Familial hyperparathyroidism

L M Sandler and M W Moncrieff

Professorial Medical Unit, Middlesex Hospital, London, and Department of Paediatrics, Radcliffe Infirmary, Oxford

Summary

A mother and her 12-year-old son both had a parathyroid adenoma. They represent a further example of dominant inheritance of isolated hyperparathyroidism.

Primary hyperparathyroidism is a rare disease in children and by 1970 only 43 cases had been reported. Familial hyperparathyroidism is also uncommon and when it occurs usually forms part of the pluriglandular syndrome. It can however be an isolated disorder, and we describe 2 such cases, a mother and her son, who each had only a single parathyroid adenoma.

Case 1

A 12-year-old boy gave a 6-week history of dyspepsia and poor appetite. Although rather thin, he looked well, but was tender in the right hypochondrium. Barium meal examination confirmed that he had a duodenal ulcer. After treatment with antacids his pain resolved and he rapidly gained weight. There was no family history of peptic ulceration. A blood sample taken at his first hospital visit showed a high serum Ca: this subsequently ranged from 2.74 to 3.0 mmol/l (10.96 to 12.0 mg/100 ml). He was given 120 mg hydrocortisone daily for 10 days and the serum Ca levels did not fall, being 2.71 mmol/l (10.8 mg/100 ml) on day 8, 2.72 mmol/l (10.9 mg/100 ml) on day 9, and 2.71 mmol/l (10.8 mg/100 ml) on day 10. The plasma phosphate, urea, albumin, alkaline phosphatase parathormone, cortisol, thyroxin, gastrin, fasting glucose, and insulin were all normal (Table). Radiologically there was no evidence of bone disease or renal calculi.

Selective venous sampling showed that the parathormone level was >10 ng/ml in the region of the inferior thyroid vein, and at surgical exploration a cystic adenoma of the left lower parathyroid gland was found within the thymus and was removed (Mr E Milroy). Normal left and right upper parathyroid glands were identified and biopsied. The right lower parathyroid was not seen. Postoperatively plasma Ca fell to normal.

Case 2

The mother, who was asymptomatic, was found to be hypercalcæmic when members of the immediate family were screened (father, mother, and brother). On direct questioning she admitted to polyuria of 4 years' duration and polydipsia for one year. She had had a thyroidectomy 18 years previously for thyrotoxicosis. Systematic examination was normal. The corrected serum Ca was 2.95 mmol/l (11.8 mg/100 ml). This failed to become normal during a standard 10-day hydrocortisone suppression test, the serum Ca being 2.90 mmol/l (11.6 mg/100 ml) on day 8, 2.87 mmol/l (11.5 mg/100 ml) on day 9, and 2.92 mmol/l (11.7 mg/100 ml) on day 10. The results of other biochemical investigations are given in the Table, and showed a low plasma phosphate, and raised parathormone level. Investigations for associated endocrine disorders were normal. Radiologically there was no evidence of bony or renal involvement.

Parathyroid venous sampling showed abnormal...
venous drainage due to previous surgery. Consistently high levels of parathormone with a maximum value of 5.2 ng/ml were found in the brachiocephalic vein, suggesting a tumour in a lower parathyroid gland. At neck exploration (Mr E Milroy) an adenoma was found in the right lower gland and was removed. Postoperative serum Ca was normal.

Histological examination of the tumour from both mother and son showed a typical parathyroid chief-cell adenoma.

Discussion

The boy is interesting in two respects. He had primary hyperparathyroidism and a peptic ulcer which in children is a rare combination. Bjernulf et al. mentioned only 2 similar cases. In addition, it is unusual for a child with hyperparathyroidism to have no bone or renal involvement.

The mother originally denied any symptoms, but early diagnosis before bony or renal complications had occurred was made when the serum Ca was measured in the child's close family. We consider that this should be done routinely in childhood hyperparathyroidism.

In neither patient was there any evidence of associated endocrine disturbance. This appears to be a further example of familial isolated hyperparathyroidism, which is rare and is said to be inherited as an autosomal dominant trait.

In familial hyperparathyroidism generalised hyperplasia of the parathyroid glands is usually found. It is interesting that both our patients had a single parathyroid adenoma, which is the usual finding in childhood.

We thank Dr J O'Riordan and Mr E Milroy for permission to publish details of these patients.

References


Correspondence to Dr M W Moncrieff, Department of Paediatrics, The John Radcliffe Hospital, Oxford OX2 6HE.

Course of rotavirus gastroenteritis in a closed community

TSUNEKO MORISHIMA, HIDEAKI YAMAGUCHI, SHOICHI NAGAYOSHI, TAKAO OZAKI, SHIN ISOMURA, AND SAKAE SUZUKI

Department of Paediatrics, Nagoya University School of Medicine, Japan

SUMMARY There were 3 outbreaks of rotavirus gastroenteritis accompanied by milky-white stools in a closed community of 21 children in Japan during a 2-year period. Several different clinical courses were observed as a result of clinical, serological, and virological observations.

There is strong evidence from many parts of the world that human rotavirus is commonly associated with acute nonbacterial gastroenteritis in infants and children. In Japan, this virus is also found in acute gastroenteritis, particularly in that form of infantile diarrhoea which is accompanied by a milky-white stool (called Hakuri in Japanese). Hakuri has long been known as a well-defined and common type of nonbacterial gastroenteritis occurring in winter. Rotavirus has been detected by electron microscopy (EM) in the faeces from as many as 90% of Hakuri patients. However, the mode of human rotavirus infection and immunity against this virus has yet to be elucidated. To clarify these points, we studied a series of outbreaks of Hakuri in a closed community during a 2-year period.

Materials and methods

The study group comprised 21 children (10 boys, 11 girls) between ages 3 and 18 months, under the care of the Shuzenki Orphanage in Nagoya. They
Familial hyperparathyroidism.

L M Sandler and M W Moncrieff

*Arch Dis Child* 1980 55: 146-147
doi: 10.1136/adc.55.2.146

Updated information and services can be found at:
http://adc.bmj.com/content/55/2/146

These include:

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/