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Causes and Clinical features of patients with Hypersplenism: a case study conducted in Ibn Sina and Soba Teaching Hospitals, Khartoum, Sudan.

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Abstract

Objective: The main objectives of this paper are to identify the main causes of hypersplenism in Sudan and also to see how far the splenic size correlates with degree of thrombocytopenia.

Methods: it is "a descriptive hospital- based study" in which a total of 100 patients meeting the diagnosis of hypersplenism were studied for a period of 12 months (from January 2003 up to January 2004).Most of these patients were admitted to Khartoum, Soba and Ibn Sina Teaching Hospitals. Patients above 15 years of age were included in the study. The data was collected through a questionnaire containing a detailed medical history, thorough physical examination and detailed laboratory investigations. The Data was analyzed using SPSS program, simple tabulations and frequency distributions were conducted.

Results: 100 patients were included in the study, 72% of them were males and 28%were females. 50% patients were due to schistosomal portal hypertension, 6% of them had liver cirrhosis beside periportal fibrosis (PPF), visceral leishmaniasis detected in 30% of patients, tropical splenomegaly syndrome (TSS) in 9%, liver cirrhosis in 6%, and in 5% no cause could be found. The majority of patients had gross splenomegaly. The main presenting symptoms were fever, symptoms of anemia, bleeding tendency and loss of weight.

Conclusion: The common causes of hypersplenism in our study are potentially preventable conditions. There was no correlation between splenic size and severity of thrombocytopenia.

Keywords: Hypersplenism, Schistosomal portal hypertension, Visceral Lesishmaniasis

Introduction

Hypersplenism is a clinical syndrome of varied causes. It is characterized by: 1-splenomegaly, which may be only moderate**,**2-pancytopenia or a decrease in the number of one of blood cells (cytopenia), however neutropenia is less common than anemia and thrombocytopenia,3- normal production or hyperplasia of the precursor cells in the marrow or a so called maturation arrest,4 decreased red blood cells survival and,5- decreased platelet survival.**1**The incidence and aetiology of splenomegaly is strongly dependent on the geographical location. The aims of this study are to know the etiology of hypersplenism and to correlate splenic size with the degree of thrombocytopenia.

Methods

This is "a descriptive, cross-sectional hospital-based study" conducted at Khartoum, Ibn Sinna and Soba Teaching Hospitals in the period between Jan 2003 and Jan 2004. A total of100 patients meeting the inclusion criteria were included in this study,. The only inclusion criteria of the patients in this study were adult of age more than 15 years with splenomegaly and cytopenia or pancytopenia with normal or hypercellular bone marrow. Cases with hematological malignancies, aplastic anemia and drug-induced pancytopenia were excluded. All patients were examined according to a clinical protocol of data-base consisting of questionnaire, clinical examination and investigations. Features in the history and examination like patient age, sex, occupation, residence and presenting complaints, past history of schistosomiasis, malaria, blood transfusion, and jaundice and alcohol consumption were

collected. General examination was conducted noting for pallor, lymphadenopathy, spleen and liver size. Investigations done included: complete blood count (CBC),and abdominal ultrasound which was done for all patients to study the echogenicity of the liver for PPF and liver cirrhosis, liver size was measured and both portal and splenic veins diameters were recorded. Splenic size and echogenicity were determined, any focal lesion was noted and presence or absence of abdominal masses. Periportal fibrosis (PPF) develop develops from an inflammatory response to the portal venous embolization of schistosoma mansoni ova. Macroscopic appearance of Schistosomal PPF is pathognomonic for this condition, sonographic pattern is equally distinctive .Endoscopy was done for those with suspected upper GI bleeding. Bone-marrow aspiration was done for all patients and lymph node aspiration and study was also done in some patients with VL looking for leishman Donovan bodies. Diagnosis of TSS in our patients was made through exclusion of other causes of gross splenomegaly, positive malaria antibodies and clinical and hematological response to anti-malarials. After designing the master-sheets, all variables were introduced into the computer using D-base III for data entry. Consistency checks and analysis was carried out using Statistical Package for Social Sciences (SPSS).

