CASE REPORT

CAPS: a rare acute abdomen

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jposmith@gmail.com**SUMMARY**

An 11-year-old girl presented to the hospital with vomiting, left upper-quadrant pain and blood in the stool. An ultrasound scan showed no blood flow in the coeliac axis or the splenic artery. A contrast-enhanced CT scan then demonstrated no flow beyond the coeliac axis origin, a large clot in the distal superior mesenteric artery (SMA), a large splenic infarct and bilateral renal infarcts. Investigations revealed a positive lupus anticoagulant and she was given a presumptive diagnosis of catastrophic antiphospholipid syndrome (CAPS). She was anticoagulated and started on combination immunosuppressive therapy. Her surgical management was by SMA embolectomy, small bowel resection and proximal stoma formation. Stoma closure was performed at 3 months, and she was eventually established on full enteral feeds. Childhood CAPS is a rare condition with a very high mortality rate, and the successful outcome in this case reflects close multidisciplinary teamwork between medical and surgical specialities.

BACKGROUND

Catastrophic antiphospholipid syndrome (CAPS) is a very rare condition in childhood. At present, there are 20 cases of childhood CAPS on the international registry, of which 5 died, a reflection of the high mortality rate of this condition.^{1 2} It is more likely to present with microvascular thrombosis in multiple organs—including lung, heart, brain and kidney.³ Large vessel thrombosis is less frequent in CAPS. CAPS only occurs in a small percentage of paediatric antiphospholipid syndrome (APS) patients.⁴

CASE PRESENTATION

An 11-year-old girl presented to her local hospital with vomiting, severe left upper-quadrant pain and blood in the stool. Although previously well, she had been taking a reducing course of oral prednisolone due to a recent infective exacerbation of asthma. Further history revealed that the girl's mother had a lower limb arterial thrombosis in childhood, with no definite diagnosis, and was being managed with life-long anticoagulation therapy.

INVESTIGATIONS

An ultrasound scan performed on admission showed no blood flow in the coeliac axis or the splenic artery. A contrast-enhanced CT scan was subsequently performed, which demonstrated a stump of non-enhancing coeliac axis origin (arrow, figures 1 and 2), a thrombus within the distal superior mesenteric artery (SMA) (arrow, figure 1), splenic (arrow, figure 2) and bilateral renal infarcts.

Urgent investigations revealed a positive lupus anticoagulant (LAC), negative anticardiolipin antibodies (aCL), normal autoimmune screen, platelets and complement.

TREATMENT

Owing to the presence of multiple thromboembolic manifestations and positive LAC, she was given a presumptive diagnosis of CAPS, anticoagulated and started with combination immunosuppressive therapy. Her initial surgical management was by SMA embolectomy and limited small bowel resection, although a more extensive resection with proximal stoma formation followed on day 3 due to SMA reocclusion.

OUTCOME AND FOLLOW-UP

After an initial stormy course, she was discharged from paediatric intensive care unit after 2 months. Stoma closure was performed at 3 months, and she was eventually established on full enteral feeds. Repeat LAC and aCL were negative after 6 months, which would have been due to the ongoing corticosteroid therapy, and prevented a definitive diagnosis of CAPS being made according to the established classification criteria.⁵ At 6 months postdischarge from hospital, she continues on anticoagulation therapy, mycophenolate mofetil and a slow weaning dose of oral prednisolone.

DISCUSSION

Preceding infection is the most common trigger of CAPS and abdominal pain is well described as a

Figure 1 Contrast-enhanced CT scan of the abdomen shows a stump of non-enhancing coeliac axis origin (arrow) and a filling defect (thrombus) within the distal superior mesenteric artery (arrow).

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Figure 2 Contrast-enhanced CT scan of the abdomen shows a stump of non-enhancing coeliac axis origin (arrow), splenic (arrow) and partial left renal infarcts.

presenting feature. Intra-abdominal complications—including renal and bowel infarction—are described in up to 1/3 of CAPS cases.³ Of the published cases in the paediatric age-group, small vessel disease was a manifestation in the majority, most commonly affecting the renal and pulmonary vessels.³ Large vessel disease can also occur as part of a mixed picture. Of the cases on the CAPS registry, 12/20 have a diagnosis of systemic lupus erythematosus and 8/20 of primary APS.^{1 2}

There is a risk of future APS complications and/or a recurrence of a ‘catastrophic’ APS event, although this is attenuated by long-term anticoagulation therapy. Follow-up of 58 patients who survived CAPS showed that 66% had no further APS-related thrombotic events at 6 years (although only 1 of these was a child), and in the CAPS registry 9 of 280 (3%) have been reported as having a relapse.^{3 6}

There is a genetic contribution to APS, primary as well as secondary forms, which has been demonstrated by observational method.⁷ A number of human leucocyte antigen alleles have been shown to be associated with antiphospholipid antibodies.⁴

A familial or genetic diagnosis has not yet been confirmed in our index case, although it may be possible to elucidate this in the future.

Childhood CAPS is a rare condition with a very high mortality rate, and the successful outcome in this case reflects the necessity of close multidisciplinary teamwork between medical and surgical specialties.

Learning points

- ▶ Recognition of catastrophic antiphospholipid syndrome (CAPS) as a rare condition in childhood.
- ▶ Management of CAPS by combining surgery, immunosuppression and anticoagulation.
- ▶ Importance of considering family history.

Contributors All the authors contributed to the care and management of the patient and also to the writing and editing.

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