NEUROLOGICAL COMPLICATIONS IN THE SCHÖNLEIN-HENOCH SYNDROME

BY

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Although a few textbooks persist in describing separately 'Schönlein's purpura' and 'Henoch's purpura' the recognition of the Schönlein-Henoch syndrome as a disease entity cannot now be disputed. The characteristic exanthem, the frequent occurrence of arthritis and intestinal symptoms, together with the possibility of renal damage, are well known.

Serious neurological complications of this condition must be rare, judging from the medical literature, and therefore the following cases are of particular interest.

The diagnosis of the Schönlein-Henoch syndrome in each patient was based on the finding of an exanthem of the type and distribution described by Gairdner (1948), namely, intestinal colic and vomiting, arthralgia and nephritis (in two of the three cases).

Case Reports

Case 1. Howard M., born on February 19, 1947, was admitted to hospital on June 29, 1955, with abdominal pain, vomiting and passage of bloody stools. The following day laparotomy was performed. There were several areas of small gut showing extensive subserous haemorrhages, and the bowel lumen was filled with blood. After five days he developed typical exanthem and on the same day complained of severe headache. Later in the day he had a generalized convulsion, his blood pressure being 160/110 mm. Hg. The C.S.F. was normal. He was given hexamethonium to control hypertension, but this resulted in gut and bladder paralysis. The blood pressure subsequently settled spontaneously without further convulsions.

On July 8 he suddenly complained of intense headache. Examination revealed much spinal rigidity, and the C.S.F. was intimately bloodstained. The symptoms cleared in 10 days, during which the right wrist became swollen for two days, and he suffered several attacks of abdominal pain. The C.S.F. was found to be xanthochromic on July 19. On July 12 the urine became abnormal for the first time (protein and red cells present). No further skin eruptions occurred.

During August there was a deterioration in the patient's renal condition. Oedema and ascites appeared

and the blood pressure again rose. On August 4 the blood pressure was 160/110 mm. Hg and next day there was some generalized twitching. Reserpine was used to control hypertension and was discontinued after a week. A trial with cortisone was started on August 15 but had to be discontinued after 48 hours owing to a dramatic increase in the oedema. The oedema disappeared equally dramatically on discontinuing hormone treatment.

The boy remained fairly well, but rather wasted, and with abnormal urine. On September 11 he suffered a mild convulsion (blood pressure 140/110 mm. Hg) and developed left hemiplegia, which has persisted. Since this time there has been a progressive improvement in his general condition. The urine remains abnormal with protein 20-100 mg. % and a few red cells and granular casts are present. The blood pressure remains normal (90/60 mm. Hg). With physiotherapy there has been some improvement in the hemiplegia, but he is still in hospital.

This case illustrates (1) convulsions, presumably hypertensive, in the absence at first of urinary abnormalities; (2) subarachnoid haemorrhage; and (3) intracerebral haemorrhage or thrombosis, presumably due to arteriolitis.

Case 2. Sheila N. was born on June 14, 1939.

She was admitted to hospital on October 16, 1949, with abdominal pain and vomiting. She had had a haemolytic streptococcal throat infection two weeks previously.

Appendicitis was suspected but on October 18 at laparotomy a normal appendix was removed. Three enlarged ileocaecal glands were noted.

The exanthem of the Schönlein-Henoch syndrome appeared over the buttocks and left forearm on October 20, and the patient complained of arthralgia with periarthral oedema.

Melaena and further crops of the exanthem appeared during the next few days, but the discomfort diminished.

On November 4 she complained of frontal headache and she became confused. Her blood pressure was 150/90 mm. Hg. Her symptoms were worse by the following day and neck stiffness was noted. Lumbar puncture revealed heavily bloodstained fluid under con-

siderable pressure. Subarachnoid haemorrhage was diagnosed and was treated conservatively.

The cerebral symptoms cleared rapidly during the next day or two, but on November 7, 1949, a trace of albumin and a few red blood cells were discovered in the urine.

During November and December the patient complained of occasional intestinal colic and arthralgia, and there was deterioration in the renal condition, marked by many fluctuations in the albumin and red blood cell content of the urine. Hyaline and granular casts were prevalent.

Early in February her urine contained 7 g. per litre of albumin. She became very oedematous early in March, and died on March 14, 1950.

A necropsy limited to the abdominal cavity was allowed. The findings were subacute nephritis with preglomerular arteriolitis in some areas of the renal cortex.

This case illustrates (1) subarachnoid haemorrhage and (2) renal failure and death.

Case 3. Caroline A. was born on April 22, 1949.

