Successful Pregnancy and Delivery in a Patient with Neurogenic Orthostatic Hypotension and Achalasia: A Case Report

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A successful pregnancy and delivery in a patient with neurogenic orthostatic hypotension and a past medical history of achalasia is described. Surprisingly, she was free from orthostatic hypotension during the last trimester, though the evaluation of adrenergic function by measurements of changes in plasma norepinephrine on standing and by the response of blood pressure to infused norepinephrine revealed no difference between function before pregnancy and that during the last trimester. Orthostatic hypotension started again just after delivery.

Key words: Acute pandysautonomia, Adrenergic function, Plasma norepinephrine

Neurogenic orthostatic hypotension (NOH) is a heterogeneous syndrome characterized by various autonomic dysfunctions such as impaired sweating, diarrhea, impotence, and so on (1). There is only one report of successful pregnancies in two patients with NOH, i.e., Riley-Day syndrome (2). We present a case of a woman with NOH and a past medical history of achalasia who was free from orthostatic hypotension (OH) during the last trimester of pregnancy and delivered successfully.

CASE REPORT

When the patient was seven years old, she was hospitalized after a 4-day history of high fever and abdominal pain. A diagnosis of appendicitis was made and an appendectomy was performed; pathologic examination showed that the appendix was normal. The patient vomited for twenty days after the appendectomy, and was thus transferred to another hospital. The diagnosis was, however, still unknown. She had a severe loss in body weight, urinary and fecal incontinence, and decreased sweating. Therefore she was taken to Osaka University Hospital. At that time she had severe malnutrition with a loss of hair. After the diagnosis of achalasia was made, a Heller type operation was performed. She recovered gradually, and was discharged two and half years later with orthostatic dizziness. In spite of the recurrent episodes of fever while walking outdoors during the summer and orthostatic dizziness, she did not have any serious problems in leading a student life and she held a job at the age of twenty-three. When she was twenty-four years old, she visited us with the complaint of various autonomic symptoms: she felt faint when standing, had chronic diarrhea, and sweating restricted to the right thigh, the hip, the left neck, and the left arm. She was 148 cm tall and weighed 37 kg. The physical examinations at that time were as follows: The pupils were anisocoric with no reaction to light. Instillation of 1.25% epinephrine solution into the conjunctival sac dilated the pupil. The nasal septum was perforated. She had artificial teeth. In recumbent position her blood pressure was

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Received for publication November 2, 1989; Accepted for publication May 30, 1990
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Jpn J Med Vol 29, No 4 (July, August 1990)
134/100 mmHg, but it fell to 58/0 mmHg on standing. There was no pathologic reflex, and no disturbance in the sensory nervous system, cerebellar function, and bladder function. No abnormal laboratory finding was observed except anemia. She got married at the age of thirty, and was pregnant at the age of thirty-one. She had no orthostatic dizziness or diarrhea during the last trimester. The blood pressure did not fall when standing. She had an uncomplicated vaginal delivery of a healthy full-term baby. However orthostatic hypotension and diarrhea started again after delivery.

METHODS

The orthostatic test and the graded (±) norepinephrine (NE) infusion test were performed as described previously (3, 4). Plasma NE and urinary metabolites of catecholamine were measured respectively by a modified procedure described elsewhere (5, 6).

RESULTS AND DISCUSSION

The decreased urinary excretion of NE and normetanephrine (Table 1), the absence of NE increase in plasma when standing with a low level of NE in plasma in the recumbent position (Fig. 1), and the hypersensitive response to the instillation of 1.25% epinephrine solution and to the graded (±) NE infusion (Fig. 2) indicated that the adrenergic deficit in this patient was in the postganglionic neurons (7, 8). These data combined with the clinical picture suggested two possible diagnoses for this NOH. One possible diagnosis is progressive autonomic failure (PAF) without associated neurologic disorders as classified by Bannister (9). Although all adrenergic dysfunctions in this patient were compatible with PAF, the age of onset and the absence of a progression in autonomic failure do not support the diagnosis of PAF in this patient. The other possible diagnosis, acute pandysautonomia (APD) (10), explains the clinical findings described above including the recovery of urinary and bowel control but not the asymmetrical disturbance in

Table 1. Urinary excretion of free catecholamine and metabolites (µg/gm creatinine).

<table>
<thead>
<tr>
<th></th>
<th>Patient</th>
<th>Controls (mean ± SD)</th>
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</thead>
<tbody>
<tr>
<td>Dopamine</td>
<td>163</td>
<td>171 ± 28</td>
</tr>
<tr>
<td>Norepinephrine</td>
<td>3.6</td>
<td>22.2 ± 7.2</td>
</tr>
<tr>
<td>Epinephrine</td>
<td>3.1</td>
<td>5.6 ± 3.8</td>
</tr>
<tr>
<td>Normetanephrine</td>
<td>4.4</td>
<td>12.7 ± 5.3</td>
</tr>
<tr>
<td>Metanephrine</td>
<td>30.8</td>
<td>22.5 ± 9.0</td>
</tr>
</tbody>
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Fig. 1. Changes in pulse rate, systolic blood pressure, and plasma norepinephrine on standing. •--• = before pregnancy, ○--○ = in the 10 month of pregnancy, ☐ = normal range.

Fig. 2. Effects on systolic blood pressure of graded intravenous doses of (±) norepinephrine (from 1.0 to 2.0 µg/min). Symbols as in Fig. 1.
sweating (usually symmetrical in APD), which might be explainable if the recovery of the autonomic nerves was incomplete due to the severe malnutrition that resulted in the artificial teeth and the perforation of nasal septum. Although none of the 26 patients hitherto reported (11) had achalasia, one can speculate that an infectious etiology might explain the occurrence of both APD and achalasia (12, 13). In our opinion APD is a more probable diagnosis than PAF.

The previous report of two successful pregnancies in patients with Riley-Day syndrome did not discuss the OH status during the pregnancies (2). In our patient the lack of a difference in plasma NE response in orthostatic test (Fig. 1) and the difference in pressor response to the graded (+) NE infusion (Fig. 2) before pregnancy and during the last trimester does not help to explain the remarkable observation that the OH was abated during the last trimester. There is no clear explanation for this occurrence. There are two possibilities however. When the uterus is full, problems in return flow from lower extremities as a consequence of compression of the vena cava inferior occur; these have a deleterious influence on orthostatic circulation in the normal late pregnancy (14, 15). On the other hand, in the late pregnancy of patients with NOH who are not able to constrict the blood vessels immediately after standing, a full uterus might block the flow of blood downward, and, as a result, blood pooling in the lower extremities might diminish. If this effect overcame the disadvantage mentioned above in normal pregnancy the observation in our patient might be explained. The other possibility is that some humoral factor synthesized in the placenta improved the OH and diarrhea by modulating not only blood pressure on standing but also bowel movements.

ACKNOWLEDGEMENT: We are grateful to Prof. J. Miller, University of British Columbia, for critically reading the manuscript.

REFERENCES