Papilloedema due to hyperparathyroidism

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Papilloedema may develop as a complication of hypoparathyroidism (Grant, 1953; Duke-Elder and Scott, 1971a), but it does not appear to have been recorded as a result of hyperactivity of the parathyroid glands. The following patient presented with unilateral papilloedema without detectable intracranial cause. She had increased bone vascularity due to secondary hyperparathyroidism. The papilloedema regressed after subtotal parathyroidectomy and healing of the bone condition, but changes in the fields of vision suggest that some degree of secondary optic atrophy has occurred.

Case report

A 19-year-old woman was referred for assessment of renal function. Oedema of the left optic disc was noted on routine examination. She had been told at the age of 11 years that she had some form of kidney trouble, but no other details are available from that time.

At the age of 19 years she had been found to have a blood urea level between 130 and 160 mg./100 ml. Urine examination showed proteinuria of approximately 100 mg./100 ml. There were 5 erythrocytes per high power field, and no growth on culture of the urine.

The highest blood pressure recorded was 120/100 mm.Hg. Serum creatinine level was 5.6 mg./100 ml., and the haemoglobin level 9.9 g./100 ml. The white cell count was normal.

A blood smear showed normochromic anaemia, and platelets appeared normal. Antinuclear factor test and LE cell preparation were negative. The patient had not complained of itching, or of ocular irritation.

On admission to hospital she had an earthy pallor; there was moderate ankle oedema, the blood pressure was 120/85 mm.Hg, the chest was clinically clear, and the abdomen normal. A neurological examination gave normal results apart from the left optic disc. This showed papilloedema, with two small haemorrhages on the disc. There were no arterial changes and no exudates. The visual acuity was normal (6/5) in both eyes. Fields of vision were normal to 2W/1000 object in both eyes, and the blind spot in the right eye was normal. The blind spot in the left eye was enlarged to approximately twice the size of that in the right eye, and the left eye also showed a small vertical crescent-shaped paracentral scotoma opposite to the blind spot. Examination with the slit lamp and hand lens confirmed the left papilloedema and the normal right optic disc. Fluorescein angiography also confirmed these findings. There was no band keratopathy or conjunctival calcification.

Repeat biochemical investigations confirmed the previous findings. In addition the serum sodium level was 144 mEq/l., potassium 4.3 mEq/l., chloride 109 mEq/l., bicarbonate 16 mEq/l., uric acid 7.2 mg/100 ml., calcium 9.7 mg./100 ml., inorganic phosphate 6.1 mg./100 ml. The serum alkaline phosphatase level was 221 i.u. at 37°C. (normal 20–54 i.u.). The total serum protein level was 7.5 g./100 ml., albumin 4.2 g./100 ml., alpha I globulin 0.2 g./100 ml., alpha 2 globulin 1.2 g./100 ml., beta globulin 0.8 g./100 ml., gamma globulin 1.1 g./100 ml. The 24-hour urinary protein was 2.3 g. The urinary excretion of lead was normal, and showed no significant increase after EDTA infusion. Random blood sugars were 102 mg. and 99 mg./100 ml. respectively. The serum protein-bound iodine level was normal.

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An infusion intravenous pyelogram showed that two kidneys were present, the right approximately 9½ cm., and the left approximately 8½ cm. in length; both showed marked impairment of function. Bilateral retrograde pyelograms showed both ureters to be free of obstruction. The micturating cystogram showed no vesico-ureteric reflux. An attempt at percutaneous renal biopsy was unsuccessful.

Chest x ray showed normal heart size and clear lung fields. The radiological changes of hyperparathyroidism were very marked in the pelvis, spine, hands, skull, ribs, and clavicles. There was no vascular or other soft tissue calcification. The paranasal sinuses and mastoid air cells were radiologically normal. Microscopical examination of the urine showed occasional leucocytes, occasional erythrocytes, no casts, and no crystals; a culture was sterile. The antinuclear factor test was again negative. The antistreptolysin titre was 250 Todd units/ml.

An electroencephalogram showed an alpha rhythm of 10 cps with a considerable amount of 4 to 5 cps activity throughout the recording. This occurred asymmetrically, with some excess over the right hemisphere posteriorly. The tracing was regarded as diffusely abnormal, and consistent with a metabolic disturbance. An echo-encephalogram showed no evidence of mid-line shift. Brain scan showed the bones to be extremely vascular, but no detectable evidence of a space-occupying lesion within the brain. The bone vascularity was noted to be as marked as in some cases of Paget's disease of bone. A left carotid arteriogram again showed changes of hyperparathyroidism in the bones, but the vascular anatomy was normal. Tomograms of the optic foramina were normal. Lumbar puncture revealed cerebrospinal fluid pressure to be normal; the fluid contained 3 leucocytes/cmm., and had a protein level of 29 mg./100 ml., sugar 52 mg./100 ml., and chloride 118 mEq/l. Colloidal gold, Kolmer, and VDRL tests were negative. An air encephalogram outlined normal ventricular systems, and the basal cisternae above the pituitary were also well filled, with no evidence of a space-occupying lesion. Electrocardiogram showed normal voltages, and normal Q-T interval.

