is considered a reasonable surgical risk. As Millard and other authors[2] have already stressed, this lesion should be considered in those cases of massive hæmatemesis in elderly males when there is no radiologically visible lesion, especially if bleeding continues after subtotal gastrectomy. A careful examination of the surgical specimen is desirable in all cases where the location of the bleeding point has not been determined before or during operation, as these lesions are small and easily overlooked.

SUMMARY

A case of aneurysm of a gastric artery which ruptured intragastrically is presented.

The lesion occurred in an elderly male and the involved vessel showed early arteriosclerosis, as in previously reported cases.

It is possible that in this case the old splenic injury may have been a causative factor.

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FAMILIAL HÆMOLYTIC ANÆMIA: CONCURRENT CRISES IN THREE MEMBERS OF A FAMILY*

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THERE ARE AT LEAST ELEVEN reports in the literature describing concurrent crises in members of families affected with familial hæmolytic anæmia. In none of these cases has there been any demonstrable precipitating factor, and the cause of the phenomenon is unknown. The present report documents another instance of concurrent familial hæmolytic crises.

The family, consisting of a father, mother, and four children, lived in a small town in rural Manitoba. All were in good health until December 8, 1952, when Clare, aged 18, became ill with nausea, vomiting and abdominal pain. She was working in Winnipeg but spent the weekend of December 6 and 7 in the country with her family. On December 13, she returned to her parents' home and she was then observed to be jaundiced. The next 10 days were spent in bed and by December 20 she had recovered. In the meantime, her younger sister Shirley, aged 14, with whom she was sleeping, became ill.

Shirley's sickness began on December 14 with vomiting and headache, followed shortly by the appearance of jaundice. On December 19 she was admitted in a semistuporous state to a country hospital. Her hæmoglobin level was stated to be 20%. She was given two bottles of blood and transferred to the Winnipeg General Hospital on the evening of December 20, 1952. She was a slight, pale girl, without definite icterus, semi-stuporous but easily roused. The temperature was 102° F., the pulse rate 125 and the respiration rate 28. The pupils were small and reactive. The mouth and tongue were dry and there were general signs of dehydration. There was no lymphadenopathy. The chest was clear; the heart was rapid with ill-defined murmurs. The liver was not palpable; in the left upper quadrant there was some resistance which was thought to be spleen, but it was soft and not definitely palpable. The skin and mucous membranes showed no hæmorrhages. There were some equivocal neurological findings. Routine urinalysis was normal. Hæmoglobin level was 5.8 g.; leukocyte count 10,800.

The presenting picture was that of high fever, severe anæmia and splenomegaly. The differential diagnosis included acute leukæmia, acute hepatitis, and some acute hæmolytic process, possibly infective. Pending the completion of tests in the morning, the following supportive therapy was ordered to be given over the next 12 hours: 1,000 c.c. of whole blood, 1,500 c.c. of 10% invert sugar in distilled water with 1 g. of terramycin added to the intravenous fluid. In the morning her condition was unchanged. At 1:00 p.m. she suddenly went into circulatory collapse, with pulmonary œdema, and died.

While in retrospect the diagnosis could have been made at the bedside from an examination of the peripheral blood smear, this was not carried out until the morning after admission and the reports became available only after death. Differential count showed mature neutro-

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phils 91; young neutrophils 0; eosinophils 1; lymphocytes 4; monocytes 4; occasional plasma cells. The average erythrocyte diameter was 6.0 μ, the range being 4-11 μ. The erythrocytes showed hypochromia 1+; poikilocytosis 1+; anisocytosis 3+; microcytosis 3+; macrocytosis 1+; polychromasia 1+; spherocytosis 3+. A reticulocyte count was ordered but the test was not completed. The cerebrospinal fluid showed a total protein of 10 mg. %, no cells, and a negative colloidal gold reaction. The serum bilirubin level was 3.0 mg. %.

