Lymphoid Hypophysitis Associated with Sudden Maternal Death: Report of a Case and Review of the Literature

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A case of lymphoid hypophysitis in a woman who died during labor is presented. From a review of the 14 previously reported cases it is apparent that this is a specific disease entity, that it involves only woman, especially in association with pregnancy, and that it may have a fatal outcome. The clinical manifestations are either related to hypopituitarism, or those of a space occupying lesion.

Attention drawn to the possible occurrence of the disease may result in earlier recognition of the disease and a better outcome of this potentially fatal disease.

Lymphoid hypophysitis is a rare disease. To the best of our knowledge, only 14 cases have been reported in the literature (1, 2, 4–8, 10–14) all the patients were women and in most of them (11 of 14) the disease was related to pregnancy.

The aim of this report is to present a 29-year-old woman who died suddenly during labor. Autopsy revealed the finding of lymphoid adenohypophysitis, which may be presumed to have been the cause of death.

In view of the paucity of similar reported cases, this rare occurrence is presented and the world literature is reviewed.

Case Report

A 29-year-old primigravida was admitted to the delivery ward in early labor, at term. Past medical history was uneventful, and the pregnancy was uncomplicated.

Immediately upon admission the patient complained of dizziness and blurred vision; soon thereafter she became disoriented. Blood pressure was 150/90 mm Hg, pulse rate 60 beats per minute, regular. Several minutes later the peripheral pulse disappeared and blood pressure dropped to 90/70. Involuntary movements of the lower extremities appeared and the patient went into deep coma. She was intubated and respirated, but in spite of vigorous resuscitative efforts the patient expired several minutes later of circulatory failure. The baby could not be saved.

All laboratory values including blood glucose and electrolytes taken on admission were within normal limits.

Postmortem examination: The body was of a well-nourished young woman. The uterus was intact and contained a well developed female baby and placenta.

The heart, liver and spleen were enlarged, weighing 300, 2400, and 560 gm, respectively. The brain appeared edematous and weighed 1300 gm. The enlarged pituitary weighed 5 gm, the thyroid—40 gm and both adrenals—10 gm. All the organs were macroscopically normal.

Histological examination of the liver, spleen, kidneys, and brain showed only congestion. A small number of trophoblastic cells were found in the lungs, but no amniotic fluid emboli were detected. The thyroid showed two foci of lymphocytic aggregates in one of three examined sections (Fig. 1). The adrenals as well as the parathyroids were normal.

The most striking histological lesion was found in
the pituitary. There was extensive cellular infiltration in the adenohypophysis. The infiltrate consisted mainly of lymphocytes, accompanied by some plasma cells. Neither granulomas, nor giant cells or bacteria could be seen. Furthermore, most of the acini in the infiltrated areas were destroyed. Foci of the uninvolved pituitary tissue were morphologically normal, yet some cells showed oncocytic transformation (Fig. 2). The neurohypophysis was normal.

Discussion

In the present case the significant pathologic finding was lymphoid adenohypophysitis similar to that found in 14 previously reported cases (1, 2, 4–8, 10–14). All patients were women, and in 12 the onset of symptoms was related to pregnancy, either during gestation (cases 9–12) or up to 6 months after delivery (cases 1, 3, 6–8, 13, 14) and in the present

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TABLE 1. Clinical and pathological data of 15 cases of lymphoid hypophysitis

<table>
<thead>
<tr>
<th>Case</th>
<th>Author (Ref.)</th>
<th>Year (Ref.)</th>
<th>Age</th>
<th>Onset (Weeks)</th>
<th>Related disorders</th>
<th>Glucose</th>
<th>Headaches</th>
<th>Visual disturbance</th>
<th>Anemia</th>
<th>Weakness</th>
<th>Weight loss</th>
<th>Symptom onset</th>
</tr>
</thead>
</table>
| 1    | Goude et al. (6) | 1961 | 22 | 39 | Hashimoto's thyroiditis | + | - | + | - | + | + | 2
| 2    | Halm et al. (7) | 1962 | 29 | 11 | Hypothyroidism | + | - | + | - | + | + | 2
| 3    | Lupp (8) | 1972 | 59 | 2 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 4    | Gellhorn et al. (9) | 1973 | 31 | 2 | Myasthenia gravis | + | - | + | - | + | + | 2
| 5    | Mayfield et al. (10) | 1974 | 12 | 11 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 6    | Quencer et al. (11) | 1975 | 23 | 6 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 7    | Lueck (12) | 1976 | 27 | 6 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 8    | Asa et al. (13) | 1977 | 33 | 8 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 9    | Guina et al. (14) | 1978 | 25 | 9 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 10   | Chou et al. (15) | 1979 | 28 | 10 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 11   | Asa et al. (16) | 1980 | 30 | 10 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 12   | Beissin et al. (17) | 1981 | 29 | 13 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 13   | Sobrino-Simoes et al. (18) | 1982 | 29 | 14 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2
| 14   | Gal (19) | 1983 | 30 | 28 | Lymphocytic thyroiditis | + | - | + | - | + | + | 2

* +: Symptom present.
| +: Not known or not mentioned.

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case during labor. The clinical data and the pathological findings are summarized in Table 1.

The clinical manifestations of lymphoid hypophysitis are either related to hypopituitarism or those of a space-occupying lesion; in fact signs of the latter were noted in seven patients. They included headaches, visual field loss, and blurring of vision. These patients seemed to have a better prognosis, probably because the diagnosis was made earlier in the course of the disease, partial or total hypophysectomy was performed, and replacement therapy was administered.

Some of the patients presented with weight loss, weakness, anemia, scanty menstruation, amenorrhea, and hair loss. All these symptoms were related to the decreased production of ACTH, TSH, and gonadotrophins. Others had hypoglycemic episodes, and this seemed to indicate a poorer outcome, as four out of five patients (cases 3, 5, 6, 14) died of the disease.

Some of the patients had an acute illness such as appendicitis, pneumonia, herpes labialis, flu-like illness, that preceded the appearance of adrenocortical insufficiency, circulatory failure, and collapse. These patients as well, had a grave prognosis. In our patient the labor itself may have been the triggering event for the circulatory collapse and demise.

Lymphoid hypophysitis was associated in five patients (cases 1, 2, 4, 5, 14) with other "lymphocytic lesions," namely: lymphocytic thyroiditis, Hashimoto's disease, parathyroiditis, pernicious anemia, and retroperitoneal fibrosis. The present case lymphocytic thyroiditis was present. These findings suggest that an autoimmune mechanism may be involved in the pathogenesis of lymphoid hypophysitis. Bottazzo et al. (3) found antibodies to prolactin secreting cells in patients with hypopituitarism. Experimentally a histological picture identical with lymphoid hypophysitis in women can be produced in rats by intracutaneous injection of pituitary tissue with immunological adjuvant (9). The prevalence of the disease in pregnant and puerperal women is also probably related to the immunologic disturbances in such periods.

Lymphoid hypophysitis has emerged as a specific disease entity, and must be considered in the differential diagnosis of women who present with signs of hypopituitarism, hypoglycemia or enlargement of the sella turcica, especially during pregnancy and in the postpartum period. This condition should also be added as a possible cause of death during labor and in the postpartum period.

Attention drawn to the possible occurrence of this disease may result in earlier recognition and successful treatment of this potentially fatal disease.

REFERENCES


