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Case Report

Isolated Splenic Sarcoidosis: A Case Report with Review of Literature

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Abstract

Sarcoidosis is an uncommon chronic multisystemic fibro-inflammatory disease of unknown aetiology, characterized histologically by the presence of non-caseating granuloma. We report the 15th case of isolated splenic sarcoidosis in a 71-year-old Nigerian female, which was discovered incidentally while being managed for acute intestinal obstruction due to volvulus. Abdominal ultrasound scan revealed a large multinodular spleen. Open splenectomy was performed and the diagnosis of splenic sarcoidosis was confirmed histologically. Her post-operative condition has been stable. Sarcoidosis should always be considered in isolated nodular splenomegaly with or without symptoms.

Keywords: Sarcoidosis, Spleen, Laparotomy, Lymphoma.

INTRODUCTION

Sarcoidosis is an uncommon multisystemic fibro-inflammatory disease, of unknown aetiology, characterized by formation of non-caseating granulomas with presence of abundant activated macrophages, and consequent interference with organ function and structure. The disease affects both gender of all ages but typically affects third to fifth decade [1]. The disease typically presents with multiple organ involvement with ninety percent of cases having pulmonary involvement. Splenic involvement in multi-organ sarcoidosis has been reported to range from 10-50% of patients [2]. Isolated splenic sarcoidosis is extremely rare and to the best of our knowledge, only 14 of such cases have been documented in medical literature. We report another case of isolated splenic sarcoidosis involving a 71-year-old female, which was discovered incidentally and was managed by performing splenectomy.

CASE REPORT

A 71-year-old Nigerian woman, and a known hypertensive, presented with three days history of abdominal pain and distension and one day history of bilious non-projectile vomiting.

She had no history of exertional dyspnoea, chronic cough or joint pain. She had appendectomy 20 years ago on account of acute appendicitis.

Examination revealed an elderly woman in painful distress. She was afebrile, anicteric and moderately dehydrated. SpO2 in room air was 97%. Her respiratory rate and heart rate were within normal range. Her blood pressure was 160/70 mmHg, and only heart sound I and II heard. Her chest was clinically clear. Abdominal examination revealed a moderately distended abdomen with generalized tenderness. A moderately enlarged spleen was detected, but no hepatomegaly, the kidneys were not ballotable or any other intra-abdominal mass detected. Digital rectal examination revealed an empty rectum.

Her full blood count, renal function test, liver function test, and pulmonary function test were within normal range. Retroviral test was negative. Abdominal X-ray and ultrasound showed small bowel obstruction and splenomegaly with multiply hypoechoic areas.

An assessment of intestinal obstruction with splenomegaly was made. She subsequently had

explorative laparotomy. Findings during surgery included small intestine volvulus over a thick fibrous adhesion and large multinodular spleen. Adhesiolysis was done, bowel viability was ascertained with evidence of peristalsis, return to normal colour and splenectomy on account of suspected case of lymphoma.

The immediate post-operative condition was satisfactory. The spleen was sent for histological evaluation. Gross examination revealed an enlarged spleen that weighed 350g, measuring 14.0 x 8.0 x 6.0cm with multinodular surface. Cut section showed multinodularity of varying sizes. Histology showed non-caseating granuloma with multiple giant cells, lymphocytes and epithelioid cells. Acid fast bacilli stain was negative. A diagnosis of isolated splenic sarcoidosis was made.

DISCUSSION

The incidence of sarcoidosis varies within countries and across regions from 1-15% per 100,000 population [3]. The actual prevalence in Nigeria is unknown because of paucity of data, lack of diagnostic facility, lack of expertise and endemicity of mimicking diseases such as tuberculosis.