A bone-marrow aspiration performed half an hour post mortem was reported by Dr. J. M. Lederman as follows: "The leukocyte series is essentially normal. There are many clumps of phagocytic reticulo-endothelial cells. Differential count of the erythrocyte series shows megaloblasts 7.8%; early erythroblasts 6.5%; late erythroblasts 6.2%; normoblasts 0.3%. The active erythropoiesis at an early erythroblastic level and the very small erythrocyte size with a high proportion of spherocytes are consistent with a congenital hæmolytic anæmia in exacerbation."

At autopsy there were no significant findings except that the spleen weighed 480 g., and on microscopic examination showed marked congestion with a loss of structural detail. A diagnosis of familial hæmolytic anæmia in acute crisis was consistent with the clinical picture and the findings in the peripheral blood and bone marrow. Up to this point the family history[1] of jaundice had not been recognized.

On December 22, 1952, Karl, aged 11, became jaundiced and began to run a fever. On December 25, he developed severe epigastric pain, vomited, and had two nosebleeds. He was admitted to the Winnipeg General Hospital on December 26. He was fully conscious and cooperative. The temperature was 104° F.; pulse 120; respiration rate 25. The scleræ were jaundiced. He had herpes facialis. There was no lymphadenopathy. The lungs were clear. There were faint aortic and apical systolic murmurs. The liver was not palpable; the spleen was felt 3 cm. below the costal margin on inspiration. There were no neurological abnormalities. There was no bile in the urine and the urobilinogen concentration was 0.45%. Urinalysis was negative. Spectroscopic examination was negative for hæmatin, coproporphyrin, and hæmatoporphyrin. The Coombs test was negative. Hæmoglobin level was 5.8 g. %; the erythrocyte count 1.9 million; the leukocyte count 10,800. Erythrocyte sedimentation rate was 62 mm. in 1 hour (Westergren). The differential count showed mature neutrophils 32; young forms 24; eosinophils 1; basophils 1; lymphocytes 25; monocytes 15; plasma cells 2. Smear showed an average red cell diameter of 5.9 μ; anisocytosis 2+; and spherocytosis 2+. Reticulocytes 0.9%. M.C.V. 69 cubic microns. M.C.H. 24 $\gamma\gamma$. M.C.H.C. 35%. Hæmolysis began at 0.5% saline and was complete at 0.42%. Serum bilirubin was 2.6 mg. %. Bone-marrow examination showed many spherocytes and marked megalo-erythroblastic hyperplasia (megaloblasts 8.5%; early erythroblasts 13.9%; late erythroblasts 13.5%; normoblasts 1.5%.) Diagnosis: Familial hæmolytic anæmia with maturation arrest.

The patient was given two pints of blood on December 26. Thereafter, his condition improved rapidly. The fever abated. The serum bilirubin level fell from 2.6 mg. % on admission to 1.6 mg. % on January 12. The following hæmoglobin levels and reticulocyte counts were recorded:

Date	*Hb (g. per cent)*	*Reticulocyte count (per cent)*
December 27	6.6	0.9
December 29	6.9	
December 30	6.4	0.5
January 2	6.9	19.6
January 4	8.4	12.2
January 6	9.5	9.1
January 12	11.2	6.8

The patient was discharged on January 14, 1953.

It was subsequently established on questioning the parents that jaundice had been observed in three of the four children on at least one previous occasion, but this had not been a matter of concern as it apparently had not affected their health. Both Clare and Karl recalled previous jaundice associated with dark urine. The father, aged 50, a weatherbeaten Swedish-born fisherman, had always been healthy and had never been aware of jaundice or of dark urine. There was no known jaundice in any of his family. His father had died at age 90 and his mother at age 50, of unknown cause. He was the only survivor of 12 children, 10 having died in infancy and 1 at the age of 14, all of unknown cause. Physical examination was normal. His hæmoglobin level was 14.5 g. %; and his reticulocyte count was 1.2%. Blood

