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EXPERIENCE IN THE EVALUATION OF CHILDREN WITH HEPATOSPLENOMEGALY AT A TEACHING AMBULATORY, SÃO PAULO, BRAZIL

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SUMMARY

Objectives: Describe cases of children with hepatosplenomegaly (HS) attended at the General Pediatric Teaching Ambulatory (AGER) of Instituto da Criança, São Paulo, identifying the main causes, evolution, necessity for hospitalization and/or referral to specialists.

Methodology: Retrospective analysis of the records of children presenting HS on admission at AGER from September 1, 1993 to August 31, 1996.

Results: Of the 89 children included (age range, 1 to 148 months; median, 24 months), 64 (72%) were referred from other services for HS investigation. Most common presenting complaints were: fever - 39 (44%); pallor - 26 (29%); weight loss - 21 (24%) and jaundice - 14 (16%). Main alterations noticed on physical examination were: pallor - 47 (53%) and short stature - 17 (19%). Anemia was diagnosed in 70 children (79%); 35 children (39%) had infections; 7 (8%) metabolic disorders and 5 (6%) neoplastic disorders. The most frequent infections were of the urinary tract - 9 (10%) and hepatitis A - 6 (7%).Thirty six children (40%) were referred to specialists, 17 of which were already diagnosed.

Conclusions: Most of the children with HS present deficiency anemia associated with infections which the general pediatrician is able to diagnose. Persistence of unexplained HS for more than 2 months, especially when there is substantial volume enlargement or alteration in the organs consistency, is an indication for referral to specialists.

KEYWORDS: Hepatomegaly; Splenomegaly; Infectious diseases; Children.

INTRODUCTION

As there are numerous causes that can lead to the enlargement of the liver and/or spleen, it is often difficult for the pediatrician to adequately realize the investigation of children presenting this condition on physical examination [2,3,5,6,17,20,21,26,27,29,31]. There are few articles in the literature reporting the experience of general pediatricians in the approach to children with hepatomegaly and/or splenomegaly (HS) [18,25,30]. Most frequently, reports by specialized services consist of cases submitted to previous selective analysis [1,4,7-12,14-16,19,22,24,27-32]. Thus, it was decided to perform a case analysis of those children with hepatosplenomegaly, attended at the General Pediatric Teaching Ambulatory (AGER) of Instituto da Criança, Hospital das Clínicas of University of São Paulo, Brazil [HCFMUSP], with the following objectives: identify the main causes of the hepatosplenomegaly; evaluate the case evolution and investigate the need for referral to specialists and/or hospitalization.

AGER is a pediatric teaching ambulatory, in a university hospital, to which children with pathologies not resolved at the primary services are referred.

CASES AND METHODS

All AGER records during the period from September 1, 1993 to August 31, 1996, of children with liver and/or spleen enlargement on the first appointment were analyzed. During this three-year period, 106 children presented with hepatosplenomegaly. Hepatomegaly was defined as when the liver edge was palpated more than 3 cm below the right costal margin (RCM) at the midclavicular line, for infants aged under 24 months, or over 2 cm for older children. The spleen was considered enlarged when palpable more than 2 cm below the left costal margin (LCM) [2,3,5,6,18,21,29,31]. To assess case evolution, the authors stipulated a minimum follow-up of two months. From the 106 children with hepatomegaly and/or splenomegaly, 17 were excluded for lack of this follow-up.

The analysis of the remaining 89 children included the following items: sex, age, geographical origin, complaints, nutritional

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condition, signs and symptoms associated with HS, laboratory analyses performed, referral to specialists and/or hospitalization, presence and duration for disease resolution. As far as the laboratory investigation was concerned, examinations were requested according to the clinical history, priority being given to the most likely diagnosis. The only laboratory test requested for all children was blood count. No serologic tests were requested for children showing no changes in the initial tests (blood count and liver function), nor for those without contact with hepatitis and that were anicteric, or for those that had spontaneous resolution of the disorder. Urine culture was requested for all children under two years of age and for those over two which showed previous urinary tract infections or alterations in the urinalysis. Fecal examination was requested for children with eosinophilia in the blood count and for those resident in a schistosomiasis endemic zone. Toxocariasis research was performed in those patients with an eosinophilia count higher than 1,000/mm³.

