Acute pancreatitis as a complication of anaphylactoid (Henoch-Schönlein) purpura

Abdominal symptoms have long been recognized as a feature of anaphylactoid purpura, and are usually attributed to involvement of the bowel wall in the pathological process. We believe this to be the first report of pancreatic involvement.

Case report

A 7-year-old girl was referred with a 5-day history of painful, swollen feet and a 3-day history of bruising around the ankles. On the morning of admission her parents had thought her to be slightly feverish. There was no relevant past history and the only family history of note was that a paternal cousin had died of leukaemia in childhood. There was no history of trauma to account for the bruising and no symptoms referable to the gastrointestinal or genitourinary tracts. She had had no recent upper respiratory tract infection and had been on no medication.

On examination she was apyrexial and in no distress. There was marked bruising in both popliteal fossae and over both ankles, with a few petechiae over buttocks and shins. Both ankle and knee joints were swollen and slightly tender to palpation and the dorsum of her right hand was also swollen.

Initial investigations showed Hb 13.5 g/dl, white cell count 10.4×10⁹/l, with a differential of 75% polymorphonuclear leucocytes, 22% lymphocytes, and 3% eosinophils; platelets 260×10⁹/l; ESR 9 mm/h, and antistreptolysin O titre 125 Todd units. A throat swab grew a β-haemolytic streptococcus Lancefield group C; urine showed no cells and no growth. Joint x-rays were normal. Anaphylactoid purpura was diagnosed and she was treated with bed rest and penicillin V 250 mg qds.

The day after admission she vomited but had no pain. 2 days later she again vomited and complained of intermittent periumbilical pain with no radiation. There was slight generalized tenderness but no localizing signs. Bowel sounds were active but not obstructive. However the vomiting and pain continued and an erect x-ray of the abdomen showed some increase in gas with one or two fluid levels in the small bowel. Because of the risk of intussusception a barium enema was performed which showed 'no evidence of obstruction but a coarse mucosal pattern and some thickening of the mucosal folds which may be due to oedema'. Other investigations gave a white cell count of 15.3×10⁹/l, with 72% polymorphonuclear leucocytes; normal liver function tests, urea, and electrolytes. Serum amylase was 4700 IU/l (normal 70–300 IU/l). Faecal occult bloods were negative.

Acute pancreatitis was diagnosed and was treated with intravenous fluids and nasogastric suction. Over the next 24 hours her condition improved, although there was some persistent tenderness in the right hypochondrium, and the serum amylase was still raised at 3700 IU/l, but after 48 hours fell to normal. During the episode of acute pancreatitis the blood glucose was normal at 5.8 mmol/l (104.5 mg/100 ml) and serum calcium normal at 2.30 mmol/l (9.2 mg/100 ml).

Over the next week she had an occasional vomit but no other acute abdominal symptoms. However, she did develop fresh purpuric lesions around her buttocks, knees, and ankles, and her peripheral joints flared up again but settled spontaneously with further bed rest. After 6 weeks in hospital she was discharged and though she had had fresh crops of the purpuric rash since then, has not been troubled with other symptoms.

Discussion

In anaphylactoid purpura (Henoch-Schönlein syndrome), apart from the usual urticarial or purpuric rash, and the joint, gut, and renal symptoms, there are many reports of unusual sites of involvement including heart (Imai and Matsumoto, 1970), lungs (Jacome, 1967), brain (Lewis and Philpott, 1956), and testicles (Fitzsimmons, 1968). In recent reviews (Cream et al., 1970; Silber, 1972) approximately 50% of patients had symptoms referable to the gastrointestinal tract; but only a few cases progressed to the serious problems of massive blood loss or intussusception. The abdominal pain is thought to be due to localized oedema and haemorrhage into the bowel wall secondary to a vasculitis of the small vessels.

This patient must have had involvement of the pancreatic vessels causing her abdominal symptoms and the large rise in serum amylase levels. A proportion of patients with abdominal pain have no evidence of blood in the stools (Gairdner, 1948; Allen et al., 1960). In this group, at least, amylase levels should be estimated so that with more accurate diagnosis we can better manage this feature of the disease which may cause a lot of concern.

Summary

A case of acute pancreatitis complicating anaphylactoid purpura (Henoch-Schönlein syndrome) is reported. In cases of this syndrome with abdominal pain, serum amylase levels should be estimated.

I am grateful to Dr. M. McLean for help and for permission to report this case.
13q- syndrome

Family study

Clinical syndromes due to duplication or deletion of part of a chromosome are much less commonly observed than those due to trisomy or monosomy. De Grouchy (1976) gives an incidence of 0·11 per 1000 live births for recognized deletion syndromes as compared with 1·29 for the trisomies. The trisomies are due to anomalous separation of parental chromosomes during gametogenesis and neither the sibs of affected individuals nor the parents show abnormalities in the chromosomes of blood lymphocytes or extragonadal tissues. Duplications or deletions, however, are often the result of translocations which commonly exist in a balanced form in one of the parents and may well affect any sibs. Cytogenetic investigation of the family is therefore important in such circumstances and this need is not perhaps sufficiently appreciated in clinical practice.

We report the clinical and cytogenetic findings in the family of an infant who presented with anomalies due to partial deletion of a number 13 chromosome—anomalies which together form a recognizable clinical syndrome.

Case report

A girl weighing 1·96 kg was born 3 weeks after term in the 37-year-old mother's tenth pregnancy. Apart from unstable lie resulting in breech delivery, pregnancy was normal. Clinical examination confirmed that the child was mature and therefore small-for-dates. The head circumference was 31 cm and the length 40 cm. The following features were noted soon after birth: a triangular-shaped skull with frontal bossing, low-set ears, broad nasal bridge, bilateral ptosis, and choroidal colobomata. The neck was short with markedly redundant skin folds. The labia minora were rudimentary. The thumbs were small and both they and the fifth fingers were proximally placed. There was partial fusion of the fourth and fifth toes on both feet, and the feet were held in correctable equinovarus position. Within the first week intermittent cyanosis and a cardiac murmur were noted. Chest x-ray and electrocardiogram showed no specific abnormality, but investigation of the congenital heart disease was not pursued further. Hypotonia and poor feeding continued throughout life and there was no apparent visual awareness. The child died at the age of 7 months of bronchopneumonia.

Both parents and all 7 older sibs were examined and were well and physically normal. 2 of the mother's previous pregnancies ended in stillbirth. The first baby was stillborn at term; he weighed 4·14 kg and at necropsy a normal physical appearance was noted. The sixth pregnancy ended at 24 weeks with the spontaneous delivery of a stillborn female infant weighing 0·54 kg; gross appearance was normal, but necropsy was not performed.

Necropsy findings

Brain. There was failure of perforation of the cribiform plates of the ethmoid bones of the skull. The brain showed two cerebral hemispheres in which there were identifiable hippocampal formations, but was externally abnormal as both olfactory bulbs and tracts were missing. Slicing the brain showed the following abnormalities: the corpus callosum was reduced in thickness and showed marked thinning at its rostral end. This structure was also reduced in length and the splenium was not well developed. There was some dilatation of both lateral ventricles and of the third ventricle. The caudate nuclei were not well developed structures and were represented by bands of grey matter bounding each lateral ventricle. The fornix, which had developed bilaterally, remained widely separated with formation of two