smear showed anisocytosis 3+; and spherocytosis 2+. Hæmolysis began at a dilution of 0.5% saline and was complete at 0.46%. The icterus index was 11, but the serum was cloudy, and the icterus was probably not above normal. The mother, aged 44, of Icelandic extraction, was born in Canada, one of five siblings. She had always been well, and there was no family history of jaundice. Studies of her blood were normal in all respects. Physical examination and blood studies of Louis, aged 17, were negative and there was no history of jaundice. Clare, aged 18, while the first one to become ill, was not seen until she had recovered, at which time she did not appear icteric. Her spleen was palpable 3 cm. below the costal margin. Her hæmoglobin level was 9.5 g.; erythrocyte count 3.5 millions; leukocyte count 5,100 and reticulocyte count 8.6%. Blood smear showed spherocytosis 2+. M.C.V. 87 cubic microns. M.C.H. 27 γγ. M.C.H.C. 31%. A red cell fragility test showed hæmolysis beginning in 0.5% saline. The serum bilirubin was 2.6 mg. %.

It was apparent, therefore, that the father was the carrier of the hæmolytic trait and that three of the four children had inherited the disease. It was unfortunately impossible to trace back to preceding generations. The family tree is outlined in Fig. 1.

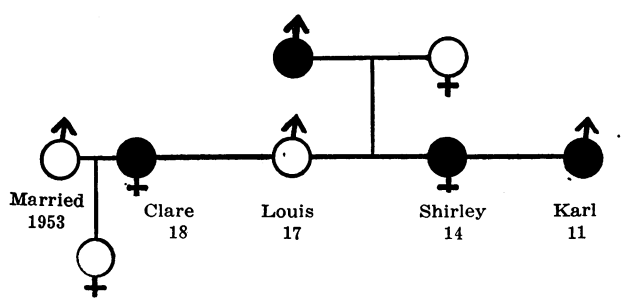

Fig. 1.—Familial hæmolytic anæmia: concurrent crises in three members of a family.

Clare underwent splenectomy on February 27, 1953, followed by an uneventful convalescence. Postoperatively the serum bilirubin level fell from 3.6 to 0.7 mg. %. Hæmoglobin level rose from 11.2 to 13.9 g. %; the erythrocyte count from 3.93 to 5.20 millions. Reticulocytes fell from 4.4 to 1%. The spleen weighed 334 g. and measured 12.5 x 10.5 x 4.5 cm. The pulp was stuffed with blood, the lymphoid tissue was relatively scanty. The endothelium lining the sinusoids was cuboidal. Clare was married shortly after her splenectomy. In May 1955 her 9-month-old female child showed a normal blood picture with no spherocytes and no increase in red cell fragility. On March 30, Karl underwent splenectomy; there were no complications. The hæmoglobin level rose from 11.2 to 12.6 g. %; the erythrocyte count from 4.26 to 4.55 millions. The serum bilirubin fell from 2.8 to less than 0.2 mg. %; the reticulocyte count from 7.1 to 3.9%.

DISCUSSION

Eleven reports have been found recording the simultaneous occurrence of crises among different members of families with congenital hæmolytic anæmia. In no case was there any demonstrable precipitating factor, such as exposure, infection, drugs, or autohæmagglutinins. As many as eight members of a family have had concurrent crises, the actual numbers reported being: four,[1] three,[2] eight,[3] four,[3] six,[4] five,[5] five,[6] four,[7] two,[8] four,[9] four,[10] two.[11] It was considered of interest to add a further instance of multiple concurrent crises in a family with hereditary spherocytosis.

SUMMARY

Over the course of 15 days, three siblings with hereditary spherocytosis developed acute exacerbations of their disease, with one fatality. The other two recovered and were later subjected to splenectomy. There was no evident precipitating factor. Study of the family showed the father to be the carrier.

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