Case Report

Cerebral Subcortical Calcification and Hypoparathyroidism
—A Case Report and Review of the Literature—

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Hypoparathyroidism is known to exhibit various neurological manifestations, but most of the neurological disorders seen in this condition are motor ones, and involuntary movement is rarely encountered. Basal ganglion calcification is a common feature in hypoparathyroidism. But calcification of the cerebral cortex is, to our knowledge, extremely rare with only nine previously documented cases reported. We report a case of both involuntary movement and extensive cortical calcification with some reference to the literature. Furthermore, we discuss the relationship between neurological disorders and intracranial calcification.

Key Words: Hypoparathyroidism, Extrapyramidal motor disorder, Cerebral subcortical calcification, Ellsworth-Howard test

INTRODUCTION

All the clinical manifestations of hypoparathyroidism may be summarized as neurologic ones, except for ectodermal changes and superficial candidiasis. The most common neurologic features in this condition are said to be various motor disorders, such as tetany, convulsion or muscle cramp, but involuntary movement such as parkinsonism or chorea is rarely encountered.

Basal ganglion calcification is recognized as a common manifestation of hypoparathyroidism, but calcification of the cerebral cortex is extremely rare.

This paper describes a case of idiopathic hypoparathyroidism with involuntary movement of the head, syncopal attack and marked calcification of the cerebral cortex.

CASE REPORT

A 69-year-old woman was admitted to our hospital in June 1983 for evaluation of frequent syncopal attacks and involuntary movement of the head. She was well until six years earlier, when involuntary movement of the head was first noted. Three years before admission, syncopal attacks started to occur several times a day. There was a history of cataract surgery at the age of 41, but she had never had an operation on her thyroid gland.

Physical examination showed a woman apparently in good health with normal features and no deformities. Neurological examination was normal except for positive Trousseau's sign and involuntary movement of the head. Psychological tests showed almost normal intelligence (WAIS IQ=82). Laboratory studies (Table 1) gave a low serum calcium level (5.8 mg/dl) and a high serum phosphate level (5.9 mg/dl). The PTH level was
Table 1. Laboratory findings

| CBC: WBC 5400/μl (St 1%, Seg 53%, Lym 43%, Mo 1%, Eo 2%, Ba 0%), RBC 357 x 10^6/μl, Hb 11.2 g/dl, Ht 33.9%, platelet 29.9 x 10^4/μl |
| CRP(−), ESR 57 mm/hr, RA(−), Wa-Rose(−), LE test(−) |
| Blood chemistry: TP 6.83 g/dl (Alb 59.1%, α₁-gld 3.3%, α₂-gld 11.9%, β-gld 9.4%, γ-gld 16.3%) GOT 14KU, GPT 9KU, Bil-T 0.56 mg/dl, Alp 3.0BLU, LDH 567WU, BUN 28.1 mg/dl, UA 4.5 mg/dl, CRN 1.2 mg/dl, Tl 589 mg/dl, T-chol 220 mg/dl, TG 135 mg/dl, β-L 470 mg/dl, NEFA 543 mg/dl, PL 182 mg/dl, Na 146.8 mEq/l, K 3.2 mEq/l, Cl 107.3 mEq/l, Ca 5.8 mg/dl, Pi 5.9 mg/dl, Mg 2.2 mg/dl |
| Parathyroid function: PTH 0.21 ng/ml, U-Ca 1.7 mg/dl, U-Pi 16.0 mg/dl, %TRP 99.1% |
| Thyroid function: T3U 28.7%, T3 84 ng/dl, T4 10.6 μg/dl |
| Renal function: Urinalysis; SG 1.020, pH 6.0, prot(±), sug(−) RBC numerous/HPF, WBC 1-3/HPF, Ccr 39.1 ml/min, PSP 15' 16%, total 44.6% |

Fig. 1. Ellsworth-Howard test showing marked increase in both urinary phosphate and cyclic-AMP after intravenous injection of 100U of 1-34 PTH.

as low as 0.21 ng/ml, but the thyroid function test was normal. An Ellsworth-Howard test (Fig. 1) showed marked increases in both urinary phosphate and cyclic-AMP after intravenous injection of 100U of 1-34 PTH.

These results indicated that the disease was in agreement with the criteria of idiopathic hypoparathyroidism. ECG showed a prolonged QT interval, which was consistent with hypocalcemia. EEG was almost normal. Roentgenograms of the hands and feet were normal. Skull X-ray film (Fig. 2) showed no calcification in the brain, but cranial CT scan (Fig. 3) disclosed marked bilateral calcification of the basal ganglion and dentate nuclei. In addition, extensive cortical calcifications of both frontal and parietal lobes were noted.

The patient was treated with a combination of dried aluminium hydroxide gel 3 g/day, calcium gluconate 2 g/day and la-OH-D₃ 1 μg/day. Consequently, the serum calcium level gradually increased and almost complete remission of syncopal attacks was achieved, but involuntary movement of the head has been continuing.