Treatment

Originally, the patient was treated conservatively, but the serum calcium rose progressively to 11.4 mg. per cent. and the serum alkaline phosphatase to 396 i.u. It was thought that the renal function was relatively stable, but that the progressive bone disease involved a risk of pathological fractures. The left-sided papilloedema remained unchanged for 9 months. It was postulated that it was due to vascular engorgement, and that the right optic disc would, in time, develop the same changes. It was also possible that, if the papilloedema persisted, it could cause secondary optic atrophy. Moreover, the rising serum calcium level suggested autonomous parathyroid activity, possibly with adenoma formation (tertiary hyperparathyroidism). For these reasons, the patient was recommended for parathyroidectomy.

Surgery

Four grossly enlarged parathyroid glands were identified and all except the lower one-quarter of the right inferior gland were removed (Dr. G. Clunie).

The combined weight of the removed glands was 3.8 g. (normal upper limits said to be 0.3 g.). All showed cellular hyperplasia with some nodularity, but without identifiable adenoma formation. Fat spaces were diminished. All cell types were included in the hyperplasia, with water-clear cells and oxyphil cells predominating. There was no cellular pleomorphism, and no mitoses were seen.

Progress

Postoperatively, there was severe and prolonged tetany, requiring repetitive intravenous injections of calcium gluconate, as well as regular oral calcium gluconate and calciferol. Injections of calcium were required for 2 months, and the total volume injected was 10,200 ml. of 10 per cent. calcium gluconate (equal to 4,590 mEq of elemental calcium, or 91.8 g. of elemental calcium). During this time there was no diarrhoea, and random 24-hour urine collections showed a maximum calcium in urine of 86 mg. per day. Presumably, although the absorption of the orally administered calcium was unknown, that which was given intravenously was rapidly deposited in bone. In support of this was a rise in the serum alkaline phosphatase level from a preoperative level of 396 i.u. to a maximum of 678 i.u. 1 week

postoperatively, and a fall in the serum inorganic phosphate level from a preoperative level of 6.9 mg./100 ml. to a minimum of 1.3 mg./100 ml., and a fall in the blood urea level to a minimum of 48 mg./100 ml. without a corresponding fall in the serum creatinine. Oral phosphate supplements were given, but the serum inorganic phosphate was still below normal 2 months after the operation, although by this time the blood urea level had returned to 157 mg./100 ml., and the serum alkaline phosphatase level had fallen gradually to 75 i.u. Some recalcification of the terminal expansions of the distal phalanges was apparent radiologically after 1 month, but residual radiological signs of hyperparathyroid bone disease were still present 6 months after the operation, and the lamina dura of the teeth was not detectable until 12 months postoperatively. Supplements of calcium and vitamin D were stopped at this time. The bone disease was therefore considered to have been very severe.

Ocular findings

The oedema of the left optic disc receded gradually, and by 3 months after parathyroidectomy the disc was flat with increased gliosis and absent physiological cup, results of the previous papilloedema (Figure 1); 2 years after surgery, the right eye was still normal. The left eye still showed the increased gliosis of the disc, but the field of vision of this eye was concentrically contracted, suggesting that some degree of optic atrophy had occurred, but was masked by the gliosis.

FIGURE *Left optic disc 3 months after subtotal parathyroidectomy. Gliosis after the papilloedema has filled in the physiological cup.*

Discussion

This patient presented with the clinical sign of unilateral papilloedema, with confirmation from the fields of vision and fluorescein angiography. She had no evidence of intracranial disease or of arterial hypertension, and the ocular condition remained unchanged until bone changes were reversed by subtotal parathyroidectomy. The papilloedema is therefore attributed to the secondary hyperparathyroidism. The mechanism is postulated to be increased orbital vascularity, associated with marked bone hyperaemia, as noted at the time of attempted brain scan. This type of orbital vascular engorgement has been thought to explain papilloedema in intracranial arterio-venous aneurysm, cavernous angioma of the orbit, and even in general increase in intracranial pressure (through shunting of carotid blood into orbital vessels) (Walsh and Hoyt, 1969). The fact that the papilloedema was unilateral also favours a local orbital cause (Duke-Elder and Scott, 1971b).

Summary

A case of unilateral papilloedema attributed to secondary hyperparathyroidism is reported. No other cause for the papilloedema was found, and it regressed after subtotal parathyroidectomy. The postulated mechanism of the papilloedema is increased orbital vascularity due to marked hyperaemia of the bones.

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