CASE REPORT

Secondary asplenia as a complication of severe Meningococcal Sepsis

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SUMMARY

We report the case of an 11-month-old infant who had a medical history of meningococcal septicaemia 6 months prior to this admission and presented to hospital with an episode of fever due to *Streptococcus pneumoniae* bacteraemia. During this second admission, an abdominal ultrasound scan revealed secondary asplenia.

BACKGROUND

Asplenia and infection with *Neisseria meningitidis* are usually perceived to have a cause-and-effect relationship since the lack of the spleen predisposes patients to infection. However, in our case, this relationship is inverted: asplenia is secondary to a meningococcal infection; a complication that has not been described so far. After an exhaustive literature review, no cases showing a causal association between meningococcal sepsis and secondary asplenia development have been found.

CASE PRESENTATION

We report the case of an 11-month-old infant who was admitted to the emergency department (ED) due to a febrile episode lasting 1 hour (39.2°C), vomiting and irritability.

The patient was born by spontaneous vaginal delivery at 37 weeks to a healthy mother after an uncomplicated pregnancy. Birth weight was 2.900 g. The baby's growth and development had been normal. At 2 and 4 months of age, she received Spanish immunisation schedule with pneumococcal conjugate vaccine 13-valent (group B meningococcal vaccine is not included). At 5 months of age, she developed group B *N. meningitidis* septic shock resulting in multiple organ failure. She was admitted to the intensive care unit where she was treated with vasoactive drugs. She also presented severe coagulopathy, compatible with disseminated intravascular coagulation (DIC), which normalised 4 days after admission. The patient developed ischaemic injuries in the limbs. As a consequence of this, the left upper limb was amputated and brain injury with diffuse cortical atrophy, right-sided hemiparesis and focal epilepsy requiring antiepileptic treatment with levetiracetam were observed.

Three months after discharge, she consulted at the ED with a new episode of fever without source, vomiting and irritability. On physical examination, only pharyngeal hyperaemia was observed. The laboratory analysis revealed leucocytosis (22 200/mm³),

C-reactive protein (7.33 mg/dL) and procalcitonin (1.74 ng/mL). The urinalysis and chest radiography showed normal results.

INVESTIGATIONS

The patient was admitted with the diagnosis of fever without source, and antibiotic therapy with cefotaxime was started awaiting blood culture results. Owing to continuous fever, worrying general condition and considering the previous disease, a cranial CT scan was carried out showing no changes when compared to previous images and, after this a lumbar puncture was performed showing normal results. Blood cultures grew *S. pneumoniae* serotype 10A.

Reviewing previous imaging for the patient, we found that during the meningococcal sepsis an abdominal ultrasound (US) scan revealed a perisplenic haematoma measuring 6.8×1.7×4 cm (figure 1), which unfortunately was not controlled with subsequent US scans, because it was assumed a spontaneous reabsorption after the episode of disseminated intravascular coagulation. A new abdominal US scan revealed a 1 cm splenic remnant (figure 2). This was interpreted as asplenia secondary to a spleen necrosis in relation to meningococcal disease. In peripheral blood smear, there was red blood cell containing a Howell-Jolly body, absent in peripheral blood smear at the first admission; nor showed sickle cell.

The immunological profile of the patient was assessed at 2 years of age and no abnormal results were found. The concentrations of immunoglobulin classes (IgG 798 mg/dL, IgM 127 mg/dL, IgA 46 mg/dL) and subclasses were normal. Analysis of the complement pathway showed normal

Figure 1 Abdominal ultrasound scan revealed a perisplenic haematoma measuring 6.8×1.7×4 cm during the first admission.

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Figure 2 Abdominal ultrasound scan revealed a 1 cm splenic remnant.

concentrations of C3 and C4, and normal AP 50 and CH 50 test. Total lymphocyte and T, B and Natural Killer cell counts were normal. Neutrophil oxidative burst test was normal. Antibody responses to tetanus toxoid and polysaccharide pneumococcal vaccine (pneumovax) were also normal, and isohaemagglutinins were present.

DIFFERENTIAL DIAGNOSIS

Sickle cell anaemia was ruled out because of the lack of sickle cell in the peripheral blood smear. Chronic granulomatous disease was excluded through neutrophil oxidative burst test with normal dihydrorhodamine 123 assay, so a normal neutrophil function is assumed and glucose-6-phosphate-dehydrogenase deficiency was also excluded. Specific diagnostic tests for hereditary spherocytosis were not performed because spherocytes were never found on the peripheral blood smear.

TREATMENT

Antibiotic prophylaxis with amoxicillin was prescribed and a 23-valent antipneumococcal vaccine from 2 years of age was due to be administered.

OUTCOME AND FOLLOW-UP

The patient improved and was discharged. After 2 years of follow-up, no other new invasive infections have occurred.

DISCUSSION

Neisseria meningitidis is a Gram-negative diplococcus pathogen that colonises the human nasopharynx. However, it can cause rapidly progressive disease and death. Fortunately, in this case, it was not fatal and only neurological deficits and asplenia, which may predispose the patient to serious infections, was observed.

Although asplenia/hyposplenia can be caused under different conditions such as haemoglobinopathies, vasculitis, chronic graft-versus-host disease, etc, there are no reported cases so far in which a direct relationship between meningococcal disease and secondary asplenia is described. Retrospectively reviewing the progress of the child, it is considered as the only clear precedent that might justify the resorption of the spleen. Regarding the pathogenic mechanism, there are several factors that could have caused it: the inflammatory reaction to the infection that caused disseminated intravascular coagulation (DIC), necrosis and/or splenic rupture.

