MENINGISM, PERICARDITIS/MYOCARDITIS UNRECOGNISED MANIFESTATIONS OF ACUTE PORPHYRIA

Pages with reference to book, From 239 To 243

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Abstract

A case of acute varigate porphyria associated with meningism, pericarditis/myocarditis is presented and relevance in treatment discussed(JPMA36:1986).

MENINGISM, PERICARDITIS/MYOCARDITIS ASSOCIATED WITH ACUTE VARIGATE PORPHYRIA

During acute porphyria (Acute Intermittent Porphyria, Varigate Porphyria and Hereditary Coproporphyria), the patient may present with abdominal pain, vomiting, constipation, tachycardia, hypertension, peripheral neuritis, fits and psychosis. [1] A case is reported of acute varigate porphyria with meningism, pericarditis/ myocarditis during an acute attack. A search in literature did not reveal such an association reported before. Focal myocardial necroses, however, have been found in acute intermittent porphyria.[2]

CASE REPORT

A 51 year old lady was admitted to the. Surgical Unit with sudden onset of severe epigastric pain radiating towards the right iliac fossa. She had nausea and vomiting of 8 hours' duration. The vomitus was undigested food only. There was no significant past medical history. The patient did not take any drugs. No other member of the family had suffered similar problems.

She was a housewife in Halifax, married for the second time. No children. The rest of her family lived in Scotland. She had lost contact with her family a long time ago. She was not aware of any illness running in her family. She was a non-smoker and teetotaler.

On examination she had tenderness and guarding in the right iliac fossa. The rest of the examination was negative. Her white cell count was 14 x 10^9. Urea and electrolytes were normal.

She had laparotomy under general anaesthesea (Cyclopentothal). Normal appendix was removed. Her ovary was cystic. Oophorectomy was performed. Histology showed benign cyst adenoma. Post-operatively she was nauseated most of the time, although the vomiting and abdominal pain had disappeared. She was discharged home on the 5th day. She came back to the Accident and Emergency Department 4 days after discharge from hospital in a confused state, unable to give history. Her husband said that she was nauseated all the time and vomited 1-2 times a day since discharge. She also complained of headache and abdominal pain which gradually got worse.

On examination she was drowsy but arousable; apyrexial; pulse 130 per minute; blood pressure 160/110. There was tenderness in the left side of the abdomen. The rest of the examination was normal. Investigations; sodium-111 mmol/1, Potassium 2.5 mmol/1, Chloride 63 mmol/1, bicarbonate-29 mmol/1, urea-10.1mmol/1.

She was started on 1.V. saline. The next morning she had a grand mal fit which was controlled by I.V. Valium and a medical opinion was sought. On examination she was drowsy, non-communicative, restless; temperature 38.5. Neck rigidity ++. Kernig's sign-negative. Fundi normal. Pulse 150 per minute regular. Blood pressure 180/1 10. Heart sounds normal. No munnur. Pericardial rub ++.

Abdomen soft. No viscera palpable. Moving all four limbs. Reflexes normal.

INVESTIGATIONS

Chest X-ray normal

ECG-LVH and sinus tachycardia

Chloride-83 mmol/1

Bicarbonate-28 mmol/1 (Figure I)