Isolated splenic involvement is however extremely rare with only 14 cases reported in literature. Previous reports showed age range of 18-76 years with overwhelming female preponderance, undermining the protective role of oestrogen against sarcoidosis [3]. Clinical features reported include nodular splenomegaly, abdominal pain, headache, night sweat, weight loss, and features of hypersplenism. Ogiwara *et al*, reported a case with hypoglycaemia due to ectopic insulin-like hormone secretion [7]. Dennis *et al.*, and Joglekar *et al.*, reported cases presenting with hypercalcemia [7, 11], while Cuilliere-Dartigues *et al*., reported a case presenting with agranulocytosis [5]. In most cases, the lesion is multiple (12 out of 14 case). Some are however clinically silent and were discovered coincidentally during radiological imaging or laparotomy for other reasons [4-15]. In our case, it was discovered while the affected patient was managed for acute intestinal obstruction.

Its aetiology is unknown although various studies have implicated various genetic and environmental triggers ultimately resulting in dysregulated immune system leading to formation of granulomas and inability to resolve them. Obesity tend to generate proinflammatory environment that tend to favour granuloma formation while oestrogen and tobacco has protective effect [3]. Sarcoidosis of the spleen has also been reported to develop following immunosuppressive effect of malignant tumour [3].

Earlier reports have shown that laboratory investigations are not helpful and radiological finding not specific [1-15]. Nodular splenomegaly has been reported in previous reports, in line with our findings which can be confirmed with USS, CT and PET-CT [1]. The presence of non-caseating granuloma on histological sections of the spleen, supported with clinical and histological exclusion of other possible causes of splenic granulomas such as tuberculosis, brucellosis, melioidosis and syphilis or occupational exposure to foreign material such as beryllium or talc is diagnostic. The absence of systemic infection, negative VDRL test and negative AAFB test are in favour of splenic sarcoidosis in the index case. A local sarcoid reaction would have further strengthened the diagnosis [1], but is not applicable because splenectomy was already performed and there was also no evidence of lymphadenopathy.

There is no established treatment protocol for isolated splenic sarcoidosis. In most of the cases of isolated splenic sarcoidosis reported, patient recovered after splenectomy was performed [1-15], while steroid administered to one of the patients [14]. Indications for splenectomy includes severe abdominal pain arising from splenomegaly, functional asplenia, hypersplenism, splenic rupture or because of strong suspicion for an alternate diagnosis. In our case, splenectomy was done because of strong suspicion for lymphoma.

In conclusion, we report a rare case of sarcoidosis in a 71-year-old Nigerian female. Sarcoidosis should always be considered in isolated nodular splenomegaly with or without symptoms and should diagnosed as sarcoidosis if there are histological features of non-caseating granuloma in addition to histological and clinical exclusion of other causes of non-caseating granuloma.

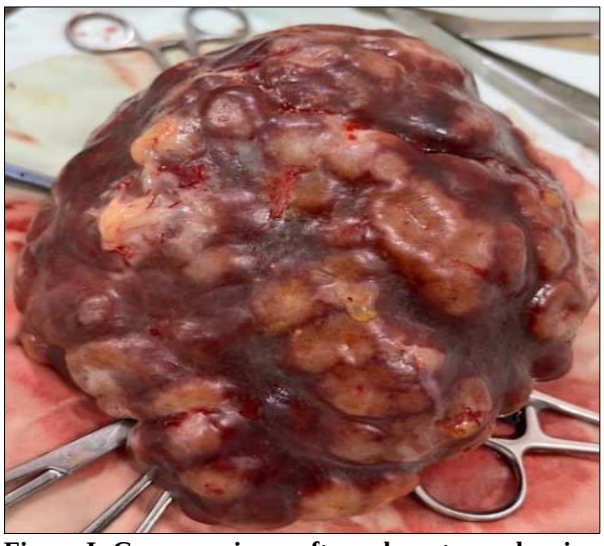

Figure I: Gross specimen after splenectomy showing an enlarged spleen with multiple nodules throughout its entire surface