Data were recorded in a previously tested form, filled and analyzed on a microcomputer, using the program EPI INFO (Version 5.01 - Centers for Disease Control. Atlanta, Georgia, USA, 1990).

RESULTS

Of the total 89 children, 42 (47.2%) were female and 47 (52.8%) male. Age ranged from 1 to 148 months (median, 24 months), on admission, most of the children were aged under 5 years (82.2%).

Sixty four children (71.9%) were referred by other services for hepatosplenomegaly investigation and/or follow-up and 25 (28.1%) presented liver and/or spleen enlargement detected during the physical examination, on admission, of children registered for other disorders. The main disorders reported on admission are listed in Table 1. Recent otitis and respiratory problems were given special consideration when they occurred within 15 days prior to admission. Besides the disorders reported in Table 1, 4 children showed positive epidemiology for AIDS (parental history of drug use or tattoos).

Most of the children (60%) resided in the city of São Paulo, but, as AGER is a referral service, 17 (19.1%) lived in other cities within the state of São Paulo and 18 (20.2%) were from other states.

On physical examination, only two (2.2%) did not present palpable liver. In the remaining, the size of the liver ranged from 2.5 to 13 cm. In 54 children (60.7%) the liver was palpable as far as 4 cm from the RCM in the midclavicular line, and in 33 (37.1%) palpable beyond 4.5 cm from RCM . Four children showed liver with altered consistency.

Sixty five children (73.3%) presented a palpable spleen, of which 26 (29.2%) had spleen tip (≤ 2 cm) and 39 (43.8%) splenomegaly. In 21 children (23.6%) the spleen was palpable between 2.5 and 4 cm and in 18 (20.2%) beyond 4.5 cm from LCM. Nine children showed predominant spleen enlargement; 3 hardened and one painful spleen. Only two children showed isolated splenomegaly.

Besides liver and spleen enlargement, the most frequent findings during the first physical examination, can be seen in Table 1. Approximately 20% of the children showed short stature and 15% had weight below the percentile 3, according to NCHS criteria.

TABLE 1
Distribution of the 89 children with hepatosplenomegaly according to complaints present at admission and physical examination

COMPLAINTS *	n	%
FEVER	39	43.8
PALLOR	26	29.2
WEIGHT LOSS	21	23.6
JAUNDICE	14	15.7
ABDOMINAL PAIN	13	14.6
LYMPHADENOPATHY	9	10.1
VOMITING	8	9.0
URINARY**	8	9.0
DIARRHEA	7	7.9
RECENT OTITIS	7	7.9
RESPIRATORY ***	6	6.7
PURPURA	4	4.5
ALTERATIONS ON PHYSICAL EXAMINATION *		
PALLOR	47	52.8
SHORT STATURE	17	19.1
LOW WEIGHT (< p 3 %)	13	14.6
CRANIOFACIES ALTERATION	10	11.2
COMPROMISED GENERAL CONDITION	10	11.2
JAUNDICE	9	10.1
LYMPHADENOPATHY	6	6.7
FEVER	5	5.6

* Several children presented more than one complaint or alteration on physical examination.
** Dysuria, polaciuria, hematuria or alteration in the urine color and history of urinary infection in the month prior to admission.
*** Tonsillitis, pharingitis, cough, bronchitis, dyspnea and/or pneumonia

Anemia (according to WHO criteria) was registered in 70 children (78.7%), this proportion being higher than that mentioned in anamnesis and the case observed in the physical examination (Table 1). Nine children (10.1%) had hemoglobin values less than or equal to 7.0 g/dL; 24 children (27.0%) showed hemoglobin concentration between 7.1 and 9.0 g/dL, and 37 (47.6%) Hb between 9.1 and 11.5g/dL.

The majority of children (71.9%) presented iron deficiency anemia and/or anemia associated to infectious and metabolic disorders. Seventeen children presented eosinophilia in the blood count, and helminthiasis was detected in 12 cases. Seven children showed thrombocytopenia; out of which 3 had kala azar, two leukemia, one idiopathic thrombocytopenic purpura and one histiocytosis X. Investigation for hemolytic disorders was performed in 17 children and confirmed in six.

