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Cardiac findings in myotonia atrophica

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THE CARDIAC FINDINGS IN MYOTONIA ATROPHICA

by

Dee R. Mattley

A THESIS

Presented to the Faculty of

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For the Degree of Doctor of Medicine

Under the Supervision of Charles A. Hamilton, M.D.

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Introduction

My original interest in myotonia atrophica was aroused by being consulted on the pre-operative cardiac status of a patient. This patient was long diagnosed as having myotonia atrophica. The cardiac involvement had progressed to the point of being clinically detectable. This circumstance helped me realize how important it was to recognize the disease and its effects on a vital organ such as the heart.

In pursuing the subject, it was found that a large number of patients having the diagnosis of myotonia atrophica and having cardiac findings were reported in the literature. The combining factor was that all the patients in the literature had a clinical diagnosis of myotonia atrophica.

In further pursuing the subject, it was found that several patients were on file at the University of Nebraska Hospital and the Omaha Veterans Administration Hospital, Omaha, Nebraska, with the clinical diagnosis of myotonia atrophica. In presenting this subject, the important factors in diagnosing and evaluating the progress of the disease will be discussed.

. . . .

Church (11) states that in 1902, Rossolino was the first to apply myotonia atrophica to the muscular disorder characterized by myotonia and by muscular atrophy. In 1909, myotonia atrophica was defined and separated from other forms of myotonia simultaneously by Steinert, according to Cannon (9) and by Batten and Gibbs (6). In 1911, Griffith (19) reported the first association of cardiac disorders with myotonia atrophica.

The salient physical findings of myotonia atrophica listed by Hurst et al (21) are cataracts, premature frontal baldness, muscle atrophy, expressionless face, testicular atrophy, and slow muscle relaxation after contraction. Hurst (21) states that the myopathy, affecting both sexes, is a somatic dominant trait. The onset is usually late, the second and third decade. The disease is progressive in nature. Cardiac effects are primarily conduction disorders and rhythm disturbances. The heart afflicted may imitate a myocardial infarction.

The literature review covers 370 cases, all having the clinical diagnosis of myotonia atrophica. In the present series, there are six patients from the University of Nebraska Hospital, Omaha, and three patients from the Omaha Veterans Administration Hospital. All of the patients in the present series have a clinical diagnosis of myotonia atrophica.

Non-Cardiac Findings

The following non-cardiac findings will be covered: sex, race, family history, frontal baldness, testicular atrophy, neurological examination, electromyography, skeletal muscle biopsy, and chest x-ray. The findings are summarized in Table I.

Sex: The sex of the patient is the first thing to be noted on physical examination and will be presented now. In 344 (90.3 percent) cases where the sex was reported, 235 (70 percent) cases were males and 90 (30 percent) cases were females. Of the present nine patients, seven (78 percent) patients are males and two (22 percent) patients

are females. No explanation is offered for the predominance of afflicted males. Hurst (21) reports that myotonia atrophica is an autosomal dominant gene affecting both sexes.

Race: Welsh et al (41) reported 35 cases of myotonia atrophica in a Veterans Administration Hospital in which 30 percent of the hospital population was Negro. None of Welsh's cases were Negro. Kilburn et al (24) reported nine cases of myotonia atrophica in Caucasian patients from a Veterans Administration Hospital where the hospital population was 25 percent Negro. Of the 103 reported cases, four (3.9 percent) cases were Negro and 99 (96.1 percent) cases were Caucasian. In the present study, there are seven (78 percent) Caucasian patients and two (22 percent) Indian patients. No explanation has been postulated as to the apparent predilection for the Caucasian race. No reports are in the literature of myotonia atrophica occurring in the Indian race.