DISCUSSION

Idiopathic hypoparathyroidism is characterized by various neurological manifestations, but the majority of neurological manifestations seen in hypoparathyroid states are motor disorders such as tetany, convulsion, muscle cramp, carpopedal spasm and laryngospasm. Extrapyramidal motor disorders such as parkinsonism, chorea and ballism are rarely seen. McKinney¹ reported 18 cases of hypoparathyroidism accompanied by extrapyramidal motor disorders such as chorea, dystonia, athetosis and torticollis in 1962.

Basal ganglion calcification is commonly seen in hypoparathyroid states; its incidence is 28% in cases of idiopathic hypoparathyroidism and 48% in cases of pseudohypoparathyroidism according to Bronsky et al.² But the true incidence would be much higher as revealed since the introduction of cranial CT scan, which allows easy detection. The causal relationship between hypoparathyroidism and calcification in the basal...
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Fig. 2. Skull X-ray film showing no calcification in the brain.

ganglia has been well established, but no patho-
genetic basis has been offered for this relationship. Further, calcification of the cerebral cortex is extremely rare, and only nine cases had been reported up to 1983. Analysis of these nine cases3-9 showed that cortical calcification occurred in conjunction with basal ganglion calcification and predominantly in the frontal lobe except for Case 43 (Table 2). The mechanism of this predilection of calcification for the frontal lobe has yet to be clarified. The combination of cortical calcification and basal ganglion calcification is also seen in our case.

Decreased intelligence is another one of the most significant features in hypoparathyroid states; it was reported in five of the nine patients cited above. As calcification in the basal ganglion causes various extrapyramidal motor disturbances, so calcification in the cerebral cortex, especially in the frontal lobe, may give rise to decreased intelligence. This idea is fascinating, but there are many cases of hypoparathyroidism showing oligophrenia without any intracerebral calcification. Thus, whether or not calcification in the frontal lobe causes oligophrenia is still in question.

Table 2. Distribution of cortical calcification in the recorded cases of hypoparathyroidism

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Authors and year</th>
<th>Diagnosis</th>
<th>Intracranial calcification</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>basal frontal other ganglia lobes cortical</td>
</tr>
<tr>
<td>1.</td>
<td>Eaton and Holness (1939)</td>
<td>IHP</td>
<td>+ + -</td>
</tr>
<tr>
<td>2.</td>
<td>Eaton and Holness (1939)</td>
<td>IHP</td>
<td>+ + +</td>
</tr>
<tr>
<td>3.</td>
<td>Schupbach and Couvoisier (1949)</td>
<td>IHP</td>
<td>+ + +</td>
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<tr>
<td>4.</td>
<td>Sekura et al. (1968)</td>
<td>IHP</td>
<td>+ ? +</td>
</tr>
<tr>
<td>5.</td>
<td>Lewin et al. (1978)</td>
<td>IHP</td>
<td>+ + -</td>
</tr>
<tr>
<td>6.</td>
<td>Savettieri et al. (1981)</td>
<td>PHP</td>
<td>+ + -</td>
</tr>
<tr>
<td>7.</td>
<td>Litvin et al. (1981)</td>
<td>PHP</td>
<td>+ + -</td>
</tr>
<tr>
<td>8.</td>
<td>Bloom et al. (1983)</td>
<td>PHP</td>
<td>+ + -</td>
</tr>
<tr>
<td>9.</td>
<td>Present case</td>
<td>PHP</td>
<td>+ + -</td>
</tr>
</tbody>
</table>

In our case, the IQ level was considered within the normal limits.

Our case disclosed both extrapyramidal motor disorder and marked intracranial calcification, but this combination is relatively rare and only 51 cases have been reported by Muenter10 in 1968 in a review of the literature up to that time. To our knowledge, no additional report has been

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Fig. 3. Cranial CT scan showing marked bilateral calcification of the basal ganglion, dentate nuclei and extensive calcification of both frontal and parietal lobes. seen since then.

The relationship among extrapyramidal motor disturbance, syncopal attack and intracranial calcification is obscure, but it is assumed that these symptoms are caused by circulatory insufficiency due to calcification or abnormal excitation of basal ganglion cells due to hypocalcemia. However, there are many cases presenting involuntary movement without basal ganglion calcification according to McKinney.1)

In our case, there is no appearance of recurrent syncopal attack in accordance with the elevation of serum calcium level, so syncopal attack seemed to be caused by hypocalcemia. In contrast, involuntary movement of the head may have been caused by irreversible calcification in the basal ganglia.

The following two points are emphasized:
1) Our patient manifested a combination of cerebral calcification and extrapyramidal motor disorders.
2) Our case is the tenth one of hypoparathyroidism presenting marked calcification in the cerebral cortex.

REFERENCES