DERMATOMYOSITIS, SUXAMETHONIUM ACTION AND ATYPICAL PLASMACHOLINESTERASE

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EISELE¹ mentions that patients with dermatomyositis may have altered responses to relaxants. Jones and Pelton² in their index of syndromes with anaesthetic implications give four references to disorders of connective tissue. However, the authors have been unable to discover any references to the use of suxamethonium in patients with dermatomyositis. The following is a short report of such a case.

CASE HISTORY

The patient was a 19-year-old girl with signs and symptoms of active dermatomyositis for the last four years. She had characteristic skin manifestations verified by skin microscopy and immunofluorescence. There was a tendency to leucopenia and thrombocytopenia as well as a slight elevation of transaminases, LDH and CPK. She had improved on prednisone, originally 20–30 mg/day, now reduced to 2.5 mg daily. In addition she took penicillamine 250 mg each day. She had suffered repeated febrile attacks, lung complications and vasoconstriction of left upper extremity leading to gangrene of the fingers. This had been improved by thoracic sympathectomy which was performed under N₂O–O₂ – halothane anaesthesia without complications.

On June 24, 1976 she was scheduled for termination of pregnancy in the eighth week. She received thiopentone 350 mg with nitrous oxide and oxygen. Suxamethonium 60 mg was given intravenously to relax the abdomen. No fasciculations were observed and she was well relaxed during the operation. As breathing did not reappear in the expected time the ulnar nerve was stimulated electrically and a neuromuscular block was demonstrated. This lasted for about 50 minutes after the injection. Breathing was not deemed adequate until one hour and fifteen minutes after injection of the suxamethonium. Because she was receiving steroid medication, hydrocortisone 100 mg was given intravenously before and after the procedure.

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A striking feature during the recovery of the muscle tone in our patient was a series of fibrillary twitchings and fasciculations, gradually increasing in intensity. These started in the extremities and progressed to the head and trunk, lasting for at least ten minutes. This made us fear that a pyrexial reaction would ensue, but the temperature did not exceed 37.5° C and her serum CPK level was virtually unchanged 24 hours post-operatively. Examination of her plasma disclosed a cholinesterase activity of only 15 per cent of normal when acetylcholine was used as substrate. Using benzoylcholine as substrate the dibucaine number (DN) was found to be 26 and fluoride number 21.* This shows that she was a homozygote for the atypical enzyme. Her parents had DN of 63 and 66 with fluoride numbers 49 and 50. This indicates that both her parents were heterozygotes for the atypical enzyme.

An inquiry in the outpatient department of the hospital for serum samples of patients with dermatomyositis has, during eight months, resulted in serum samples from four patients. Two of these were young men and two were young women. All had typical signs of dermatomyositis. The two men had normal DN but one of the women had DN of 66 and fluoride number of 50 indicating that she was a heterozygote. She also was pregnant.

COMMENTS

The absence of fasciculations after injection of suxamethonium in our patient is of interest in view of recent discussion as to whether or not such fasciculations occur in patients with atypical plasmacholinesterase.^{3,4} The abnormal fasciculations observed during the delayed recovery of muscle tone in our patient has not been described before. It might be connected with her muscle disease. Surprisingly no pyrexial reaction and no elevation of serum CPK occurred following these fasciculations. These reactions might

*Normal values in this laboratory are: activity: 0.8 to 1.12 micromoles/ml/min of Benzoylcholine. DN, 78 to 83; FN, 56 to 69.

have been suppressed by the prednisone and hydrocortisone she had received. It is a clinical experience in patients with dermatomyositis that steroid therapy suppresses elevated serum enzyme levels.⁵

A definite connection between atypical plasma cholinesterase and genetic susceptibility to certain diseases has so far not been found. A high incidence of atypical enzyme has been reported in patients with leprosy⁶ but this could not be confirmed by another group of workers.⁷

The coincidence of atypical plasma cholinesterase in a patient with dermatomyositis is interesting because both conditions are rather rare. The incidence of homozygotes for the atypical enzyme is 1:2800.⁸ In a regional population of 3.3 millions Rose and Walton⁹ collected 89 cases of dermatomyositis, giving an incidence of 1:36000. According to the theory of statistics the chances of coincidence would be 1:100 millions. Atypical plasma cholinesterase has recently been described in Huntington's chorea,¹⁰ a condition even rarer than the disease of our patient. Both Huntington's chorea and atypical plasma cholinesterase are transmitted by autosomal genetic defects while the cause of dermatomyositis is still unknown. No explanations can be offered for the coincidence of two such rare conditions.

SUMMARY

A 19-year-old girl suffering from active dermatomyositis was given suxamethonium 60 mg during anaesthesia for termination of pregnancy. A prolonged suxamethonium action occurred which was explained by the finding of homozygous atypical plasma cholinesterase in her blood. Although no fasciculations were seen immediately after injection of the drug, a period of fasciculations progressing from the extremities to the head and trunk occurred during recovery of muscle tone. No hyperpyrexia or elevation of serum creatine phosphokinase occurred. This was ascribed to the steroid therapy she received. Plasma from four other patients suffering from dermatomyositis was also investigated and one young woman, also pregnant, was found to be heterozygous for the atypical enzyme.

RÉSUMÉ

Une jeune fille de 19 ans souffrant de dermatomyosite en activité, a présenté une apnée prolongée après une injection de 60 mg de

suxaméthonium au cours d'une intervention (interruption d'une grossesse de huit semaines). Les analyses postopératoires ont démontré qu'elle était porteuse homozygote d'une cholinestérase atypique.

Il n'y a pas eu de fasciculation après la dose de suxaméthonium, mais le retour du tonus musculaire s'est fait avec des fasciculations, d'abord aux extrémités, progressant ensuite vers la tête et le tronc. On n'a pas observé d'hyperthermie et les CPK n'ont pas été modifiées (effet des stéroïdes que recevait la malade?).

Nous avons eu l'occasion d'investiguer quatre autres malades atteints de dermatomyosite et y avons trouvé une jeune fille, également enceinte, porteuse hétérozygote de la forme atypique de la cholinestérase.

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