Letter to the Editor (Viewer's Choice)

A RARE CAUSE OF FAILURE TO THRIVE

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A female infant was born to healthy unrelated parents following a normal pregnancy. Birth weight was 4100g (91st centile). She was breast fed from birth and thrived initially, regaining her birth weight by 2 weeks of age and increasing to 4400g by 7 weeks of age (50th centile). Concerns were first raised at 13 weeks when she had dropped to the 2nd centile. She was an alert and happy baby, although thin. High calorie formula milk top ups by bottle and syringe were started along with weekly multidisciplinary outpatient reviews. By week 23, she weighed 4470g (<0.4th centile), with length on the 50th centile and head circumference just above the 25th centile. Despite her thin appearance she did not cry for feeds and was increasingly aversive towards bottles. Developmentally she was age appropriate, and was noticeably bright and visually alert. She was admitted for nasogastric (NG) feeding, empirical ranitidine, domperidone and vitamin supplementation. She was given 120% of her calculated requirements. Blood tests, barium swallow and follow-through and pH probe study were all grossly normal, although a growth hormone level was at the upper end of normal (18.9 micromol/L). By week 25, after 2 weeks of NG feeding with minimal weight gain to 4650g, small amounts of pureed high calorie solid foods were added in. Subsequently, the milk was changed to an elemental infant formula in case of allergy. At week 26, she developed subtle horizontal nystagmus. An MRI demonstrated a large hypothalamic lesion causing significant mass effect, but without local invasion, hydrocephalus or additional intracranial masses. This was identified as a low grade astrocytoma of the optic chiasm and hypothalamus.

The association between hypothalamic tumours and faltering growth was first recognised in 1951 (1) and termed diencephalic syndrome. Signs and symptoms of diencephalic syndrome typically appear at around 6 months. (1) They classically include severe weight loss with decreased subcutaneous adipose tissue despite normal to increased calorie intake, with maintenance of linear growth and normal or even precocious development. Hyper-alertness, hyperkinesis, vomiting and nystagmus are variably described. (2) The features of hyperemesis, hypoglycaemia, pallor or hypotension reported in early series are less consistently described in more recent case series. (3) The underlying process may involve raised growth hormone levels with partial resistance, resulting in a highly catabolic state. (3) The most common tumour associated is pilocytic astrocytoma. (2) Definitive surgical removal is often impossible; however partial resection to reduce hypothalamic compression and improve symptoms may be advocated. (4) Chemotherapy is used for low grade gliomas (5), and despite its potentially deleterious effects in young children, radiotherapy may be used. (4) In conclusion, in those who fail to thrive despite multidisciplinary intervention an investigation of possible underlying causes should be undertaken including when appropriate neurological imaging - particularly in infants who maintain linear growth and normal development.

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