Abdominal ultrasonography was performed in 66 children (74.2%): 17 (25.6%) were normal and 14 (21.2%) showed problems, while in 35 (53.0%) the ultrasonography served only to confirm non specific hepatomegaly and/or splenomegaly.

Aminotransferases, serologic tests and urinalysis were carried out in 59 children (66.3%). Elevation in the aminotransferases was observed in 24 out of 59 examinations (40.7%). Urinalysis revealed alterations in 10 out of 59 cases (16.9%). Serologic tests for infectious disease research have been requested for 59 children, however, most of these have not yet undergone all the examinations necessary to exclude the probability of infection [1,7-9,13,19,21-25,27,29-32].

Protein electrophoresis was performed in 41 children (45.6%), indicating hypergammaglobulinemia (over 1.6 g/dL) in 13. Hypoalbuminemia (below 3.5 g/dL) was observed in 7 children, all of which with weight below percentile 3.

Urine culture was performed for 34 children, indicating 9 cases of urinary tract infection.

Stool examination of 29 children revealed: schistosomiasis mansoni - 3 cases; giardiasis - 7 cases; ascariasis - 5 cases; trichocephaliasis - 3 cases; amebiasis and ankilostomiasis, one case each.

Bilirubin rate was determined in 25 children (28.1%): 9 presented high levels corresponding to 5 infectious diseases; 3 sickle cell anemia and one extrahepatic biliary atresia.

Bone marrow aspiration was indicated for 9 children, indicating: two cases of kala azar, two Gaucher's disease, one acute lymphocytic leukemia and one suspected of leukemia (84% lymphocytes). Three other children had been admitted with a confirmed diagnosis by bone marrow aspiration: two cases of kala azar and one idiopathic thrombocytopenic purpura.

TABLE 2
Distribution of diagnoses made in children with hepatosplenomegaly (*)

DIAGNOSIS	NUMBER
ANEMIA	70
INFECTION	42
METABOLIC	7
NEOPLASTIC	5
OTHERS:	
CONGESTIVE	3
"IDIOPATHIC" NEONATAL HEPATITIS	2
CHRONIC HEPATITIS	2
EXTRAHEPATIC BILIARY ATRESIA	1

(*) Excluding malnutrition

Nine children (10.1%) were referred to specialists for histological examinations. From the 8 histological examinations done, 6 were liver biopsies which confirmed the following diagnosis: Niemann-Pick disease; extrahepatic biliary atresia; primary sclerosing cholangitis; glycogen storage disease type 1; hemangioendothelioma; and chronic hepatobiliary disease. A skin biopsy was carried out enabling the diagnosis of histiocytosis X and a bone marrow biopsy confirmed the possibility of acute lymphocytic leukemia in the child whose bone marrow aspiration revealed lymphocytosis of 84%.

Other examinations were requested such as: ophthalmoscopy (11); cranium ultrasonography (5); chest roentgenogram (10); skeletal roentgenogram (4); screening for inborn errors of metabolism (3); rate of alpha-1 antitrypsin (3); serum acid phosphatase levels (3). Only one ophthalmoscopy showed alteration (cherry-red spot on the macula) and a test for inborn errors of metabolism resulted positive for mucopolysaccharidosis.

Serum isohemagglutinins were requested for 2 children with suggested toxocariasis and these were elevated in both.

Several diagnoses associated with hepatosplenomegaly were identified (Table 2). A total of 42 infectious pathologies were diagnosed in 35 children (Table 3); seven children had metabolic diseases (excluding malnutrition) and five neoplastic disorders.

Excluding iron deficiency anemia and malnutrition, 11 children showed more than one cause associated with hepatosplenomegaly, as can be seen in Table 4.

