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ORIGINAL ARTICLE

Partial splenectomy in sickle cell disease☆

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KEYWORDS
Sickle cell disease;
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Abstract
Introduction: Total splenectomy in sickle cell disease is related to a high risk of fulminant sepsis and increased incidence of other events, which have not been reported in patients with partial splenectomy. In this study we examined the patients with sickle cell disease and partial splenectomy and compared the clinical and laboratory results with non-splenectomised patients.
Materials and methods: We studied 54 patients with sickle cell disease who underwent partial splenectomy in childhood from 1986 until 2011 at the Institute of Hematology and Immunology. They were compared with 54 non-splenectomised patients selected by random sampling with similar characteristics.
Results: Partial splenectomy was performed at a mean age of 4.1 years, with a higher frequency in homozygous haemoglobin S (70.4%), and the most common cause was recurrent splenic sequestration crisis. The most common postoperative complications were fever of unknown origin (14.8%) and acute chest syndrome (11.1%). After splenectomy there was a significant increase in leukocytes, neutrophils, and platelets, the latter two parameters remained significantly elevated when compared with non-splenectomised patients. There was no difference in the incidence of clinical events, except hepatic sequestration, which was more common in splenectomised patients.
Conclusion: Partial splenectomy was a safe procedure in patients with sickle cell disease. There were no differences in the clinical picture in children splenectomised and non-splenectomised except the greater frequency of hepatic sequestration crisis in the first group.

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PALABRAS CLAVE
Drepanocitosis;
Esplenectomía parcial;
Esplenectomía total

Esplenectomía parcial en pacientes con drepanocitosis

Resumen
Introducción: La esplenectomía total en la drepanocitosis se relaciona con riesgo de infecciones sobreagudas y con aumento de la incidencia de otros eventos, lo que no se ha comunicado en pacientes con esplenectomía parcial. En este estudio se caracterizó a los pacientes con drepanocitosis y esplenectomía parcial, y se comparó el comportamiento clínico y de laboratorio con los pacientes no esplenectomizados.
Materials y métodos: Se estudió a 54 pacientes con drepanocitosis sometidos a esplenectomía parcial durante la edad pediátrica, desde 1986 hasta el año 2011, en el Instituto de Hematología e Inmunología. Se compararon con 54 pacientes no esplenectomizados seleccionados por muestreo aleatorio con características similares.
Resultados: La esplenectomía parcial se realizó a una edad media de 4,1 años, con una frecuencia mayor en la anaemia drepanocítica (70,4%) y su causa más común fue la crisis de secuestro esplénico recurrente. Las complicaciones posoperatorias más frecuentes fueron: fiebre de origen desconocido (14,8%) y síndrome torácico agudo (11,1%). Después de la esplenectomía, aumentaron significativamente los leucocitos, neutrófilos y plaquetas; estos 2 últimos parámetros se mantuvieron elevados de manera significativa cuando se compararon con los pacientes no esplenectomizados. No hubo diferencias en la incidencia de los eventos clínicos, excepto el secuestro hepático, que fue más frecuente en los esplenectomizados.
Conclusión: La esplenectomía parcial constituyó un proceder seguro en los pacientes con drepanocitosis. No hubo diferencias en el cuadro clínico entre los niños esplenectomizados y los no esplenectomizados, salvo la mayor frecuencia de crisis de secuestro hepático en los primeros.

Introduction

Sickle cell disease is a multisystemic condition, and splenic complications are among its most prominent manifestations.[1,2] Spleen enlargement is common in the first decade of life, but vascular occlusive events and recurrent splenic infarctions lead to autosplenectomy. However, splenomegaly persists in some patients to later ages, and other patients require splenectomy for various reasons, such as recurrent splenic sequestration crises (SSCs), hypersplenism, massive splenic infarction, and splenic abscess.[2,3]

The most common indication for splenectomy in sickle cell disease is SSC.[3,4] Total splenectomy is the classical treatment for SSC in children older than 4 years, but the procedure often carries a high risk of fulminant septicaemia, which may be fatal.[5] The risk is greater in younger children, in whom the preferred approach is a transfusion regime until at least 2 years of age. Aiming to avoid or reduce the adverse effects of total splenectomy, the Institute of Haematology and Inmunology (IHI) started to perform partial splenectomies in 1986, with excellent results. The remaining spleen remains functional, at least partially, for a period of time that has yet to be determined, and SSCs do not recur, which supports the indication of partial splenectomy in patients with sickle cell disease.[2,4–6]

For a long time, total splenectomy was only associated to increased risk of overwhelming infections.[7] Recent publications have described an increased incidence of vaso-occlusive pain crises (VOPCs), acute chess syndrome (ACS), and stroke following total splenectomy.[8,9] Today it is also associated with thrombotic complications, such as deep-venous thrombosis, especially portal venous thrombosis, and with pulmonary hypertension (PHT).[10–12] These and other complications have not been reported in patients with partial splenectomy.[2,4–6]

Due to all the risks currently known to be associated with total splenectomy, there is a growing interest in partial splenectomy.[13] The aim of this study was to characterise the clinical and laboratory features of patients with sickle cell disease that had undergone partial splenectomy, and to compare them with those of non-splenectomised patients.

Materials and methods

We performed an observational, retrospective, comparative study in patients with sickle cell disease who received ongoing care at the haemoglobinopathies clinic of the IHI between September 2010 and December 2011. The source population consisted of patients with sickle cell disease receiving care at the paediatric and adult departments of the IHI. From it we selected a group of patients aged 1–18 years who had undergone a partial splenectomy. We excluded patients who did not want to participate or whose parents or guardians did not give consent, and patients whose records did not include any postoperative clinical data.