Family History: Myotonia atrophica being an autosomal dominant gene, it is expected that a family history should be obtained in all cases. In the 120 reported cases, 108 (90 percent) cases had a positive family history and 12 (10 percent) cases had a negative family history. The positive family histories ranged from grandparents, aunts, uncles, siblings, to offsprings. Of the present nine patients, five (55 percent) patients have a positive family history and four (45 percent) patients have a negative family history. One patient was questioned extensively but no history could be elicited. Negative family history raises the possibility of somatic gene mutations and the possibility of this being a recessive gene. McKusick (28)

states that in most or all of the cardiovascular syndromes the reason the manifestations are multiple and diverse is that the normal gene which underwent mutation determines a substance or process with widespread significance to the body's economy. Payne et al (31) discussed the possibility that myotonia atrophica was a genetically determined biochemical abnormality, because of dominant inheritance or involvement of multiple systems.

Frontal Baldness: The incidence of frontal baldness is high among patients with myotonia atrophica. Frontal baldness was reported in 106 (83 percent) cases of 128 (34.5 percent) cases reported and absent in 22 (17 percent) cases. Six (66 percent) patients in the present study had frontal baldness and three (33 percent) patients did not have frontal baldness.

Testicular Atrophy: Testicular atrophy was reported in 104 (78 percent) cases of 133 (49 percent) cases reported and absent in 29(22 percent) cases. Testicular atrophy was found in four (57 percent) patients and absent in three (43 percent) patients of the male patients in the present study. Determining the presence or absence of testicular atrophy is an arbitrary task.

Neurological Examination: The neurological examination was reported as typical in 288 (100 percent) reported cases. In the present nine (100 percent) patients, the neurological examination was typical. The patient presents with masked facies, swan neck, and weakness peripherally in the arms and legs. As the disease progresses, wasting of the peripheral muscles occurs. The hand grasp is myotonic. The patient frequently has upper eye lid ptosis.

and dysphagia. The typical neurological examination contains many variations. However, the overall examination is used in establishing the diagnosis.

Electromyogram: Electromyographic examination was typical of myotonia atrophica in 175 (98 percent) cases of 179 cases reported, and normal in four (2 percent) cases. Of the present nine patients, seven electromyographs were taken and 11 (100 percent) were typical of myotonia atrophica. Two of the nine patients were not tested. The electromyographic examination of skeletal muscle shows chains of oscillations of high potential waning slowly and giving characteristic sound in the loud speaker rather like that of a dive bomber (10). The examination must be carried out in an effected muscle to be characteristic.

Muscle Biopsy: Muscle biopsy of striated muscle was positive in 55 (95 percent) cases of the 58 (16 percent) reported cases and negative in three (5 percent) cases. In the present nine patients, five (55 percent) biopsies were taken; of these, four (80 percent) were positive and one (20 percent) was negative. Four patients in the process of being worked up did not have muscle biopsies done. The typical biopsy findings are stated by Wohlfart (43) after analyzing 25 biopsies of 18 cases of myotonia atrophica. The findings are:

- (1) The only constant histopathologic changes in myotonia atrophica is an inward migration of nuclei from the periphery of the muscle fibers to the center, where they divide and form long chains. The migrating cells lose their

immediate contact with the capillaries. Capillary loops and surrounding tissue are inclined to grow into the muscle fibers toward the nuclear cores.

- (2) In myotonia atrophica, the myofibrils are often destroyed without corresponding decrease in the sarcoplasm content. Consequently 12 of 25 biopsied muscles exhibited numerous muscle fibers with a thick peripheral layer of sarcoplasm.
- (3) The striated fibrillar rings are rather common in myotonia atrophica and were found in 14 of 25 biopsies of this material. However, they are also to be found in many muscles of advanced age and especially in the extrinsic muscles of the eye as well as in muscle atrophy from disease and in progressive muscular dystrophy. The striated fibrillar rings mentioned above are, on cross section of striated muscle, a central column of longitudinal fibrils enclosed by a spiral fibrillar fasciculi.