TABLE 3
Distribution of the 42 infectious causes associated to hepatomegaly and/or splenomegaly, according to the number of diagnoses

INFECTIOUS CAUSES	NUMBER OF DIAGNOSES
URINARY TRACT INFECTION	9
HEPATITIS A	6
VISCERAL LEISHMANIASIS (KALA AZAR)	4
TOXOCARIASIS*	4
HEPATITIS B	3
AIDS	3
SCHISTOSOMIASIS	3
TOXOPLASMOSIS	2
RUBELLA	2
SYPHILIS	2
PNEUMONIA	2
LEPTOSPIROSIS	1
WHOOPING COUGH	1

Note: * one case without serology

TABLE 4
List of children with more than one diagnosis related to hepatosplenomegaly *

CASE	DIAGNOSIS
1	URINARY TRACT INFECTION + PNEUMONIA
14	HEPATITIS A + SCHISTOSOMIASIS
41	TOXOCARIASIS + PORTAL VEIN THROMBOSIS
43	HEPATITIS A + HEMOLYTIC ANEMIA
45	CYTOMEGALIC VIRUS + SC HEMOGLOBINOPATHY
47	WHOOPING COUGH + URINARY TRACT INFECTION
69	CONGENITAL LUES + URINARY TRACT INFECTION
76	HEPATITIS A + SICKLE CELL ANEMIA
77	LEPTOSPIROSIS + TOXOCARIASIS
81	HEPATITIS A + TOXOCARIASIS + SCHISTOSOMIASIS
83	URINARY TRACT INFECTION + TOXOCARIASIS + PNEUMONIA

* Excluding iron deficiency anemia and malnutrition.

EVOLUTION AND REFERRALS TO SPECIALISTS

Approximately half of the children (56.7%) showed hepatosplenomegaly resolution from one to twelve months (median, 3.5 months).

During the follow-up, 36 children (40.5%) were referred to specialists, 17 (47.2%) already with diagnosis established. Fifteen were referred to the hepatology group, seven (46.7%) of which already diagnosed: two cases of hepatitis by virus B; two with Gaucher's disease; one with Niemann Pick's disease; one with portal vein thrombosis and one case of mucopolysaccharidosis.

Eleven children were referred to the oncology group: 9 for bone marrow aspiration; one with hemangioendothelioma and another child because abdominal ultrasonography detected a tumor in the right suprarenal region, diagnosed later as Wilms' tumor.

Six children with hemolytic anemia were referred to the hematology group five of which with the diagnosis established at AGER: sickle cell anemia (3); SC hemoglobinopathy (1) and thalassemia (1). Half of the children with hemolytic anemia also presented associated infectious processes.

Three children were referred to the immunology group, after acquired immunodeficiency syndrome was confirmed. One child was referred to the nephrology group for follow-up of rickets resistant to vitamin D, and another to the gastroenterology group for follow-up of cystic fibrosis.

DISCUSSION

Regarding similar studies in the literature, only three were found [18,25,30] and these involved a low number of hospitalized children (n = 57 [18], 63 [25] and 18 [30]). Furthermore, a significant comparison was made difficult by the fact that the research used different methods and took place in other countries and times with consequent differences in the incidence of endemic diseases such as kala azar and schistosomiasis. Nevertheless, it is worth noting that HS was presented predominantly in children under 5 years of age and with a low incidence of isolated splenomegaly.

Due to the large number of conditions leading to liver and/or spleen enlargement, it is important that the pediatrician is able to identify, beforehand, those diseases which are potentially treatable, and to differentiate the benign and self-limited diseases from those with a worse prognosis [5-7,10,11,13,16,20,26,27,31,32]. Clinical evaluation is of extreme importance, and age range, presence of other complaints in the clinical history and epidemiological data should be evaluated. Alterations found during the physical examination should be considered, especially compromised general appearance and condition, magnitude of liver and/or spleen enlargement and consistency alterations. Another important factor is persistency of hepatomegaly and/or splenomegaly for more than 2 months [21,22,25-32].

Liver function tests present low specificity for the diagnosis, since they are affected by several factors such as hemolysis, nutritional condition, osseous and renal problems, among others. The predictive value of positive results for the presence of hepatic disease varies from 56% to 88%, and the predictive value of negative results is higher (87% to 97%) [17]. Therefore, they should be carried out more in order to investigate the presence or absence of liver disease than as an aid to etiologic diagnosis or to monitor the severity, course, and response to treatment of hepatic diseases and also to detect the hepatotoxic effects of medicines and drugs [5,17-22,27,29-32]. Hypergammaglobulinemia helped in the identification of chronic inflammatory processes [1,9,11,17,19-22,26,27,32], being high in HIV cases, kala azar, toxocariasis and chronic hepatitis.

