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A Rare Case of Retroperitoneal Sarcoidosis

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Abstract **Case Report**

Sarcoidosis is a systemic granulomatosis of undetermined etiology characterised by the formation of epithelioid and gigantocellular granulomas without caseous necrosis in the affected organs. Our case has a history of sarcoidosis with left flank pain. CT scan founded a left hydronephrosis secondary to a retroperitoneal mass encasing the lumbar ureter which was shunted by a double j stent. Biopsy of the mass showed fibrous tissue with epithelioid and gigantocellular granuloma without caseous necrosis. Our patient is an extremely rare case report.

Keywords: Sarcoidosis, gigantocellular granulomas, hydronephrosis.

INTRODUCTION

Sarcoidosis is a systemic granulomatosis of undetermined etiology characterised by the formation of epithelioid and gigantocellular granulomas without caseous necrosis in the affected organs, the most frequently involved being the mediastinal lymphatic system, lungs, skin and eyes [1]. The diagnosis is based on a combination of clinical, paraclinical and anatomopathological evidence [2].

Retroperitoneal localization is extremely rare and there are only a few cases reported in the literature. We report here a case of retroperitoneal fibrosis and ureteral stenosis secondary to sarcoidosis.

CASE

A 52 years old women with a history of sarcoidosis treated with corticosteroids; last cure received in 2015; presented with recurrent left flank pain computed tomography of the abdomen revealed a left hydronephrosis secondary to a retroperitoneal mass encasing the lumbar ureter which was shunted by a double j stent (fig1). Biopsy of the mass showed fibrous tissue with epithelioid and gigantocellular granuloma without caseous necrosis (Fig 2). The biological evaluation showed a persistent inflammatory syndrome with lymphopenia in favour of the activity of the disease. Without evidence of infection, malignancy or any additional inflammatory process as an explanation for the pathologic findings, the diagnosis of retroperitoneal sarcoidosis was retained and a treatment with corticosteroids was prescribed.

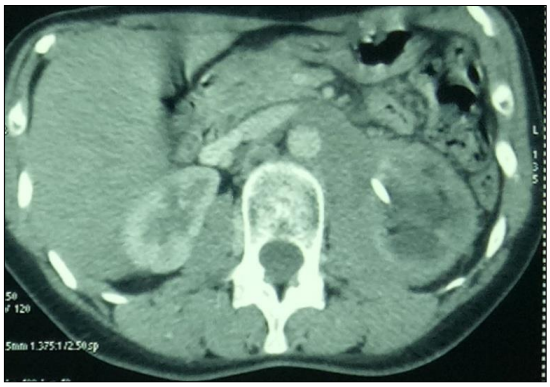

Fig. 1: Computed tomography of the abdomen, left hydronephrosis secondary to a retroperitoneal mass encasing the lumbar ureter

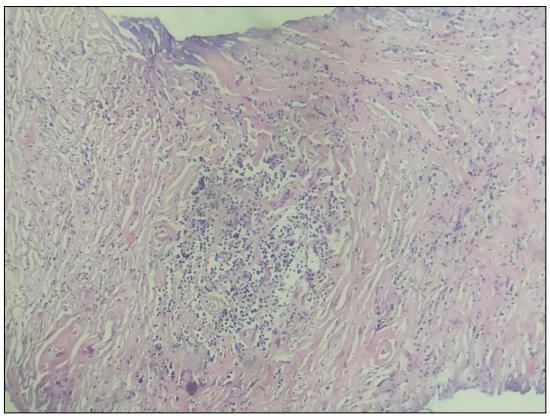

Fig. 2: Fibrous tissue with epithelioid and gigantocellular granuloma without caseous necrosis

DISCUSSION

Sarcoidosis is a systemic disorder characterized by the aberrant development of granulomas within various organs in the body. The lungs are involved in 90% of patients, and the skin, eyes, and heart are affected in a significant fraction of patients. The disease remits within 3 years in most patients, whereas 10% to 30% of patients develop chronic disease requiring ongoing treatment [3].

According to the American Thoracic Society and the British Thoracic Society, despite the absence of objective measures to establish it, the diagnosis of sarcoidosis must be based on three major criteria (i) a compatible clinical presentation; (ii) the presence of non-caseous granulomatous inflammation in one or more organ tissues; and (iii) the exclusion of alternative diagnoses of granulomatous disease [4].

The triad of a CD4/CD8 ratio greater than 4:1, lymphocytes greater than 16% on BAL and non-caseating granulomas in EBUS-guided ganglion biopsy, has a positive predictive value of 100% and of 81% in distinguishing sarcoidosis from pulmonary interstitial disease and other diseases, respectively [4].

Sarcoidosis is a challenging pathology due to the potential multisystemic involvement, making rare and atypical manifestations particularly difficult to interpret and associate with the disease. A high index of clinical suspicion is needed in order to recognize them [4].

The histopathological hallmark of sarcoidosis is the presence of non caseating granulomas in involved organ systems. The lungs are the most common site of granuloma formation, with up to 97% of patients having intrathoracic involvement [5].

The most common sites of extrapulmonary involvement are the skin, eyes, liver, and reticulo-endothelial system with rarer renal, cardiac, and neurologic involvement. Only 8% of patients present with isolated extrapulmonary disease in the absence of pulmonary involvement with the most common presentation in this group being isolated cutaneous sarcoidosis [5].

Differential diagnosis are: neoplasms, including lymphoma; mycobacteriosis, such as tuberculosis; other infectious aetiologies (fungi, cytomegalovirus, EBV and HIV); drugs and connective tissue diseases [4].

Corticosteroids remain the first line of treatment although specific doses and tapers remain controversial. Second line therapies such as immunomodulating therapy with hydroxychloroquine, methotrexate, azathioprine, mycophenolate mofetil, leflunomide, and cyclophosphamide are occasionally used as corticosteroid sparing therapies to lower the risk of adverse events associated with long term steroid use [5].

Patients with sarcoidosis have a shorter life expectancy than the general population [6].

In Western countries, most sarcoidosis deaths are due to advanced pulmonary fibrosis leading to respiratory failure, pulmonary hypertension or both and less commonly, cardiac and CNS sarcoidosis or portal hypertension. In Japan, the main cause of mortality in sarcoidosis patients is cardiac involvement, which is responsible for 77% of deaths in people with sarcoidosis [6].

Our case report a very rare case of retroperitoneal fibrosis and ureteral stenosis secondary to sarcoidosis, the diagnosis was found on clinical and histological criteria after the exclusion of alternative diagnoses of granulomatous disease.

CONCLUSION

Retroperitoneal fibrosis can be secondary to multiple etiologies, sometimes very rare and difficult to diagnose, such as sarcoidosis; clinical imaging and biopsy most often make it possible to find the diagnosis.

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