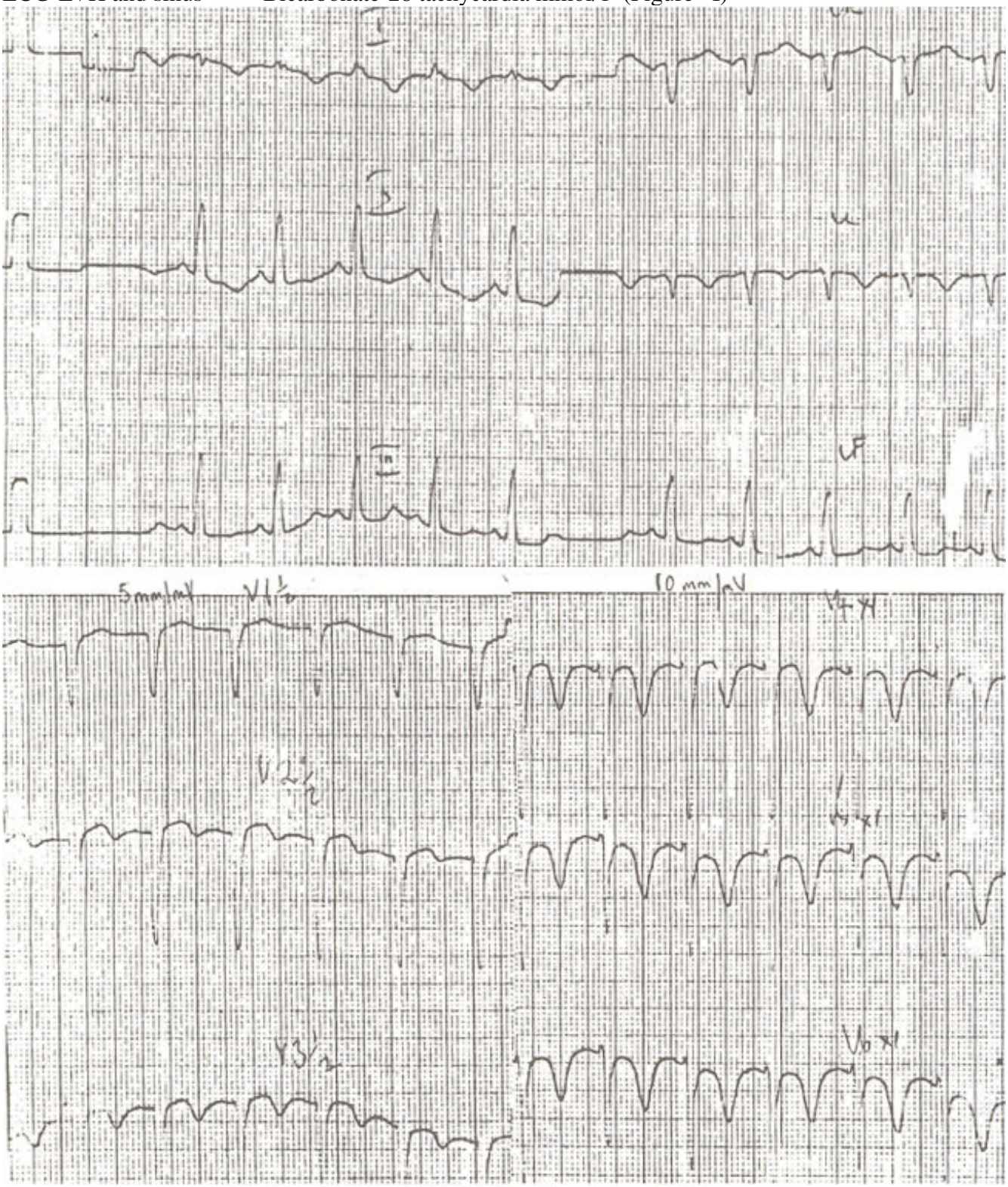

Figure 1. ECG Showing LVH and Sinus tachy cardia

WCC-17.5 x 109 Blood sugar- 4.9 mmol/1
Hb - 13.5 gm/dl Serum calcium - 2.2 mmol/1
Serum sodium - 112 mmol/1 Serum phosphate -.7 mmol/1
Urea - 6.3 mmol/1 Brucella & myco-plasma antibodies -negative
Potassium - 3.7 mmol/1 Cold agglutinins - negative
Creatinine 76 mmol/1 Coomb's test - negative Clotting screen -normal
Heaf test- negative Urine porphobilino-gen - 98 mmol/1
Influenza virus A & B (normal 0-.25 adenovirus mmol/1)
Chlamydia, Psittacosis, C. Serum osmolality -Burneti, 252 mosmol/kg
Coxsackie virus antibodies Urine osmolality AST LDH
less than 32 in paired sera. 573 mosmol/kg
No virus was isolated from blood, urine and faeces.

