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A young boy with abdominal pain

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Acute Intermittent Porphyria - A Diagnosis to Consider

Pages with reference to book, From 45 To 45

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Acute intermittent porphyria (AlP) is iatrogemc, a disease of medical progress and development. Serious clinical manifestations are often precipitated by ingestion of prescribed drugs. Like syphilis or hysteria, AlP may be termed as "little imitator[1]. We are presenting the clinical spectrum of AlP, with a view to highlight the possible misdiagnoses and important management issues.

Patients, Methods and Results

A computer search for AIP patients was made from the medical records of cases admitted between Januaiy, 1991 and December, 1993. The charts of AlP patients were reviewed for presenting features, biochemical abnormalities and provisional diagnosis on first admission. There were 24 patients witha mean age of 32.4 years (range 16-54 years). The clinical characteristics were abdominal pain and vomiting in 24 (100%), mental confusion in 19 (79%) and constipation in 18 (75%) patients. Other presenting features were backache, diarrhoea, chest pain and unconsciousness in less than 20% cases. Common clinical signs were tachycardia in 21(87.5%), fever in 17 (71%), dehydration in 17 (7 1%) and hypertension in 7 (29%) patients. Syndrome of inappropriate antidiuretic hormone (SIADH) secretion was present in 8 (33%) and depletional hyponatremia was present in 7(29%) patients. There were on an average 1.8 hospital admissions before the diagnosis of AlP could be made. Only six (25%) patients were diagnosed on their first admission as suffering from AlP. The other diagnoses on first admission were acute encephaliiis in eight (33.3%), intestinal obstruction due to antispasmodics in three (12%), faecal impaction in five (21%) and hysterical motor weakness in two (8%) patients.

Comments

The manifestations of AlP in the present series are comparable to the large series[2,3]. The clinical features were mostly neurovisceral, highly variable and non-specific. This highlights the fact that AlP can be easily misdiagnosed and a high degree of clinical suspicion is required to make an early diagnosis.

Hyponatremia has come up as a special management problem in this review. It is recommended that before presuming the SIADH as the cause of hyponatremia, every attempt should be made to exclude any fluid and electrolytes loss causing hyponatrernia so that the unnecessary water restrictionbe avoided. Another important point inthe management is the screening of the family members to detect the career gene. These individuals are potentially at a high risk of developing an attack of AlP.

We conclude that AlP should remain as one of the differential diagnosis in the clinical spectrum and its manageinent should be in the hands of expert professionals.

References

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