Chest X-Rays: Chest x-rays were abnormal in 47 (26.5 percent) cases of the 178 (48 percent) cases reported. The chest x-rays were normal in 131 (73.5 percent) cases reported. The abnormalities were 33 (70 percent) cases of cardiomegaly, four (8.5 percent) cases of right hemidiaphragm elevation, four (8.5 percent) cases of small heart, and four (8.5 percent) cases of left ventricular hypertrophy. In addition, there was one case each of left hemidiaphragm elevation and of right ventricular hypertrophy. In the present series of nine patients, eight (89 percent) patients had normal chest x-rays and one (11 percent) patient had bilateral elevation of the diaphragm. The

elevation of the diaphragms are attributed to the involvement of the diaphragms with myotonia atrophica.

Cardiac Findings

The following cardiac findings will be covered: significant cardiac symptoms, blood pressure, pulse rate, significant cardiac findings on physical examination, and electrocardiograms. The findings are summarized in Table II.

Significant Cardiac Symptoms: Cardiac symptoms were present in 67 (83 percent) cases and absent in 13 (17 percent) cases of 81 (22 percent) cases reported. Comprising the symptoms were 40 (60 percent) cases of shortness of breath, nine (13 percent) cases of palpitations, six (9 percent) cases of pedal edema, and 26 (18 percent) cases of assorted symptoms. In the present nine patients, one (11 percent) had cardiac symptoms consisting of pedal edema and hepatomegaly. Eight (89 percent) patients had no cardiac symptoms. The literature reports several cases where the cardiac symptoms preceded the diagnosis of myotonia atrophica. Kohn et al (25) reports a case in which the patient had a ten year history of progressive heart trouble consisting of recurrent atrial flutter before myotonia atrophica was diagnosed. One patient (Omaha Veterans Administration Hospital, #120-07-5354) of the present series reported hand weakness at age 25 years getting progressively worse. At the age of 50 years, he was admitted with the chief complaint of chronic lower leg ulcers. He was diagnosed as having myotonia atrophica and a right bundle branch block on electrocardiogram. The two preceding cases illustrate

the variation in the presenting symptoms of myotonia atrophica. Either the cardiac symptom or myotonia atrophica symptoms may be present first with an earlier history of one or both being elicited.

Blood Pressure: Blood pressures were reported in 121 (33 percent) cases. Eight (6.6 percent) cases had a hypertensive blood pressure of ≥ 140 mm Hg systolic or ≥ 100 mm Hg diastolic. Nineteen (15.7 percent) cases had a hypotensive blood pressure of ≤ 90 mm Hg systolic. Ninety-four (77.7 percent) cases had a normotensive blood pressure of < 140 mm Hg and > 90 mm Hg systolic; and < 100 mm Hg diastolic. In the present nine patients, nine (100 percent) patients had normotensive blood pressures (systolic pressure of < 140 and > 90 mm Hg; and a diastolic pressure of < 100 mm Hg). Accounting for part of the hypertensive blood pressures may be a complete heart block; the heart in this condition has a large stroke volume.

Pulse Rate: Of the 267 (72 percent) cases reported, bradycardia (pulse ≤ 60 beats per minute) was reported in 68 (25.5 percent) cases. Tachycardia (pulse ≥ 140 beats per minute) was reported in three (1.0 percent) cases. Normal pulse (> 60 and < 140 beats per minute) was reported in 196 (73.5 percent) cases. In the present series of nine patients, eight (89 percent) patients had a normal pulse and one (11 percent) patient had a bradycardia. The etiology for the varying pulse rate will become evident in the section on electrocardiograms.

Significant Cardiac Findings on Physical Examination: Of the 308 (83 percent) cases reported, 247 (80 percent) cases did not have any significant cardiac findings on physical examination. Sixty-one

(20 percent) cases had significant cardiac findings. These findings were systolic murmurs in 38 (62 percent) cases, distant heart sounds in 18 (29 percent) cases, cardiomegaly in six (9.8 percent) cases, and assorted findings in 11 (17 percent) cases. In the present nine patients four (44 percent) did not have any significant cardiac findings on physical examination. Five (56 percent) of the nine patients did have significant cardiac findings consisting of two (40 percent) patients having a systolic murmur and of three (60 percent) having distant heart sounds. The systolic murmurs could be on the basis of more complete filling of the ventricles during the prolonged diastolic phase. This would give rise to a functional ejection murmur due to the rapid ejection of a large volume of blood.

