Correspondence

Enigmatic death of an infant

Sir,

The report of Hirschberger and Kleinberg (Archives, 1976, 51, 977) was stimulating; it is always a challenge to read about enigmatic cases. The authors describe an infant who failed to thrive and died at the age of 4½ months; on repeated occasions they found high urinary excretion of homovanillic acid (HVA) and vanillylmandelic acid (VMA). On microscopical examination of necropsy material the brain, lung, thyroid, liver, adrenal glands, and intestinal tract were normal. From their data (HVA and VMA, opsoclonus on one occasion, 'abnormal vertical eye movements'), even in the absence of a detailed report of neurological findings, we would like to suggest a diagnosis of myoclonic encephalopathy of infants (Kinsbourne, 1962).

Although failure to thrive, without accompanying diarrhoea, is not a usual symptom of myoclonic encephalopathy of infants, this could represent an age-specific manifestation. In their report the authors do not mention microscopical examination of the neural crest; therefore a possibly very small tumour (ganglioneuroblastoma) was not completely ruled out. However, even if the authors would agree with our diagnosis, there remains another enigma: as to the relationship between the hormones (HVA, VMA) and the clinical symptomatology—in this as well as in other patients. Nevertheless, in a future similar case, a therapeutic trial with ACTH would seem to be justified.

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Dr. Kleinberg comments:

We are grateful for the reference by Drs. Kopp and Burck to myoclonic encephalopathy, but hasten to point out that there are significant differences between our case and those presented in that report (Kinsbourne, 1962). Our case did not have myoclonic movements. There were occasional episodes of opsiphotonic posturing and opsoclonus of the eyes, but myoclonus was simply not a feature of the neurological exam of our infant. Secondly, the youngest infant in Kinsbourne's series was 6 months old and almost all were considerably older. None presented with failure to thrive and none died. Lastly, of course, we are concerned about raised HVA and VMA levels which are not reported in Kinsbourne's paper.

Reference