Results

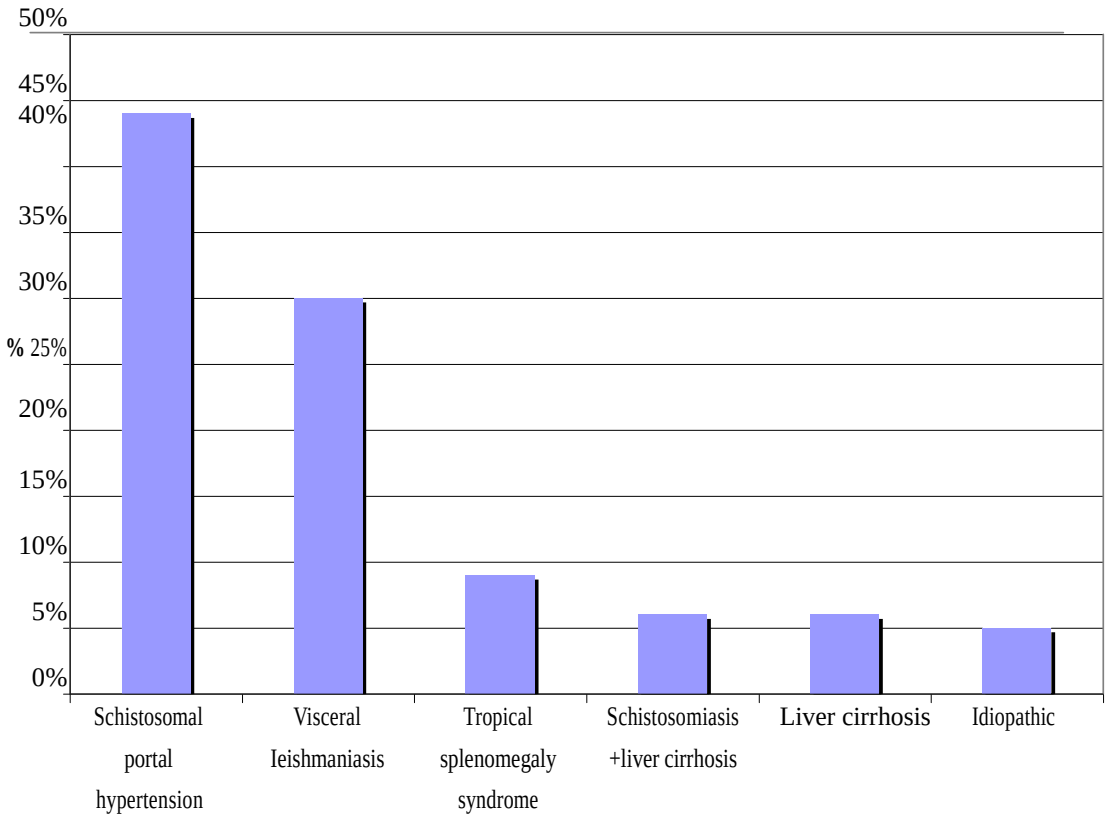
Most of patients were young adults; half of the patients were from central Sudan.–as shown in table 1

Table 1: Some demographic characteristics of the patients

Characteristics	***N=%***
Gender	
Males	72
Females	28
Regions	
Central Sudan	50
Khartoum State	20
Western Sudan	11
Eastern Sudan	10
Southern Sudan	9
Age groups	
15-20	9
20-40	60
40-60	28
>60	3
Total	***100***

The etiological diagnosis in these patients of hypersplenism is shown in Fig. 1

Figure (1): The aetiological diagnosis in 100 Sudanese patients with hypersplenism



The Majority of patients have splenomegaly >8cm and hepatomegaly as shown in table 2.

Table 2: Liver size and the spleen size measured clinically in cm in 100 patients with hypersplenism in Sudan.

