

MYOTONIA ACQUISITA (Talma's Disease)

Report of a Case

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Myotonia acquisita is a rare disease first described by Talma in 1892. It is characterized by an increased muscular rigidity and spasm when movement is initiated, as well as a decrease in the power of relaxation even when the muscle is at rest. The myotonic reaction is also present, namely, a normal mechanical and electrical excitability of the motor nerves but an abnormally heightened mechanical and electrical excitability of the muscle. It differs from Thomsen's disease or myotonia congenita in that it is not congenital and usually develops in adult life following trauma, acute infection, or intoxication. Also it tends to improve spontaneously or go on to complete recovery, whereas, Thomsen's disease progresses through childhood to become stationary in adult life. The onset of the disease is variable and may be characterized by weakness, pain, or flaccid paralysis.

Krabbe¹ in 1934 reviewed the literature and found 34 cases of myotonia acquisita, while Moore² added another to the series in 1935. All of these cases were in men, the ages varying from 15 to 50 years in the 29 cases where the ages were reported.

CASE REPORT

The case reported here is that of a 45 year old white housewife who came to the Clinic with the complaint of "cramping of muscles and swelling of the joints." In 1936 following a panhysterectomy for menorrhagia she developed her first symptom of cramping pain in the right calf with radiation down to the foot. This pain was not related to exercise and would often awaken the patient during the night. The pain would be so severe that the foot would become inverted and plantar flexed. Four months later she developed a severe depression for which she was confined to a sanatorium. While she was in this institution, the pains became worse and spread to the other calf and to both arms. Fibrillary twitching of the muscles was noted in 1937, but it soon disappeared even though the cramping sensations persisted.

During the year previous to admission the patient noticed swelling of various joints of the body, particularly those of the hands, shoulders, and ankles. Motion of the involved joints was painful. During this same period, the patient gained 20 pounds. She stated that she had had practically a constant "spasm" of her right calf for 4 years. She then developed a cramping sensation in the left arm and shoulder girdle, and in the muscles of her back. The right side was also involved for several years, but only during the year previous to admission did it retard the movements of that side.

Personal history revealed that the patient was married and had 4 children living and well, ranging from 11 to 24 years of age. No previous illnesses other than a "nervous breakdown" in 1937 was elicited. She had had shortness of breath on exertion as well as ankle edema for the past 4 years. Occasional attacks of paroxysmal nocturnal dyspnea had been noted during the past 10 years. The patient had always been of a nervous

temperament. There had been an exaggeration of this symptom following the surgical menopause in 1936. The past history was otherwise negative.

The general physical examination was essentially normal except for (1) an upper dental plate; (2) inflamed fauces; (3) slightly enlarged cervical lymph glands; (4) tenderness to pressure over left lower ribs; (5) pitting edema of the legs. The neurological examination was normal.

Concerning the myotonic phenomena, the right calf was in a continuous state of contraction. The grip of the right hand was the same as that of the left although it became much slower on repeated opening and closing. Striking the muscle induced a slow contraction. Reflexes were all brisk and nonpathological. Faradic and galvanic stimulation of the motor nerves was normal. She was seen by one of us (W.A.N.), and the diagnosis of myotonia acquisita was made.

Her heart was normal. Accordingly it was thought that the edema might be improved by a low sodium high potassium diet in that the edema could be due to retention of electrolytes.* The patient was started on this therapy and lost 16 pounds even though the caloric intake was not changed. However, the pain and muscle cramps increased in frequency and severity under this form of treatment.

A piece of the right gastrocnemius muscle was taken for biopsy. Sections of it showed normal striated muscle with areas of degeneration and hyalinization of muscle fibers. Some fibrosis of the intermuscular septa was present without a sign of active inflammatory reaction, parasites, or neoplasm.

SUMMARY

There were no apparent etiological factors such as infection or intoxication in this case, although the surgery she had undergone might be interpreted as the traumatic incident which occasionally precipitates this syndrome. Pain was the outstanding symptom.

There was no hypertrophy of the muscles although the myotonic syndrome was present. The patient was unable to relax the affected muscles or to initiate a rapid movement. The myotonic reaction to electrical and mechanical stimulus was observed.

Although Krabbe¹ stresses the presence of a neuritis or polyneuritis in the majority of cases of myotonia acquisita. No evidence of such a lesion was found in this patient.

The patient was older than most of the men who had developed this condition. This case is of unusual interest in that we believe it is the first of its kind reported to have occurred in a woman.

**Under the direction of Dr. E. P. McCullagh.*

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