these presentations, which is pulmonary sarcoidosis with cannonball pattern, whose main differential diagnosis is metastatic pulmonary localization.

A rigorous diagnostic approach will eliminate an evolving neoplasia and retain the diagnosis of sarcoidosis. Regular and close monitoring of the patient as well as the progress under treatment will confirm this diagnosis.

Conflict of Interest:

The authors declare that they have no links of interest.

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