**Letter**

**Idiopathic familial basal ganglia calcification associated with juvenile hypertension**

_SIR,—We describe a family in which two sisters with idiopathic basal ganglia calcification had severe hypertension, detected at an early age for which no cause was found. The parents (who were not consanguineous) and two brothers, did not have hypertension or basal ganglion calcification. None showed any neurological abnormality.

The younger sister was noted to have a blood pressure of 180/140 mmHg when admitted unconscious during an attack of hypertensive encephalopathy at the age of 16 years. The cerebrospinal fluid was slightly blood stained. Electroencephalogram and carotid angiogram were normal. The skull x-ray showed bilateral dense basal ganglia calcification and this prompted us to investigate the other family members of whom only the sister showed similar skull x-ray changes.

The elder sister was noted to have a blood pressure of 180/120 mmHg at the age of 13 years when admitted for typhoid fever. Both sisters had no neurological deficits and had been healthy with normal physical and mental development and no history of fits or tetany. No obvious cause for the hypertension was found in either case. Renal causes, renal artery stenosis, and coarctations of aorta were excluded. Investigations including intravenous pyelogram, aortogram, renal arteriogram, presacral air insufflation, and retrograde pyelogram were normal. Twenty-four-hour urine excretion of vanillylmandelic acid was normal and ro tinine test negative. Renal functions were normal. There was no cardiomegaly, and fundoscopy was normal. Renal biopsy done recently showed the changes, due to arteriosclerosis.

Causes for basal ganglia calcification such as hypoparathyroidism and pseudohypoparathyroidism were excluded in both sisters by several years of clinical observation and the absence of physical stigmata such as short metacarpals, cataracts and subcutaneous calcifications. Biochemical investigations showed normal calcium metabolism.

**Case 1:** Serum calcium was 2.52 mmol/l; phosphate 1.5 mmol/l; alkaline phosphatase 42.5 iu/l. Twenty-four-hour urine calcium excretion was 1.01 mmol.

**Case 2:** Serum calcium was 2.27 mmol/l; phosphate 0.99 mmol/l; alkaline phosphatase 134 iu/l. Twenty-four-hour urine calcium excretion was 2.42 mmol.

Toxoplasma haemagglutination test was negative in both cases. The cases have been followed up for 14 years.

Idiopathic familial basal ganglia calcification is a rare condition. Only about 10 families have been reported. All these families had extrapyramidal symptomatology such as parkinsonism and athetosis associated with epilepsy and mental retardation, but the family described above was unaffected neurologically.

The interesting association of this condition with a peculiar severe hypertension in childhood in the same members of the family has not been described. Whether the hypertension could be related to such calcification is not known. It is possible that this represents a genetic disorder of some unknown metabolic type where hypertension and basal ganglia calcification are associated.

**References**


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**Notice**

12th World Congress of Neurology to be held 20 September–25 September 1981. Address for further information: Secretariat, 12th World Congress of Neurology, c/o Simul International, Inc., Kowa Bldg. No. 9, 1-8-10, Akasaka, Minato-ku, Tokyo 107, Japan.

**Book Review**


This substantial neuroradiological text book is subdivided into sections. After a very short preliminary account of the clinical manifestations of disease in the region and some generalisation about the radiological methods available for their diagnosis, a full chapter is devoted to normal anatomy and pathology on a topographic basis; the size and shape of the sella, anatomy and diseases of the optic canal, the ethmoid sinuses and so on. There follow six chapters, each orientated around a different category of disease, its clinical signs, radiological manifestation and investigation, pathology and treatment.

Most of these are well designed for the young general radiologist seeking a balanced introduction to a career in neuroradiology. If one were to criticise their content it would be chiefly on the account of CT investigation, but CT has advanced so fast that it is difficult for any text book to keep up with improvements in technique, interpretation and usage.

Perhaps the authors have been carried away a little too far in widening the subject matter to include virtually the whole head, and have thereby made the book rather longer than it need have been to fulfil its purpose. The sections not really concerned with the sellar region, though frequent, inevitably fall short of a comprehensive treatise and for some readers will be a duplication of effort.

The book is extensively illustrated and most of the pictures do succeed in making their point, though in my copy many of them are spoiled by the printer's ink rubbing off from one page on to another. They are rather dark and need good daylight viewing.

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