Cerebro Spinal Fluid

Pressure normal, and clear 18.12.85 180 183
Colourless 19.12.85 217 163
Proteins-.4 gm/dl 20-12.85 166 90
No RBC's, white cells or 23.12.85 78 87 organisms
Glucose - 4 mmol/1 31.12.85 59 69
No white cell castes or RBC's
Mid Stream Specimen of urine Serum alkaline phosphatase - 7KA/dl
Bilirubin - 6 mmol/1
No proteins. No growth
Stools, high vaginal swab, Total proteins - 72 throat swab g/1
did not grow any pathogens Albumin - 34 g/1
Blood cultures x 6 - no Total globulins - 38 bacterial g/1
growth EEG - an excess of slow wave activity
Thyroid function tests over both hemis
normal pheres
anti-nuclear factor- negative
Rheumatoid factor positive 1:60
ASO titre less than 200 IU/ml

ECG

A. Sinus tachycardia. Deep T wave inversion in all leads (pericarditis/myocarditis) and LVH (ECG Pictures).

A diagnosis of acute porphyria attack with Syndrome of Inappropriate Antidiuretic Hormone (SIADH), meningism, pericarditis/myocarditis was made. She was given 10% Dextrose 500 ml 6-hourly alternating with .9% saline 500 ml 6-hourly. I.V. Ceftazoxime lgm twice aday. She was sedated with chlopromazine. Her tachycardia was controlled with Inderal I.V. She gradually regained consciousness. Her pyrexia settled but she retained pericardial rub. She was handling water load well and did not need water restrictions for SIADH.

She started eating on the 4th day. She was well orientated, her pericardial rub disappeared. She was mobilised and discharged home to be reviewed in the Outpatient Department in four weeks. Sodium on

discharge was 141 mmol/1, potassium 4.7 mmol/1, Chloride -106 mmol/1, bicarbonate-27 mmol/1, urea -7.2 mmol/1.
She remained well at home for four weeks had no neck stiffness and the pericardial rub had disappeared. At her Out-Patient check-up her blood pressure was 110/60. Repeat ECG had no T wave inversions.
B. 23.1.86. Sinus rhythm. Twaves back to normal(Figure 2)

