Acute Encepholopathy Associated with Acute Hepatitis - Reye's Syndrome

A Case Report

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Since Reye's original description in 1963[1], more than 85 cases of this syndrome have been reported from several parts of the world [2,3]; but to the best of our knowledge this is the first report of Reye's syndrome from this country.

Case Report

A.Ç. (HCH 68/68066), a six-year-old Turkish boy, was admitted to our hospital with a three-day history of vomiting and malaise, but with no rise in temperature. One day prior to admission he had twitching movements of the legs, and became listless and stuperous. No history of trauma was elucidated.

On admission the patient was unconscious and unresponsive to external stimuli, however the pupils of his eyes were equal in size, and reacted sluggishly to light. His temperature was 37°, and his pulse and respiratory rates were 200 and 36 per minute respectively. The initial blood pressure was 120/60mm/Hg. His height was 117 cm and weight 16 kg. Eye fundi were normal and the neck was supple.

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The right ear drum was perforated, but no pus was seen; the throat was hyperemic and an accumulation of saliva was observed. Transmitted rales were heard over both lung fields. The abdomen was soft and the liver was two cm below the right costal margin, but the spleen was not palpable. Deep tendon reflexes were hyperactive in both lower extremities, but normal in the upper extremities. Both the Babinski and clonus signs were positive, and the patient had clonic seizures at the time of admission.

Laboratory examination indicated granulocytosis (15.000 and 16.000 WHB per cmm on two occasions with 85 per cent segs). Hb was 11.7 gm per 100 ml, and platelets on the peripheral smear were abundant. The cerebrospinal fluid (CSF) was clear and under normal pressure.

Urinealysis revealed a trace of albuminuria, NPN was found to be elevated (78 and 120 mg per 100 ml on two occasions). Electrolytes (Na:138mEq/L; $Co_2$26mEq/L; K:5.6mEq/L; Cl:100mEq/L) were within the normal limits. However, the patient later developed acidosis (Co_2:17.5mEq/L) and mild hyponatremia (Na:133mEq/L). SGOT:1160 u; SGPT:1500 u; bilirubin 1.8 mg per 100 ml (0.8 mg conjugated and 1 mg unconjugated), alkaline phosphatase 23.4 Bodansky units, ammonia 260 mg per 100 ml; and Quick prothrombin time 12 seconds. Blood sugar at the time of glucose effusion was 81 mg per 100 ml. EEG revealed generalized cortical and subcortical dysrythmia with some suppression bursts. Skull and chest x-rays and an EKG did not show any abnormalities. CSF and blood cultures were negative, and no pathogenic organism was cultured from the throat; PPD was negative.

It was concluded that the patient had Reye's syndrome, and corticosteroid therapy (Prednisolone 2.5 mg/kg I.V.) was initiated. His seizures were kept under control with phenobarbitol and diazepam and by keeping his temperature below 36°. The boy's state never improved and he died four days after admission. Post mortem examination of the liver, kidney and brain confirmed the diagnosis; the liver was found to be enlarged and pale.

Comments

This is a clinicopathological entity which has been described under such different headings as Reye's syndrome, White Liver disease,[4] encephalitis syndrome with fatty degeneration of the viscera,[5] etc. Usually two to three days, but sometimes up to six, after a mild upper respiratory tract infection or gastro-intestinal prodromata, or both, the child presents sudden onset of pallor, convulsions, coma, acidosis, NPN elevation and electrolyte disturbance. Different viruses and toxic etiologic agents have been implicated,

and in one case varicella was the most likely etiologic factor.[6] Patients are not often pyrexial as in our case, and do not necessarily have enlarged livers. Death or apparent improvement may occur within 36 to 72 hours after the complete clinical picture has been established, but the mortality rate is more than 80 per cent. Jaundice is not a part of the syndrome, and normal CSF findings separate it from infectious encephalitis

Since these patients have elevated transaminase values as well as blood ammonia, the encephalopathy finding might be explained by hepatocellular dysfunction. But the fatty degeneration of the liver cell, renal distal tubules and the cerebral blood vessels are the morphologic findings differentiating Reye's syndrome from acute hepatic coma in which electrolyte disturbance is generally found. The blood sugar is often low in these cases and there is no notable response to intravenous administration of glucose.

The history, clinical findings and laboratory data of our patient all favored Reye's syndrome, and for that reason corticosteroid and intravenous glucose treatment was started following his admission. Exchange transfusion was considered the last night, but the patient died before this could be done.

Summary

A case of Reye's syndrome, diagnosed clinicall y and confirmed by macroscopic and microscopic examination, is presented.

REFERENCES

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