CHRONIC IDIOPATHIC HYPOPARATHYROIDISM ASSOCIATED WITH MONILIASIS

By

MURIEL M. McLEAN

From the Royal Aberdeen Hospital for Sick Children

(RECEIVED FOR PUBLICATION MAY 12, 1954)

Idiopathic hypoparathyroidism is probably less rare than is commonly thought. Steinberg and Waldron (1952) in a comprehensive review of the literature collected 51 cases and added one of their own, but they did not include several reported cases whose serum inorganic phosphorus did not exceed 5.0 mg. %. Few reports have come from this country. Humphreys (1939), Himsworth and Maizels (1940), Jordan and Kelsall (1951) each report one case, and de Mowbray, Llewellyn Smith and Symonds (1954) add three more. In Steinberg’s 52 collected cases symptoms had appeared by the age of 12 years in 50%, but symptoms frequently persisted for several years before the diagnosis was made. Convulsions occurred in more than 50% of the cases, and an earlier diagnosis might be made if idiopathic hypoparathyroidism were considered in all children presenting with convulsions.

In 10 reported cases fungus infections have been found along with parathyroid deficiency. Thorpe and Handley (1929), Sevringhaus and St. John (1943), Talbot, Butler and MacLachlan (1943), Gotta and Odoriz (1948), Collins-Williams (1950) all report one case, and Sutphin, Albright and McCune (1943) add five cases, three in siblings.

In the case reported here chronic parathyroid insufficiency was associated with chronic moniliasis.

Case Report

In August, 1953, a girl aged 11 years was admitted to hospital for investigation of convulsions. She was an only child. Her mother was healthy but her father had been killed shortly before her birth. In 1946, at the age of 4½ years, she had an acute illness with pains in the legs and arms. She was admitted to an infectious diseases hospital as a case of poliomyelitis. On examination she had some carpopedal spasm which passed off rapidly. Trousseau’s and Chvostek’s signs were not present. The serum calcium level was 7.8 mg. %. She had a thrush infection of the mouth. There was no evidence of poliomyelitis and the final diagnosis was alkalosis.

Soon after discharge from hospital she developed ptosis of both eyelids. Photographs taken before her first admission to hospital show that she had no ptosis then. About March, 1947, the nails of the right hand became rigid and thickened, and cracks appeared at the angles of the mouth. In January, 1951, the nails of the left hand and of the toes became similarly affected. In May, 1951, she had three typical grand mal fits over a period of two days. In June, 1951, she was re-admitted to hospital for investigation of these fits.

FIG. 1A.

FIG. 1B.

FIGS. 1A AND B.—Photographs showing ridging and cracking of finger and toe nails.
On examination she was a well built, well nourished girl with ptosis of both eyelids. The angles of the mouth were cracked and the mucous membrane was heaped up into plaques, the nails were ridged and brittle, and the thumb nail beds were inflamed. No other abnormalities were noted. The cerebrospinal fluid was normal, and the blood Wassermann reaction was negative. A radiograph of the skull showed a small area of calcification in the left cerebrum in the region of the basal ganglia, but the significance of this was not appreciated. The lesions of the nails and mouth were ascribed to a metabolic upset, and she was discharged on treatment with a multi-vitamin preparation.

She remained well until June, 1953, when she had two convulsions during dental extraction. These were accompanied by carpopedal spasm.

In August, 1953, she was again admitted to hospital for investigation.

She was a tall, well nourished girl. Her skin was smooth and her hair was dry but not scanty. The finger and toe nails were ridged and broken (Fig. 1), and the mucous membrane at the angles of the mouth was cracked and thickened (Fig. 2). She had already lost all four six-year molars, and the lower incisors were carious, but radiographs showed that the roots of erupted and unerupted teeth were all normal. Ptosis of the eyelids was still present. Chvostek's and Trousseau's signs were present on most occasions but at times negative responses were elicited. Urine examination gave specific gravity readings up to 1028, no abnormal substances were found in any specimen, but Sulkowitch's test for calcium was repeatedly negative. A swab from the angles of the mouth grew Candida albicans. Serum calcium was 6.7 mg. % and serum phosphate 7.6 mg. %. A radiograph of the skull now showed bilateral symmetrical calcification in the region of the basal ganglia (Fig. 3). Radiographs of the long bones showed no abnormality. An electrocardiogram showed a prolonged Q-T interval of 0.36 second at a heart rate of 100 per minute. The Ellsworth Howard test gave a marked rise in the urinary excretion of phosphate following the intravenous injection of parathyrome (Fig. 4). On the history of chronic tetany and convulsions, the low serum calcium, high serum phosphate readings, the absence of renal insufficiency, the normal bones found in radiographs and the positive response to the Ellsworth Howard test, the diagnosis of chronic idiopathic hypoparathyroidism was made.

Treatment was started with A.T.10 (dihydrotachysterol). Two doses of 0.5 ml. were given with a two-day interval between doses, and the dose was then increased to 1.0 ml. every second day. After four days on treatment small amounts of calcium began to appear in the urine. After three weeks' treatment A.T.10 was discontinued and treatment with 25,000 units of 'calciferol' daily, and 60 grains of calcium lactate three times daily, was started. On this treatment she continued to excrete calcium, the serum calcium level rose to 8.18 mg. %, but the phosphate still remained high at 8.6 mg. %.

Within a few weeks of starting treatment the child became much brighter and very talkative, whereas previously she had been very quiet and a little morose. On treatment with vitamin D and calcium by mouth, she remains well and continues to excrete calcium in the urine.
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Comment

The frequency with which monilial infection has been reported in idiopathic hypoparathyroidism indicates that it is more than a chance finding. It has not been reported in parathyroid deficiency following thyroparathyroidism, and in some cases has preceded the appearance of symptoms of hypoparathyroidism.

It seems unlikely that the fungus infection is responsible for depression of parathyroid activity, but there may be some factor which depresses the parathyroids and also encourages fungus infections.

The occurrence of ptosis following the first episode of tetany in this child has not been explained. The changes in the nails may be due to monilial infection, or to lack of calcium, but there is no doubt that the mouth lesions have been due to chronic moniliasis.

Summary

A case of chronic idiopathic hypoparathyroidism associated with moniliasis is reported.

REFERENCES