Abdominal ultrasonography was in effect only helpful for the diagnosis of congestive, neoplastic and metabolic problems [2,5,6,12,14-16,21,22,25-32].

Bone marrow aspiration is indicated if the child presents pancytopenia or significant hepatosplenomegaly [2,5,6,9,14-16,20,26,27,29-32]. This examination is essential for the diagnosis of kala azar, leukemia and Gaucher's disease.

Blood count was the only examination carried out for all children, allowing the detection of anemia and suggesting the investigative course to be taken. Alterations in the white and megakaryocytic series also helped in the diagnosis of infectious, hematological and neoplastic problems [5,6,17-19,26,27,29-32].

Anemia has been the most frequent diagnosis associated with HS. Although only one third of the parents complained that the child presented pallor, it has been observed in half of the children (Table 1) and the blood count revealed anemia in almost 80%. Six children presented hemolytic anemia, 5 of which had a history of jaundice.

In 29 children (32.5%) who had hipochromic and microcytic anemia with low reticulocytis count, iron deficiency anemia was

diagnosed as the possible cause of hepatosplenomegaly; 20 children (22.5%) presented mild liver and/or spleen enlargement improving in less than two months. Other hematological tests and investigation for infectious diseases were carried out for 9 children (10%) who had hemoglobin rates between 4.3 and 10.4 g/dL and a quite slow resolution of HS. In all these children iron deficiency anemia was confirmed. The HS only disappeared after 5 to 12 months, after the anemia was corrected with iron therapy. It is not possible to exclude the hypothesis that in this group of 29 children, the HS besides being associated to the iron deficiency anemia, was also associated with infectious processes. Eleven children of these (37.9%) had recent antecedents of acute infections (UTI, otitis, pneumonia and diarrhea) [6-9,11,18-25,27,29-32].

Urinary tract infection (UTI) was the most frequent infection associated with hepatosplenomegaly and children presenting a liver ≤ 5 cm and spleen ≤ 3 (Table 3). In all cases HS subsided in less than two months. It is very important to investigate urinary tract infection in children under 2 years of age, because this infection occurs with no specific signals or symptoms in infants [6,18,2125,27,29-32].

During the course of viral diseases, the children presented a small enlargement of the liver and/or spleen of less than 4 cm, and the hepatic consistency was normal. Another interesting point is that the majority of the children with hepatitis were anicteric. Of the nine children with hepatitis A or B, only two had jaundice and only three presented an increase of aminotransferases on the admission date [19,21-23,27,32].

The great variety of infectious agents associated with hepatosplenomegaly renders specific research for all possible agents unfeasible in practical terms [5-9,19,21,24,27,29-32]. In this study, specific diagnostic tests were only requested for those cases where persistent clinical alterations were detected, or when there was positive epidemiology for the suspected infection or alteration in the initial examination. It should be pointed out, however, that viral hepatitis occurs without jaundice in most infants and young children, and that in 30 to 50% of the individuals infected with the hepatitis A and B viruses, it is not possible to identify the source of infection[19,23,27,32]. In viral etiology hepatitis aminotransferases persists high for 2 to 3 weeks and can normalize afterwards, therefore, even when the rates of aminotransferases are normal, it is not possible to exclude viral hepatitis. The same can be stated about other agents responsible for hepatitis. It should be stressed that hepatitis caused by viruses A, B, C, D and E cannot be clinically distinguished from infections caused by other agents[6-9,11,13,19,21,22,24,27,29-32]. The authors strongly recommend research into the possibility of infection by HIV in children with HS and positive epidemiology for AIDS, as this clinical manifestation is frequently found in children with AIDS. CARVALHO et al. (1997)[7] state that of 103 children with AIDS, 84.5% presented hepatomegaly and 64% splenomegaly. It is important to remember that specific treatment is effective in altering the prognosis of HIV-1 associated disease[11].

The most frequent infections by non viral agents were toxocariasis, kala azar and schistosomiasis.

Hepatomegaly is a common finding in visceral larva migrans or toxocariasis infection [9,19]. In this study, toxocariasis diagnosis was confirmed via specific serologic tests (ELISA) in 3 children. In another child, it was not possible to collect serum for confirmation of toxocariasis; nevertheless, this child probably had toxocariasis too, as results confirmed positive epidemiological data, anemia (Hb = 6.7 g/dL), leucocytosis and marked eosinophilia (29%), associated with an increase of isohemagglutinins.