Characteristics	**N=%**
Liver size	
Enlarged	61
Shrunken	12
Normal	27
Spleen size	
< 8 cm	8
8-15 cm	68
> 15 cm	24
Total	100

The main symptoms were: fever which was present in 64 patients, symptoms of anemia in 84 patients, bleeding tendency in 60 patients and loss of weight in 54 patients. Hemoglobin was found to be less than 50% in 78 patients and more than 50% in 22 patients White blood cell count was below 2000/cm in 60 patients between 2000 and 3000/cm in 28 patients and more than 3000/cm in 12 patients. Platelet was less than 50.000/cm in 49 patients between 50.000-100.000/cm in 41 patients and between 100.000-150.000/cm in 10 patients.

All patients underwent bone marrow aspiration and study, hypercellularity seen in 55 % of patients, while normo-cellularity was seen in 45% of cases. Hypercellular was found in 55 patients and normal cellularity in 45 ,Leishman donovan bodies were detected in 28 patients . Ultrasonographic findings detected periportal fibrosis in 50 patients, liver Cirrhosis in 6 patients, while combined P.P.F. and liver cirrhosis in 6 patients, as cites in 38 patients.

Discussion

Hypersplenism can result from splenomegaly due to any cause. It is commonly seen with splenomegaly due to haematological disorders, portal hypertension, and rheumatoid arthritis, Felty's syndrome and lymphoma.**2**However there is geographical variations. There are very few studies on hypersplenism. In this study we found that the common causes of hypersplenism were schistosomiasis which occurred in 50% of patients, 6 of them had liver cirrhosis beside PPF. VL occurred in 30%, TSS in 9%, liver cirrhosis in 6% and in 5% no cause was found. These results are different from the study conducted by Mustafa *et al* in 1965 in which he found schistosomiasis as a cause of gross splenomegaly in 20%, visceral leishmaniasis in 16% and TSS in 26%.**3**

This discrepancy in the results can be explained by the more extension of irrigation projects during the last 3 decades, beside collapse of the national programmes that aimed to combat infectious diseases, in addition to that, our data was collected from 3 major hospitals in Khartoum, moreover improvement in diagnostic techniques during the last 3 decades will no doubt add more to this disagreement in results.

Another two studies conducted done in Sudan & Yemen in 1996,1997 by Dahawi *etal and* Abdalhafeez *et al* showed that schistosomal portal hypertension was the commonest cause of gross splenomegaly. **4,5**

A fourth study done in India by Balji *et al* and colleagues in 2005 showed etiological and clinical results similar to ours with non-cirrhotic portal hypertension being the commonest cause. **6**

True cirrhosis in PPF does not exist and could occur only if there is concomitant cause. 7,8. In our study 6 patients with periportal fibrosis (PPF) also had liver cirrhosis which is Possibly due to coexisting cause like viral hepatitis or alcohol consumption.

Most of patients in our study (60%) were young adults their ages ranging between 20-40 years, This reflect how such problems have negative impact on the economy of the country by affecting the most productive groups.

Associated symptoms and signs of hypersplenism are typically related to the underlying disorder and may include fever, Pallor, dyspnea, weight loss, bruising, and petechiae. **9** The main symptoms and signs in our study are fever which was found in 64% of patients, anemia in 80% o, bleeding tendency in 60% and weight loss seen in 54% of cases.

In portal hypertension, hypersplenism is common and thrombocytopenia is the most common feature. Platlet counts are usually around 100,000/cumm and counts below 50,000/cumm are rare. Leucopenia occurs occasionally. The spleen is the principal producer of antibodies aimed at circulating blood cells but it has confirmed only for antiplatelet antibodies, anaemia in patients with splenomegaly is in part due to dilution of red cells in an expanded plasma volume. **10, 11** So in our study to correlate the size of the spleen with the haematological findings it is more reliable to take platelet counts as an example. We had found that 30% of those with splenic size between 8-15 cm their platelet count was less than 50.000/cm, whereas 32.5% of those who had splenic size < 8 cm their platelet was less than 50.000/cm, so this indicates that no relation between the size of the spleen and the severity of cytopenia, this result is supported by Balji*et al* study in which he found that no significant correlation between splenic size and severity of hypersplenism. Also it is well documented that spleen size is not a reliable guide to splenic function.**12**

Conclusion

Schistosomal portal hypertension is the commonest cause of hypersplenism, and there was no direct correlation between the splenic size and degree of thrombocytopenia.

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