First, DIC plays an important role in this case. In an US scan a perisplenic haematoma, which presumably evolved to massive

asymptomatic necrosis, was seen. It is known that DIC can cause bleeding and thrombotic events (due to fibrin deposition in the microcirculation, or more rarely in large vessels, such as the splenic artery or vein).¹ In our case, there are several predisposing haemodynamic factors: systemic hypoperfusion, which increases the formation of thrombin, and liver hypoperfusion, which prevents clearance of procoagulant factors. Although splenic haematoma was demonstrated, the occurrence of splenic vein thrombosis may have also occurred.

Furthermore, even in the absence of thrombosis or haemorrhage, inflammatory mechanisms appearing in sepsis may cause or contribute to spleen necrosis. The case of a 75-year-old woman who suffered from typhoid fever 30 years ago and who had a liver abscess caused by *Salmonella typhi* is described. A massive splenic necrosis was incidentally found. The authors conclude that the splenic alteration could have originated years ago, and that functional splenectomy may have contributed to the maintenance of the state of carrier and the development of the new infection.²

This inflammatory reaction can be so intense that it can even cause *atraumatic splenic rupture*. In the literature, the case of a 13-year-old adolescent who had symptoms of meningococcal infection and abdominal pain has been described.³ During her admission, the patient revealed progressive abdominal distension and a laparoscopy was performed. This examination showed bleeding splenic rupture, which was eventually attributed to the said infection.

In most cases, splenic rupture is symptomatic (sudden abdominal pain with signs of shock, anaemia etc). Nevertheless, some oligo-asymptomatic cases have been described: the case of an 18-year-old woman who developed a subcapsular haematoma in the context of infectious mononucleosis, the case of spontaneous splenic rupture as the presenting form of acute lymphoblastic leukaemia and another case of failure as a complication of an infectious endocarditis.⁴⁻⁶ In addition to the aforementioned mechanisms of oedema and cellular hyperplasia, rupture can be caused by a tear in the fibrous capsule (due to cellular infiltration), bleeding into the infarcted spleen, rupture of a mycotic aneurysm or a splenic abscess.

An abdominal US scan can fail to show small or poorly organised splenic haematomas, with superior sensitivity and specificity of a CT scan. It is, therefore, possible that, in our case, the splenic rupture had occurred before the first US scan was performed.

In a case of asplenia, there is less clearance of bacteria from the circulation. Humoral immunity also weakens. (IgM production is reduced in response to polysaccharides.)⁷ All this increases the risk of rapidly progressive septicæmia and death. Patients are more susceptible to certain microorganisms such as *S. pneumoniae*, *H. influenzae* and *N. meningitidis*. Other pathogens Gram-positive (*S. aureus*, other streptococci) and Gram-negative (*E. coli* and other bacilli, *Salmonella* spp., *Klebsiella* spp., *Pseudomonas* spp. etc) may less commonly be implicated. Moreover, the risk of infection by *Campylobacter jejuni*, *C. cynodegmi*, *Babesia* and *Bordetella holmesii* is also increased. Although the highest risk of infection occurs in the first year after splenectomy, it can remain in the same level for the whole life. For this reason, when such a patient develops fever without source early antibiotic treatment effective against all the above-encapsulated bacteria is recommended after blood and other appropriate cultures are obtained. In addition, complete immunisation and daily antimicrobial prophylaxis against pneumococcal infection at least in the early years of secondary asplenia is also required.⁸

Learning points

- We report the first case described in the literature of asplenia probably secondary to disseminated intravascular coagulation in the context of previous meningococcal sepsis.
- Although it is a rare complication bearing in mind the high risk of severe infections, we think that septic shock should be considered as a predisposing factor to asplenia and that early diagnosis is important in these situations.
- Accurate diagnosis will allow for preventive measures to be taken such as immunisation, antimicrobial prophylaxis and early antibiotic treatment of febrile episodes in order to prevent serious infections and their complications in these patients.

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Contributors LC-C designed and organized the report and wrote the conclusions. LB-C wrote the discussion and chose the US images. MJM-S wrote the clinical case. MIG-T reviewed the report.

Competing interests None declared.

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REFERENCES

- 1 Corne P, Borie F, Gallix B, *et al*. Infarctus splénique révélant une méningococcémie. *Gastroentérol Clin Biol* 2004;28:197. <http://www.em-consulte.com/en/article/99871>
- 2 Collazos González J, García Pérez J, *et al* [Liver abscess caused by *Salmonella typhi* associated with splenic ischemic necrosis and renal vein thrombosis]. *Med Clin (Barc)* 1992;98:595.
- 3 Buist MD, Power RG, Keeley SR, *et al*. Severe meningococcal septicaemia associated with splenic rupture. *Med J Aust* 1991;155:713–14.
- 4 Gorosquieta A, Pérez Equiza E, Gastearena J. [Asymptomatic pathological rupture of the spleen as the presenting form of acute lymphoblastic leukemia]. *Sangre (Barc)* 1996;41:261–2.
- 5 Baibars M, Ohrum P, Alraiyes AH, *et al*. Asymptomatic spleen rupture in patient with endocarditis. *QJM* 2013;106:1047–8.
- 6 Stahlknecht T, Majewski A. [Asymptomatic subcapsular splenic hematoma in infectious mononucleosis (Pfeiffer disease)]. *Rontgenblatter* 1986;39:264–5.
- 7 Di Sabatino A, Carsetti R, Corazza GR. Post-splenectomy and hyposplenic states. *Lancet* 2011;378:86–97.
- 8 Rubin LG, Schaffner W. Clinical practice. Care of the asplenic patient. *N Engl J Med* 2014;371:349–56.

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