She was first seen at home on October 29, 1955. Two weeks previously she had a mild sore throat lasting two days. Five days previously she became listless and vomited once the following day. A day later she complained of pains in several joints, but no swelling was noticed. The day before being seen she developed a rash on the arms, legs and buttocks. The Schönlein-Henoch syndrome was diagnosed and the case was left to the care of the family physician. The next day severe abdominal pain developed, and dark fluid stools were passed for three days. On November 1 she complained of a 'funny feeling', her expression became vacant and later she lapsed into unconsciousness but did not convulse. After one and a half hours she awoke and was rational. Two hours later a similar episode occurred, and after another two hours a third episode, in which she had a major convulsion lasting 10-15 minutes. Subsequently she was admitted to hospital on the same day.

On admission the blood pressure was 130/80 mm. Hg. Treatment with phenoxymethyl penicillin and phenobarbitone was given. A further generalized convulsion occurred on the day after admission. This was rapidly controlled with intramuscular paraldehyde. She continued to have crops of skin lesions and occasional attacks of abdominal pain, with bloody stools. The blood pressure began slowly to settle, but on November 18 it rose suddenly to 145/120 mm. Hg. She had a further convulsion which was promptly controlled with intramuscular paraldehyde. Subsequently the blood pressure gradually subsided without specific treatment. On December 8 the blood pressure was 90/40 mm. Hg and only slight fluctuations were encountered subsequently.

In view of the continued skin and abdominal features, treatment with prednisone was begun on November 11, but this had no dramatic effect on the course of the disease. Between December 6 and 13 the drug was

gradually withdrawn. After December 16 no further skin eruptions or attacks of abdominal pain occurred. On December 17 she was discharged for further surveillance at home. During the first two weeks at home she had several mild attacks of abdominal pain, but her stools remained normal and there was no further exanthem. Her blood pressure remained steady. After a further week she was allowed up and has remained well ever since.

At no time during the course of the illness or subsequently has any abnormality in the urine been detected, and the blood urea has been normal.

This case illustrates convulsions, probably hypertensive in origin, in the absence of urinary abnormality.

Discussion

Little mention of involvement of the central nervous system can be found in the literature dealing with the Schönlein-Henoch syndrome. Osler (1914) in the last of his four articles on the visceral lesions of purpuras and allied conditions described neurological complications, but only one case undoubtedly suffered from the Schönlein-Henoch syndrome. This patient, a 13-year-old boy, had nephritis as well as intestinal colic and the exanthem, and had a transient hemiplegia on several occasions. Gracie (1924) described the case of a 13-year-old boy who suffered from colic, arthralgia, exanthem and macroscopic haematuria. The child had several convulsions but later appeared to make a full recovery. Other cases of convulsions have been mentioned in connexion with this disease, but they have all had nephritic features (Levitt and Burbank, 1953; Wedgwood and Klaus, 1955). Case 3 reported above had no signs of renal involvement at the time of the convulsions.

Green (1946) described a girl aged 7 years who suffered from a subarachnoid haemorrhage during the course of an attack of the Schönlein-Henoch syndrome. This would appear to be the only previous recording of such a complication, although he quoted a personal communication from Davis who claimed to have seen two similar cases in 1,100 (adults and children) studied by him. Davis (1948), however, makes no mention of these in his own paper.

That the disease process is essentially an arteriolitis was made plain by Gairdner (1948). The skin is invariably affected and the gut and joints are frequently involved. It is not certain what lesion is present in the kidneys, which are diseased in a large number of cases (Philpott, 1952). The course and prognosis of the kidney lesions suggests something different from glomerulonephritis, probably the same arteriolitis which involves the skin and gut. Widespread arteriolitis and arteritis have been

described in necropsy reports by Wassilieff (1937), Gairdner (1948) and Levitt and Burbank (1953). It would be surprising if the cerebral vessels escaped and the cases recorded in this paper strongly suggest that they are involved. Headache was noted in 25% of 116 cases of the Schönlein-Henoch syndrome (Lewis, 1956). By no means all these patients with headache had either hypertension, fever or nephritic features, and it seems likely that the symptom is due to the same vascular changes as found in the skin and elsewhere.

The convulsions suffered by Cases 1 and 3 were presumably hypertensive in origin, although they may have resulted directly from arteriolitis in the cerebrum. Case 3 is particularly interesting in that her urine never became abnormal. (Addis counts, however, were not performed in this case.) It must remain a matter of speculation as to whether she had preglomerular arteriolitis or not.

Summary

The cases of two children who developed sub-arachnoid haemorrhage during the course of an attack of the Schönlein-Henoch syndrome are described, together with a report of a case of hypertensive encephalopathy in the same condition without, however, renal abnormality.

The literature is discussed.

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