Spontaneous rupture of the spleen as a result of infectious mononucleosis in two siblings

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INTRODUCTION

The first confirmed case of splenic rupture complicating infectious mononucleosis was reported by King (1941) and is well recognized as a rare complication. Enlargement of the spleen and intrinsic changes in the splenic parenchyma during the course of the disease make the spleen susceptible to rupture without a notable traumatic insult.

For the first time we report splenic rupture in two siblings with infectious mononucleosis and review the clinical presentation of this potentially life-threatening complication.

DISCUSSION

Spontaneous rupture of the spleen in infectious mononucleosis is rare and has been estimated to be 0.1–0.5% (Lee et al, 1976; Farley et al, 1992). The criteria for establishing splenic rupture in infectious mononucleosis, outlined by Rutkow (1978), are:

- No history of recent trauma
- Haematological and serological evidence of infectious mononucleosis
- Recent clinical symptoms of infectious mononucleosis
- Histology showing changes consistent with infectious mononucleosis.

These two cases fulfill these criteria.

Eighty per cent of patients with infectious mononucleosis are under the age of 25 years (Farley et al, 1992). Abdominal pain is a rare presentation of infectious mononucleosis, but occurs in 90% of cases complicated by splenic rupture (Konvolinka and Wyatt, 1989). Kehr's sign, pain referred to the left shoulder as a result of diaphragmatic irritation caused by the presence of intraperitoneal blood, is present in more than 50% of patients with splenic rupture from whatever cause.

Splenic enlargement has been noted in 50% of patients with infectious mononucleosis and is universally associated with splenic rupture. This reflects the fact that the normal spleen weighs approximately 198 g but the spleen that ruptures often weighs more than 750 g (Konvolinka and Wyatt, 1989).

The mechanism for spontaneous rupture of the spleen in infectious mononucleosis is unclear, but about 2 weeks after the onset of the illness the enlarged spleen shows capsular and trabecular infiltration with normal and atypical lymphocytes. The fibromuscular and vascular structures of the trabeculae may be so diminished that a tear can easily occur (Hyun et al, 1972). The degree of

CASE REPORT 1

A 15-year-old girl presented with acute onset of left shoulder tip pain and left-sided pleuritic pain while lying in bed. She had been unwell for 2 weeks with a sore throat and cough. On examination she had cervical lymphadenopathy and white plaques on the posterior pharyngeal wall but was haemodynamically stable. There was reduced air entry into the left lung base, and tenderness and guarding throughout the abdomen, maximal in the left upper quadrant. Her haemoglobin level was 9.9 g/dl with a white cell count of 14 x 10^9/litre. A Paul Bunnell test was positive. Abdominal ultrasound demonstrated an enlarged spleen with free fluid around the liver and in the pelvis. A diagnosis of splenic rupture with haemoperitoneum was made and she underwent emergency splenectomy following deterioration in her vital signs.

The spleen weighed 454 g, measured 19x12x4 cm and had an 8 cm capsular slit at the posterior border. Histology showed the splenic tissue to contain very small B cell follicles and a massive expansion of the red pulp by large activated T lymphocytes, consistent with infectious mononucleosis.

CASE REPORT 2

One year later the 15-year-old brother of the previous patient presented with vomiting and periumbilical pain which spread to the left shoulder tip. Infectious mononucleosis had been diagnosed that morning following a chorizal illness. On examination he had cervical lymphadenopathy and was tachycardic, with a low blood pressure which initially responded to fluid resuscitation. He had reduced air entry in both lung bases, and tenderness, rigidity and guarding in the central abdomen and left upper quadrant. His haemoglobin level on admission was 11.9 g/dl, white cell count was 15.3x10^9/litre and a Paul Bunnell test was positive. Ultrasound demonstrated an enlarged spleen with a haematoma on the lower pole and free fluid in the pelvis, consistent with splenic rupture. He underwent splenectomy on the same evening, having deteriorated despite fluid resuscitation.

The spleen weighed 553 g and measured 17x12x6.5 cm with two capsular tears. Histology showed marked expansion of the red pulp by immunoblasts, confirming a histological appearance of infectious mononucleosis and splenic rupture. Both children made an uneventful recovery and received pneumococcal and *Haemophilus influenzae* type B (Hib) vaccines before discharge.

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rupture varies from subcapsular haematomas to exsanguinating haemorrhage.

Although non-operative management is preferable in patients who remain haemodynamically stable (Johnson et al, 1981; Ali, 1993), laparotomy with splenectomy is a safe therapeutic approach and is preferred by some authors because of the extent of the histological changes in the spleen (Konvolinka and Wyatt, 1989). Unfortunately, we were unable to preserve the spleen in either of these cases.

CONCLUSIONS

This is the first report of spontaneous splenic rupture caused by infectious mononucleosis in siblings. The likelihood of this extremely rare complication occurring in two siblings by chance alone is infinitely small, although it is possible. This raises the possibility of an associated genetic or familial predisposition, although the nature of this, if it does exist, is merely speculative. HM

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