Electrocardiograms (Table III): Electrocardiograms were carried out on 287 (77.6 percent) cases in the literature with a total of 435 electrocardiographic abnormalities. Electrocardiograms taken on 90 (31.4 percent) cases were normal and on 197 (68.6 percent) cases were abnormal. The abnormalities by group consisted of PR interval \geq 0.20 in 75 (38.1 percent), left axis deviation in 61 (31 percent), QRS interval \geq 0.10 in 43 (21.8 percent), low P waves in 30 (15.3 percent), premature ventricular contractions in 26 (13.2 percent), left bundle branch block in 20 (10.2 percent), T wave changes in 20 (10.2 percent), atrial flutter in 18 (9.1 percent), atrial fibrillation in 17 (8.6 percent), abnormal intraventricular conduction in 14 (7.1 percent), right bundle branch block in 12 (6.1 percent), non-specific ST changes in 12 (6.1 percent), incomplete heart block in ten (5 percent), left ventricular hypertrophy in ten (5 percent),

notched QRS complex in ten (5 percent). Remaining are 57 (28.5 percent) electrocardiographic abnormalities which are varied and are small in number.

In the present nine patients, eight patients had electrocardiograms taken and all eight (100 percent) had electrocardiographic abnormalities. One patient being in the process of workup did not have an electrocardiogram. Considering the abnormalities by group, they consisted of PR interval \geq 0.20 in seven (88 percent), QRS complex \geq 0.10 in five (62 percent), premature ventricular complexes in four (50 percent), left intraventricular block in two (25 percent), non-specific ST changes in two (25 percent) and left axis deviation in two (25 percent). In addition, the following abnormalities were noted with one patient each; U waves, left bundle branch block, left ventricular hypertrophy, premature atrial complexes, and second-degree atrioventricular block with Wenckebach phenomenon. However, the small numbers are not significant. They are listed to show that in the comparisons of the present patients to the findings in the literature, the majority of findings correlate rather well.

Payne et al (31) suggested that the etiology of the electrocardiogram abnormalities may be on the basis of focal pathology of the Purkinje network, biochemical changes of the muscle proteins, or physiological alternations in the conduction properties in the cell membrane.

Early in the search for the etiology of the electrocardiographic abnormalities, Waring et al (39) stated that the type of change observed would suggest coronary sclerosis rather than a dystrophic process

involving the myocardium. Several researchers then set out to prove or disprove this hypothesis. The following are several studies conducted with the coronary arteries being a possible factor.

Fisch and Evans (17) found on autopsy of a myotonia atrophica patient that there was fibrous replacement of the myocardium in the presence of normal coronary arteries. This patient had an abnormal electrocardiogram. Orndahl et al (30) carried out exercise tolerance tests on 16 patients with myotonia atrophica. None of the 16 patients developed angina. Of these 16 cases, six were over 45 years of age with three of these having abnormal resting electrocardiograms and three having normal resting electrocardiograms. Ten patients were 40 to 45 years of age. Of these, four patients had abnormal resting electrocardiograms and six had normal resting electrocardiograms. Orndahl et al (3) concluded that the moderate atherosclerotic vessel changes usually found in the coronary arteries cannot account for the high incidence of conduction abnormalities noted in myotonia atrophica patients. Because the diffuse type of the intraventricular conduction defects encountered in these patients, focal damage of the cardiac conduction system is unlikely, whereas diffuse changes within the bundle branches and the Purkinje network may be a more adequate explanation.

Petkovich (32) reports an abnormal electrocardiogram in a myotonia atrophica patient with normal coronary arteries after examination while implanting a pacemaker. Myocardial biopsy at the same time showed muscle bundles separated by loose connective tissue in excess of normal, lymphocytes and macrophages were predominant in the stroma and the borders of the muscle fibers were indistinct.