Visceral leishmaniasis (kala azar) is common in northeastern Brazil [9] and all four children with kala azar were from the state of Bahia. They presented malnutrition and compromised general condition, with a history of fever and weight loss. On physical examination, they showed marked pallor and massive splenomegaly, between 6 and 13 cm. Blood count revealed pancytopenia. Two of these children were hospitalized on the admission day for investigation, because this clinical picture is indistinguishable from other serious diseases, such as leukemia, lymphoma, prolonged Salmonella bacteremia, acute schistosomiasis, typhoid fever, miliary tuberculosis and other infections. A definitive diagnosis of kala azar depends on the demonstration of *Leishmania* in tissue or isolation of the organism in culture. Bone marrow aspiration is the safest diagnostic procedure for confirming this [9].

Two of the three children with schistosomiasis resided in the state of Minas Gerais (an endemic area) [9], and one had traveled to that state a month before the initial symptoms appeared. Besides clinical and epidemiological data, all these children had eosinophilia in blood count.

In the cases of kala azar, schistosomiasis, toxocariasis and leptospirosis, very slow improvement of hepatosplenomegaly was observed, in all cases the HS persisted for over 5 months.

Although iron deficiency anemia and infectious processes were more commonly associated with HS it should be pointed out that 19% of the children had short stature and 15%, low weight. Malnutrition can also lead to hepatomegaly [5,6,30,31], and in this study it has been associated with infection, neoplastic and other metabolic processes, and also to a significant presence of multiple causes associated to HS (Table 4).

Metabolic diseases (excluding malnutrition) constituted the third group of pathologies associated to HS. It is known that some metabolic diseases appear during the neonatal period, while others become apparent at a later date. Clinical manifestations that suggested the possibility of metabolic disease were: neonatal cholestasis; significant hepatosplenomegaly; fulminant hepatic failure; bleeding (coagulopathy); failure to thrive; short stature; dysmorphic features; developmental delay/psychomotor retardation; hypotonia and rickets. Further clues are provided by a family history of similar diseases [5,6,10,14,18,19,20,21,25,27,29-32].

The pediatrician should be alert to the possibility of metabolic diseases in early childhood, to direct therapeutic measures to avoid accumulation of toxic substances in the body and indicate genetic counseling, when necessary [27]. However, some of these diseases may be apparent only with hepatomegaly and/or splenomegaly at the

beginning of the symptoms, such as alpha-1 antitrypsin deficiency, Gaucher's disease and Wilson's disease[4,5,10,20,21,27,29].

In this study, seven children presented metabolic disorders; all had height and weight below percentile 10. There were two cases of Gaucher's disease in which the children presented enlarged spleen and hardened consistency. Splenomegaly is usually the first clinical sign of Gaucher's disease. Hepatosplenomegaly can lead to hypersplenism, causing anemia and thrombocytopenia, and this diagnosis should be suspected in children with persistent hepatosplenomegaly and unexplained anemia[5,6,14,20,21,27,31]. In this study, serum acid phosphatase levels were greatly elevated and roentgenograms helped in identifying osseous complications associated with Gaucher's disease. In both, the diagnosis was confirmed by identification of Gaucher's cells in bone marrow and in one case, by the measurement of glucosylceramide ß-glucosidase activity in leukocytes. There was a case of Niemann-Pick's disease in a child with massive splenomegaly and failure to thrive that had a typical cherry red spot in macula. The diagnosis was confirmed by bone marrow aspiration[5,6,15]. There was one case of glycogen storage disease, confirmed by liver biopsy, and the other metabolic diseases were cystic fibrosis, mucopolysaccharidosis, and vitamin D resistant rickets.

Neoplastic diseases were diagnosed in 5 children. Although the rate of incidence of malignant disease in children is low, the impact of cancer makes it imperative that the pediatrician maintains an alert position regarding the possibility of cancer. It is important to emphasize that an early diagnosis improves the outcome[26]. The clinical data suggesting severe processes was: high fever, marked pallor with normocytic anemia, and compromised general condition.

