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Pregnancy and Stiff Person Syndrome

To the Editor:

Stiff person syndrome is an uncommon disorder characterized by fluctuating, progressive muscle stiffness, contractions, rigidity, and spasm usually involving the axial muscles.[1] Although the cause is unknown, it has been reported to be frequently associated with autoimmune conditions such as diabetes mellitus, pernicious anemia, thyroiditis, and vitiligo. Three different forms of stiff person syndrome are recognized: autoimmune, paraneoplastic, and idiopathic.[2]

The autoimmune form of the disease accounts for approximately 60% of the cases and is associated with circulating anti-glutamic acid decarboxylase antibodies.[3] These antibodies target gamma aminobutyric acidergic neurons and their nerve terminals and are detectable in the serum. The other 40% rely on clinical testing and history for diagnosis[4] because there are no consistent characteristic serologic or image abnormalities.

CASE REPORT

A woman (gravida 3 para 2) at 10 weeks' gestation presented to the triage area with several days of progressive muscle stiffness, body spasms, rigidity, and disabling axial muscle pain. The patient was known to have autoimmune stiff person syndrome with anti-glutamic acid decarboxylase antibodies and was being treated with diazepam. She had exhausted her prescription of diazepam the previous month. After her clinical evaluation, she was given 5 mg of diazepam intravenously, which promptly alleviated her symptoms. The patient was admitted to the hospital for internal medicine and neurologic evaluations. The neurology consultant recommended discontinuing diazepam and starting the skeletal muscle relaxant methocarbamol at 750 mg twice per day. She was discharged symptom-free after 1 day, only to return to triage 3 weeks later with difficulty swallowing and inability to close her right eye. Another neurology consultant diagnosed Bell's palsy and stiff person syndrome exacerbation. She was given artificial tears and prescribed gabapentin at 300 mg 3 times per day. The patient was once again discharged. She returned 6 weeks later at 19 weeks gestation with intractable facial paralysis and muscle pain. The neurology consultant prescribed diazepam again at 20 mg 3 times per day and discontinued the methocarbamol. The duration of her pregnancy was characterized by gestational diabetes but no exacerbation of her muscle spasm.

The patient returned to the hospital at 35 weeks' gestation with spontaneous rupture of membranes in active labor. Nonreassuring fetal heart tone changes occurred, and she delivered by cesarean a 9-lb infant with Apgar scores 3 and 9. The postpartum course was complicated by endometritis treated by intravenous antibiotics and muscle stiffness treated with oral diazepam. She left the hospital on postoperative day 4 in stable condition.

DISCUSSION

To our knowledge, this is the first case report to describe an autoimmune variant of stiff person syndrome with positive anti-glutamic acid decarboxylase antibodies diagnosed before pregnancy. Benzodiazepines are considered the optimal initial treatment because they are thought to modulate the levels and activity of gamma aminobutyric acid.[5] In this patient's case, discontinuing the diazepam probably contributed to her critical spasticity at 10 weeks, and restarting it at 19 weeks probably contributed to her lack of debilitating spasm through the rest of her pregnancy.

CONCLUSIONS

Other methods of treatment for the autoimmune variant of Stiff person syndrome have been described. Baclofen is a gamma aminobutyric acid-modulating drug commonly used for people with stiff person syndrome with mixed success, but it could not be applied in this patient secondary to a previous allergic reaction to this drug. Physical therapy has been shown to be helpful in some cases and may attenuate spasm. Other suggestions for

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treatment include immunosuppressive therapy, plasmapheresis, and intravenous gammaglobulin.

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References

1. Helfgott SM. Stiff-man syndrome: from the bedside to the bench. *Arthritis Rheum*. 1999;42:1312-1320.
2. Shaw PJ. Stiff-man syndrome and its variants. *Lancet*. 1999;353:86-87.
3. Solimena M, Folli F, Aparisi R, et al. Autoantibodies to GABA-ergic neurons and pancreatic beta cells in stiff-man syndrome. *N Engl J Med*. 1990;322:1555-1560.
4. Ellis TM, Atkinson MA. The clinical significance of an autoimmune response against glutamic acid decarboxylase. *Nat Med*. 1996;2:148-153.
5. Lorish TR, Thorsteinsson G, Howard FM Jr. Stiff-man syndrome updated. *Mayo Clin Proc*. 1989;4:629-636.