Using age as a possible index of coronary artery sclerosis, De Wind et al (12) reports 38 (57 percent) abnormal electrocardiograms in 67 patients less than 45 years of age and ten (62 percent) abnormal electrocardiograms in 16 patients greater than 45 years of age. DeWind concluded that coronary artery sclerosis was not the etiology of the abnormal electrocardiograms in patients with myotonia atrophica.

Shearn et al (34) states that intermittent bundle branch block is usually an expression of underlying myocardial disease and probably represents a transitional stage before permanent bundle branch block supervenes. The most frequent cause of intermittent bundle branch block is coronary artery sclerosis followed by hypertensive cardiovascular disease.

Ferrington et al (15) compared electrocardiograms, vectorcardiograms, and autopsy results. Ferrington et al (15) studied 17 myotonia atrophica patients. There were 12 (70 percent) abnormal electrocardiograms and five (30 percent) normal electrocardiograms. The results of the vectorcardiograms were ten (60 percent) abnormal and seven (40 percent) normal. The vectorcardiogram was found to reflect abnormalities of the myocardium activation process more accurately than did the electrocardiogram. His observations tend to support the clinical hypothesis that suggests the presence of involvement of the myocardium similar to the involvement of skeletal muscle in myotonia atrophica. Ferrington et al (15) reviewed 17 autopsy cases in the literature and concluded that the findings of conduction defects in the electrocardiogram and of significant changes in the ventricular activation process in 60 percent of the patients

in the vectorcardiograms, would seem to be consistent with the reported autopsy findings of myocardial involvement. Hurst et al (21) states the typical autopsy findings to be diffuse myocardial fibrosis, separation of the myofibers by fibrous connective tissue, and scattered hypertrophic muscle fibers with large nuclei.

Conclusion

The first step in determining the complications of a disease is to diagnose the disease process. The data shows that the typical case of myotonia atrophica does not normally exist. The usual case presents with sufficient changes to be diagnosed as having myotonia atrophica. However, not all the changes occur in the same patient. Again, the feature that makes this collection of cardiac findings significant is that all of the patients have a clinical diagnosis of myotonia atrophica.

The high incidence in the male sex is not explained. The accepted principle of myotonia atrophica being an autosomal dominant gene does not explain the apparent predilection for males. By accepting this principle, it is expected that equal numbers would occur in males and females.

The apparent predilection for the Caucasian race will bear watching in the future as more cases are documented in the literature. The present difference is significant if it is assumed that all cases of myotonia atrophica in the non-Caucasian races have been diagnosed and reported. As noted earlier, there are no reports in the literature review of myotonia atrophica occurring in the American Indians.

Cardiac symptoms were reported to be present or absent in only a minority of cases from the literature. Significant cardiac findings on physical examination were sparse. The combination of few symptoms and normal physical examination indicates that there is slight gross change in the heart of a myotonia atrophica patient.

The electrocardiogram is the more sensitive detector of cardiac changes commonly available. Upholding this statement are the 287 (66 percent) cases from the literature which had abnormal electrocardiograms and eight (100 percent) patients in the present series having abnormal electrocardiograms. The high frequency of increased PR interval and increased QRS interval should suggest the possibility of primary myocardial involvement.

Orndahl et al (30) and others have shown that the electrocardiographic changes are not on the basis of coronary artery disease. Fearington et al (15) showed a good correlation of electrocardiograms, vectorcardiograms, and autopsy findings in patients with myotonia atrophica. Having ruled out coronary artery sclerosis as the cause of the abnormal electrocardiograms, myotonia atrophica can be considered as being the disease process responsible. It is presently accepted that the same pathological process that affects the skeletal muscle also affects the myocardial muscle. Having demonstrated myocardial changes with an abnormal electrocardiogram and myocardial biopsy in the face of normal coronary arteries supports the hypothesis that there is primary involvement of the myocardium by myotonia atrophica.

The final conclusion to be drawn is that myotonia atrophica is not going to present in a typical form and after diagnosing the syndrome, the cardiac status should be evaluated carefully. A corollary is if an electrocardiogram is obtained on an undiagnosed patient which shows an increased PR interval and an increased QRS interval; primary myocardial involvement must be entered into the differential with myotonia atrophica being among the processes considered.