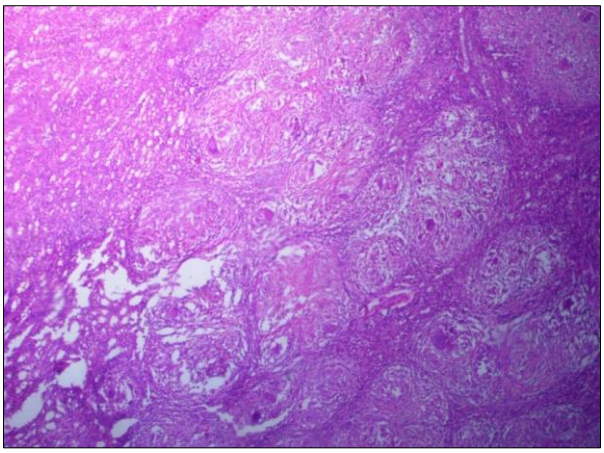

Figure II: Histopathology showing splenic parenchyma with a discrete, round to oval non-caseating granuloma composed of epithelioid cells, macrophages, fibroblasts, and multinucleated giant cells (x40 magnification)

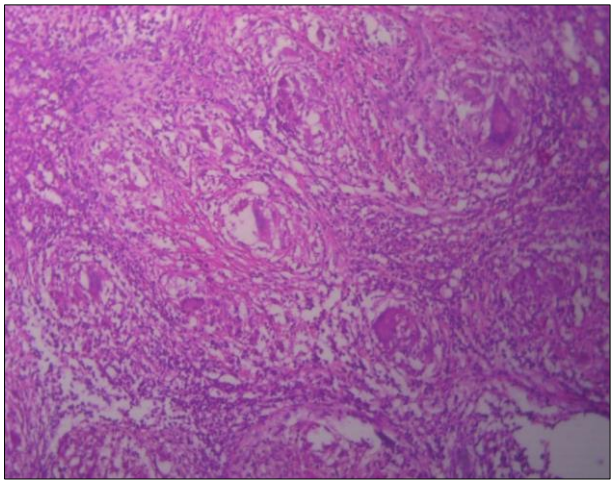

Figure III: Histological sections of splenic parenchyma showing non-caseating granuloma. (x100) magnification

Table 1: Summary of 14 Reported Cases of Isolated Splenic Sarcoidosis

Number	Author/year	Age (Years)	Gender	Symptoms	Number of lesions	Treatment	Ref
1	Giovinale *et al.*, 2009	32	F	Epigastric pain	Multiple	LS	4
2	Giovinale *et al.*, 2009	53	F	Abdominal pain	Single	LS	4
3	Joglekar *et al.*, 2009	46	F	Back and leg pain	Multiple	OS	5
4	Cuilliere-Dartigues *et al.*, 2010	18	M	Night sweat	Multiple	LS	6
5	Ogiwara *et al.*, 2010	74	F	Night sweat Palpitation	Single	OS	7
6	Palade *et al.*, 2012	66	F	Anaemia	Multiple	LS	8
7	Bauones *et al.*, 2014	37	F	Chronic abdominal discomfort	Multiple	LS	9
8	Souto *et al.*, 2014	29	F	-	Multiple	LS	10
9	Ruiz Serrato *et al.*, 2015	37	F	Abdominal pain	Multiple	LS	11
10	Dennis *et al.*, 2015	65	M	Headache, Weight loss	-	HALS	12
11	Sreelesh *et al.*, 2017	50	F	Weight loss	Multiple	OS	13
12	Bachmeyer *et al.*, 2017	56	F	Weight loss	Multiple	Steroid	14
13	Gaudemer *et al.*, 2018	42	F	Epigastric pain	Multiple	LS	15
14	Kobayashi *et al.*, 2020	76	F	None	Multiple	LS	1
15	Present case	71	F	None	Multiple	OS	

Key: LS, laparoscopic splenectomy; OS, Open splenectomy; HALS, Hand assisted laparoscopic splenectomy; Ref, reference; F, female; M, male

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