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Sinus histiocytosis with massive lymphadenopathy, (Rosai-Dorfman Disease) with cholastatic jaundice in an HIV positive patient

Kgomo Mpho[1,2], Elnagar Ali Ahmed[1], Mashoshoe Kgataki Sam[1], Thomas Preetha[1], Ngwata Portia[1], and Bida Mishack[2]

1. Department of Gastroenterology, University of Pretoria, South Africa
2. Department of Anatomical Pathology, University of Pretoria, South Africa

CASE STUDY

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Corresponding Author:
Kgomo Mpho
Department of Gastroenterology and Anatomical Pathology, School of Medicine, University of Pretoria, Gezina, Pretoria, 0001 South Africa
Email: kgomomk@worldonline.co.za

ABSTRACT

Introduction
Sinus histiocytosis with massive lymphadenopathy is a rare histiocytic disease first described by Rosai and Dorfman in 1969. The typical presentation is that of cervical and often widespread painless lymphadenopathy.

Case report
We present a 33-year-old gentleman, with one-month history of pruritus, abdominal pain, jaundice and dark urine, jaundice, scratch marks, large inguinal lymph nodes, an enlarged liver and spleen. CT scan confirmed ultrasound findings and suggested that splenic lesions were micro-abscesses. Biopsy of the lymph nodes revealed features in keeping with sinus histiocytosis with dermatophatic lymphadenopathy.

Discussion
This is our second case of sinus histiocytosis in two years.

Key Words
Sinus histiocytosis, HIV, cholastatic jaundice

Implications for Practice:

1. What is known about this subject?
It is a rare disease with no known cure.

2. What new information is offered in this case study?
This is the first case of cholestatic and HIV reported in the literature.

3. What are the implications for research, policy, or practice?
We need to look for sinus histiocytosis as a cause of generalized lymph nodes in HIV patients.

Background

Sinus histiocytosis with massive lymphadenopathy is a rare histiocytic disease first described by Rosai and Dorfman in 1969. It predominantly affects children and young adults with a mean age being 20.6 years.

The typical presentation is that of cervical and often widespread painless lymphadenopathy with associated fever and weight loss.[1,2]

The clinical course is usually self-limited, with a tendency to resolve spontaneously.[3] It has become evident that it is not always limited to lymph nodes but that it can involve a large variety of extra-nodal sites. The incidence of extra-nodal involvement is approximately 40 per cent in recent series.[3] The commonest sites to be involved by Rosai-Dorfman syndrome include bone,[4] skin and soft tissue,[5,6] central nervous systems,[7] eye and orbit,[8] and respiratory tract.[9]

The site least likely to be involved by Rosai-Dorman syndrome is the digestive system as reported in the Rosai Dorfman Disease (RDD) Registry.[3]

Liver involvement is even less common. Gregory et al., reviewed 11 cases whereby digestive was involved and found liver involvement in only five cases.[10] We previously reported a case of Sinus histiocytosis with cirrhosis of the liver in our institution[11] and this is our second case of sinus histiocytosis with liver involvement in two years.

Case Details

We present a 33-year-old gentleman who presented with one-month history of pruritus, abdominal pain, jaundice and dark urine. There was no nausea loss of weight or appetite. He had no significant past medical history and was of sober habits.

Clinical examination revealed jaundice and scratch marks, large inguinal lymph nodes, an enlarged liver and spleen with normal vital signs.

Ultrasound revealed a 17cm hepatomegaly with normal biliary ducts with hyperechoic nodules in the right lobe, a 13cm by 8cm spleen with multiple hyperechoic nodules of varying size.

Blood tests showed an obstructive picture with an elevated INR. HIV test was negative.

CT scan confirmed ultrasound findings and suggested that splenic lesions were micro-abscesses Figure 1.

The patient was started on anti-TB therapy and phenergan cream and tablets plus ursodeoxicholic acid. He came back after 10 days with worsening pruritus at which stage an MRCP was done. This showed beading of both intra and extra-hepatic ducts. ERCP was then done to identify any dominant strictures and it was found to be normal. Biopsy of the lymph nodes revealed features in keeping with sinus histiocytosis with dermatophatic lymphadenopathy. No malignant or infectious agents were found Figure 2.

Patient was started on steroids, anti-histamines and ursodeoxycholic acid. He continued to deteriorate over a six months' period at which time HIV was repeated which came back positive with a CD 4 of 146. He was started on HAART but demised a few months later.

Discussion

Sinus histiocytosis is a rare disease with lessons only from case reports. The cause of this disease is unknown. It may affect any system in the body; it however commonly involves the lymph nodes. There is no known treatment however chemotherapy, radiation and steroids have been tried. Involvement of the liver is very rare; we have however seen two cases in two years. Our first case was a 62-year-old woman who was HIV negative but had advanced cirrhosis of unknown aetiology. We think the cirrhosis may have contributed to her depressed immunity however there is currently no link between immune status and sinus histiocytosis as the condition is rare. The more cases reported the more we are likely to understand this condition. This can only be achieved by improving awareness. In both cases we tried steroids with poor outcomes. Even though it is a self- limiting, the presence of other secondary diseases may have contributed to poor outcome in these patients. There is currently no known link between sinus histiocytosis and HIV and because there have only been 6 cases of liver involvement with this being the seventh, it is not known what the link between the liver disease and this condition is. Our patient was a heterosexual who wasn't married with no history of IV drug use. It is unclear whether sinus histiocytosis contributed to these patient's outcome as they both had life threatening diseases.

Conclusion

Sinus histiocytosis even though it is rare, clinicians should be aware of it and look for it as this may help us discover a cure.

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PEER REVIEW

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CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

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None

PATIENT CONSENT

The authors, *Kgomo MK, Elnagar AA, Mashoshoe KS, Thomas P, Ngwata P, Bida M*, declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).
3. This submission is compliant with the requirements of local research ethics committees.

Figure 1: CT scan abdomen showing splenic abscess

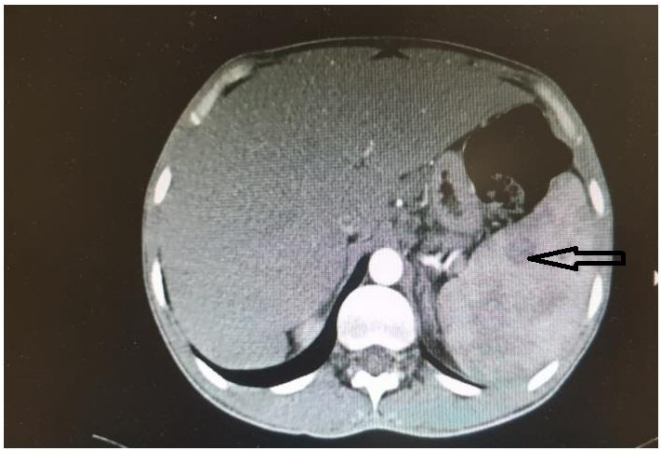

Figure: 2 Histology slide, sinus histiocytosis