Hepatic tumors are quite rare in childhood[5,6,18,25-27,30,31]. In the case of hemangioendothelioma, the main clinical finding was enlarged liver (13 cm). Hepatosplenomegaly is not common in cases of Wilms' tumor, these are often asymptomatic and present as an abdominal mass often discovered on routine examination or by parents, even though the child does not generally appear to be ill[26,27]. In this case, it is possible that the hepatosplenomegaly had been associated with some non-identified infection.

Histiocytosis is a rare disorder of histiocytic proliferation, with variable and often unpredictable behavior. The child with hystiocytosis presented marked pallor, fever, anemia and thrombocytopenia, besides hepatosplenomegaly, dermatitis and a poor general condition that suggested this diagnosis[16].

Regarding other etiologies (Table 2), congestive causes were not very frequent in this study. As in other studies[18,25,30], abdominal ultrasound examinations proved of great value for the diagnosis of congestive causes[5,21]. Besides the long evolution (> 6 months) one of the physical examination findings that most suggested chronic hepatitis was the alteration of hepatic consistency. Among the laboratory data, hypoalbuminemia and hypergammaglobulinemia stand out.

As AGER is a referral service, children aged under 30 days are rarely registered, and as a consequence, some pathologies which arise during the neonatal period were not commonly found in the cases studied.

Screening for congenital infections is crucial in children that show HS in the neonatal period, especially when the newborn presents other clinical signs[5,6,12,19,23,27,29,32]. In the present study, 8 from the 42 infection processes confirmed (19%) were congenital: three cases of AIDS, two cases of syphilis, one of hepatitis B, one of rubella and one of toxoplasmosis.

Extrahepatic biliary atresia is the most common structural abnormality resulting in neonatal jaundice and the pediatrician should be alert to this and quickly refer suspected cases to a specialist because, if not treated, this disease develops with progressive obliteration (obstruction) of the extra hepatic biliary system, even after surgical correction[27-29,31].

In this study, extrahepatic biliary atresia was diagnosed in a two-month old child, which, in spite of surgical correction, developed several outbreaks of cholangitis.

CONCLUSIONS

This study showed that we can classify children with HS into 2 groups: children with a small increase in liver and/or spleen size, without signs and symptoms of serious diseases and in which the problem can be solved within 2 months; and children with substantial volume enlargement or alterations in the liver and/or spleen consistency, with other signs and symptoms suggesting chronic disorders. In the first and largest group, the most frequent problems were anemia and infectious diseases and the general pediatrician can reach a diagnosis with few examinations, but in the latter group, it is necessary to refer the child to a specialist for a comprehensive investigation.

RESUMO

Experiência de um ambulatório de ensino na avaliação de crianças com hepatoesplenomegalia, São Paulo, Brasil

Objetivos: Descrever os casos de crianças com hepatoesplenomegalia (HE) matriculadas no Ambulatório Geral (AGER) do Instituto da Criança, São Paulo, identificando as principais causas, evolução, necessidade de hospitalização e/ou encaminhamento a especialistas.

Metodologia: Estudo retrospectivo dos prontuários de crianças matriculadas no AGER para investigação de HE de 01/09/93 a 31/08/96.

Resultados: Das 89 crianças incluídas (idade: 1 a 148 meses; mediana, 24 meses), 64 (72%) foram encaminhadas de outros serviços para investigação de HE. As queixas mais freqüentes foram: febre - 39 (44%); palidez - 26 (29%); emagrecimento - 21 (24%) e icterícia - 14 (16%). As principais alterações ao exame físico foram: palidez - 47 (53%) e baixa estatura - 17 (19%). Setenta crianças (79%) apresentavam anemia; 35 (39%) tinham infecções; 7 (8%) doenças metabólicas e 5 (6%) doenças

neoplásicas. Trinta e seis crianças (40%) foram encaminhadas a especialistas.

Conclusões: A maioria das crianças com HE apresenta anemia carencial associada a infecções que o pediatra geral é capaz de diagnosticar. A persistência de HE inexplicada por mais de 2 meses, especialmente quando existe grande aumento e/ou alteração na consistência dos órgãos é indicação para encaminhar ao especialista.

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