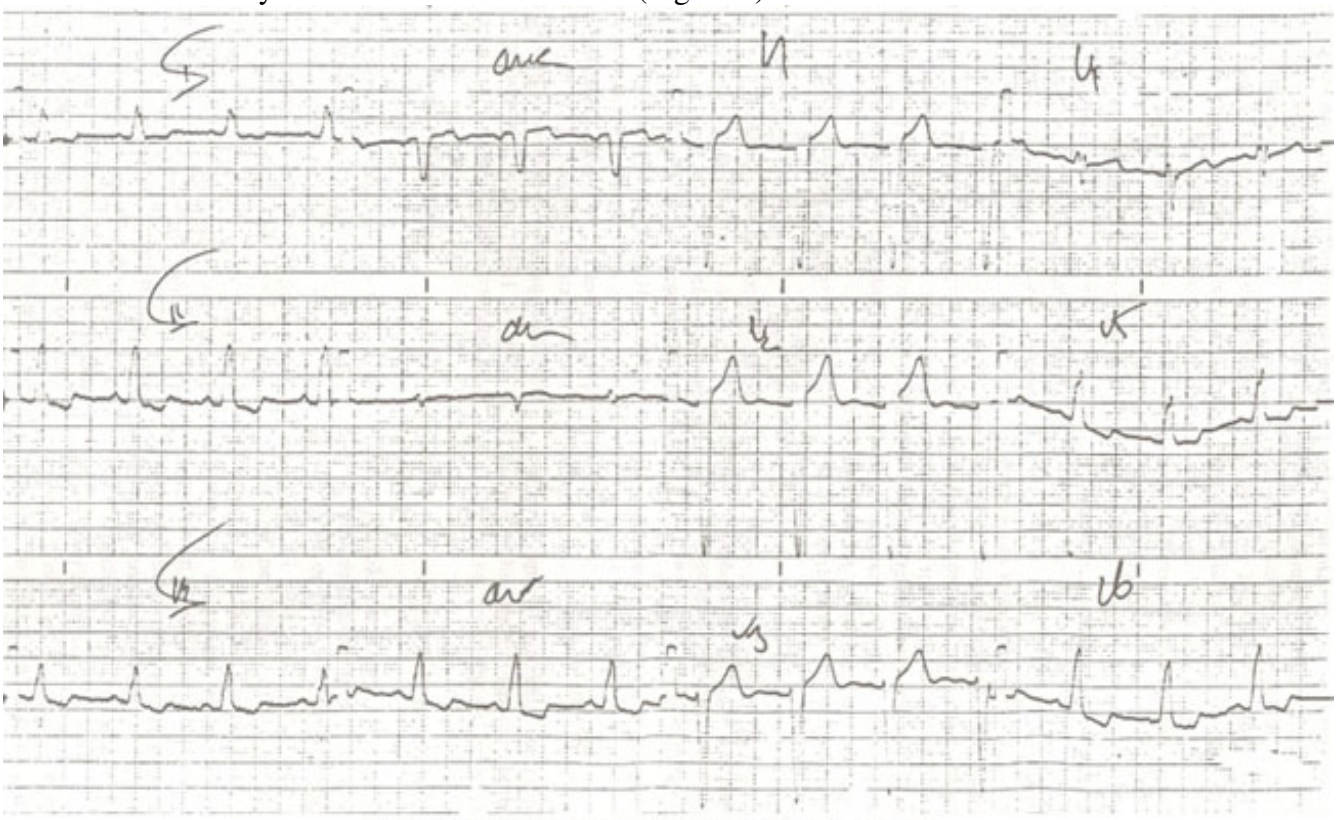

Figure 2. (23-1-86) ECG Showing Sinus rhythm T Waves back to normal.

Further tests on her urine showed marked increase in coproporphyrin and some increase in uroporphyrin. Faeces showed raised protoporphyrin and coproporphyrin. These results confirmed that she had varigate porphyria.
Although no significant past medical history was available at the time of admission, it was later revealed that she was admitted to hospital in 1977 with vomiting, headache, pyrexia and neck stiffness. Meningitis was provisionally diagnosed but CSF was normal. Sinusitis was also suspected. Her sinuses were washed out and were found to be clear. No cause for her symptoms was found. She had been suffering from headaches repeatedly. She also had vomiting and abdominal pain frequently when she went on holiday and it was put down to change of food. Her family was contacted in Scotland and it was discovered that her sister and niece both had varigate porphyria.

COMMENTS

This patient had an acute attack of varigate porphyria associated with SIADH and meningism pericarditis/myocarditis. She had one attack of meningism in 1977, which seems retrospectively to be associated with acute porphyria attack. There was no other aetiology of meningism, penicarditis/myocarditis in this case. Her SIADH, meningism and pericarditis/myocarditis recovered when she came out of the acute episode. She did not need any special treatment for it.

SIADH is a known complication of acute porphyria[3] but meningism, pericarditis/myocarditis has not been reported in literature before. Necrotic lesions of myocardium occur with acute intermittent porphyria[2].

Hypertension and sinus tachycardia are the most common cardiovascular manifestations of acute porphyria.[1] Rarely have cardiac failure and postural hypotension been found. Respiratory failure due to muscle weakness is one cause of mortality in acute porphyria. Myocarditis/pericarditis may be another one, especially when large amount of parental fluids are administered. Awareness of this manifestation and care in I.V. fluid administration should reduce mortality in acute porphyria attacks.

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