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Table I Non-Cardiac Findings*

		# of Reported Cases in the Literature (% of same)	# of Patients in Present Series (% of same)
Sex	Reported	344 (90.3%)	9 (100%)
	Males	235 (70%)	7 (78%)
	Females	99 (30%)	2 (22%)
Race.	Reported	103 (27.8%)	9 (100%)
	Negro	4 (3.9%)	0
	Caucasian	99 (96.1%)	7 (78%)
	Indian	0	2 (22%)
Family History.	Reported	120 (32.5%)	9 (100%)
	Positive	108 (90%)	5 (55%)
	Negative	12 (10%)	4 (45%)
Frontal Baldness.	Reported	128 (34.5%)	9 (100%)
	Present	106 (83%)	6 (66%)
	Absent	22 (17%)	3 (33%)
Testicular Atrophy.	Reported	133 (49%)	7 (100%)
	Present	104 (78%)	4 (57%)
	Absent	29 (22%)	3 (43%)
Neurological Examination. .	Reported	288 (78%)	9 (100%)
	Typical	288 (100%)	9 (100%)
Electromyograms	Reported	179 (48%)	7 (100%)
	Typical	175 (98%)	7 (100%)
	Normal	4 (2%)	0
Skeletal Muscle Biopsy. . .	Reported	58 (16%)	5 (55%)
	Typical	55 (95%)	4 (80%)
	Normal	3 (5%)	0
Chest X-Ray	Reported	178 (48%)	9 (100%)
	Normal	131 (73.5%)	8 (89%)
	Abnormal	47 (26.5%)	1 (11%)

* References 1,2,3,4,5,6,7,8,9,11,12,13,14,15,16,17,18,19,20,22,23,24,25
26,27,29,30,31,32,33,35,36,37,38,39,40,41,42,43.

Table II	Cardiac Findings*	# of Reported Cases in the Literature (% of same)	# of Patients in Present Series (% of same)
Significant Cardiac Symptoms	Reported	81 (22%)	1 (11%)
Blood Pressure	Reported	121 (33%)	9 (100%)
	Normotensive	94 (77.7%)	9 (100%)
	Hypotensive	19 (15.7%)	0
	Hypertensive	8 (6.6%)	0
Pulse Rate	Reported	267 (72%)	9 (100%)
	Normal	196 (73.5%)	8 (89%)
	Tachycardia	3 (1%)	0
	Bradycardia	68 (25.5%)	1 (11%)
Significant Cardiac Findings on Physical Examination.	Reported	61 (20%)	5 (55%)

*References: (Same as Table I)

Table III Electrocardiogram Findings *

	# of Reported Cases in the Literature (% of same)	# of Patients in Present Series (% of same)
Total Reported	287 (77.6%)	8 (89%)
Abnormal	197 (68.6%)	8 (100%)
Normal	90 (31.4%)	0
PR \geq 0.20	75 (38.1%)	7 (88%)
Left Axis Deviation	61 (31%)	2 (25%)
QRS \geq 0.10	43 (21.8%)	5 (62%)
Low P Waves	30 (15.3%)	0
Premature Ventricular Contractions	26 (13.2%)	4 (50%)
Left Bundle Branch Block	20 (10.2%)	1 (12.5%)
T Wave Changes	20 (10.2%)	0
Atrial Flutter	18 (9.1%)	0
Atrial Fibrillation	17 (8.6%)	0
Abnormal Intraventricular Conduction	14 (7.1%)	2 (25%)
Non-Specific ST Changes	12 (6.1%)	2 (25%)
Right Bundle Branch Block	12 (6.1%)	0
Incomplete Heart Block	10 (5%)	0
Left Ventricular Hypertrophy	10 (5%)	1 (12.5%)
Notched QRS Complexes	10 (5%)	0
Assorted Other Findings	57 (28.5%)	4 (50%)

*References (Same as Table I)