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DR. FOSTER KENNEDY presided

PATHOLOGICAL FINDINGS IN THE HEART IN PROGRESSIVE MUSCULAR DYSTROPHY

JOSEPH H. GLOBUS, NEW YORK CITY

Some years ago I had the opportunity to observe the pathology of the heart in a small number of cases of progressive muscular dystrophy. My attention was drawn to one feature, particularly in two cases: viz., after a long period of *status quo* there suddenly developed a sequence of events which ended fatally. The patients were fairly well, when without warning, there suddenly appeared pulmonary edema, hydrothorax, hydropericardium, with death in twenty-four hours after onset of symptoms.

The opinion expressed for several years has been that death in these cases of progressive muscular dystrophy is due to paralysis of the diaphragm. I believe that in some cases there is another cause operative, namely cardiovascular disturbance.

On studying the heart very carefully anatomically, I found definite changes in the heart muscle, not differing in any way from those in the skeletal muscle. One case was not sufficient to base definite conclusions, and another case was waited for and careful examination made. Lesions were again found which correspond to changes in the skeletal muscle. Dr. Goodhart and I reported this case, but did not sufficiently stress the cardiac lesions. Later another case was examined and well-defined changes were found.

In the literature two types of observations are recorded. Oppenheim in 1911 recorded these lesions as a disease of the central nervous system, but many other workers have interpreted them as myopathic in character. Oppenheim does not feel that the heart is affected similarly to the skeletal muscle, while other workers take the opposite view. Thus two viewpoints are reported: those that record clinical observations without anatomical findings; and *vice versa*.

Among the clinical reports were those of enlargement of the heart, irregularity, and increase of rate and intensity of the heart beat, palpitation on exertion.

On going through the literature I find only eleven cases reported since 1879 in which the hearts were studied. Eight of these recorded definite myocardial lesions, in no way differing from changes found in the skeletal muscle. The other three cases were not so definitely

grouped. There were changes, however, in color, consistency and invasion of the subepicardial fat.

I will report the findings of a boy with progressive muscular dystrophy, ten years of age, who died twenty-four hours after the extraction of a tooth. On autopsy, the muscles were found infiltrated with fat, including the diaphragm. No excess of fluid was found in the thorax. Several small pneumatic areas were found in the lower lobes of the lungs which were edematous.

The heart, pale yellowish red, was flabby and friable. On section, the wall of the left ventricle showed many translucent patches, irregularly distributed, apparently areas of fibrosis which bore no relation to the blood vessels. The thymus was large, solid, and rich in lymphoid tissue. The liver showed marked fatty infiltration.

Microscopically, the heart showed many connective tissue scars. Individual heart muscle fibers were seen in various forms of degeneration surrounded by massive bundles of connective tissue. Fat was found in the connective tissue and about the blood vessels. Many small round cells were seen, interpreted as a reactive phenomenon to the degenerative changes nearby. The muscle fiber changes were edema with swelling or atrophy, hyalinization and fragmentation.

In the thymus were a large number of vessels, some almost completely occluded by proliferation of endothelium.

(Lantern slides were used to demonstrate the various pathological conditions found microscopically in the heart, thymus and skeletal muscles.)

Conclusions: The heart does not escape myopathic processes in progressive muscular dystrophy.

The changes are milder in degree than those in the skeletal musculature. The changes would be more frequently found if the heart were studied more systematically during life, as well as anatomically after death. The heart plays an important rôle in causing death in progressive muscular dystrophy.

DISCUSSION

DR. WALTER TIMME said: I think what Dr. Globus has told us to-night emphasizes the protean character of the disease and its involvement of practically every organ, but not necessarily in every case. You will find cases in which there is marked myocarditis demonstrable in life, and many cases in which it is not demonstrable until after death. I have seen a case of a man eighty-two years, who had the disease since infancy. He was a preacher and had to be carried to the pulpit. He had no sign of myocardial disease. The dystrophy manifested itself in fourteen descendants in three generations, but in none of them was there myocardial disturbance. It is a slowly progressive disease in the milder forms, and in these there is no myocardial-disturbance. Another form of the disease, as described by Gowers, in which death occurred early, from the sixteenth to the nineteenth year, there is usually a cardiac condition and one in which fatty degeneration assumes marked proportions. I think we have got to look upon this disease not exclusively as a muscular one. It is not a pure myopathy, but the myopathies which

occur are the most outstanding feature. Other atrophies, other dystrophies are seen, but not universally seen. You may have progressive muscular dystrophies with only slight muscle changes. But at autopsy you will find disturbances in the anterior horn cells and in Clarke's column, in the anterior gray substance, in Lissauer's zone, which are not at all correlated to the slight muscular disturbance. In other cases at autopsy there are no spinal cord changes, but there have been marked muscular dystrophies. So far as changes in the thymus go, that is very interesting. I radiograph all my cases for thymus, and a very large proportion of cases have persistent thymus and certain cases in early life show a pineal shadow. Marburg has published a paper on early pineal disturbance, accompanied by abnormal fat deposits about the body. I think we are coming more and more to see this disease as one in which the degenerative process is dependent upon deficiency disturbances, probably in the control of some one or other or a group of the internal glandular units. Dr. Globus has made a splendid presentation and I wish to thank him.

DR. FOSTER KENNEDY said: Dr. Globus stated that a certain number of the cases studied died from a water-logging of the chest. I think it would be interesting to know if the case examined by him had such a death: I saw a similar case, a child thirteen years old, who died at nineteen, with pseudodiaphragmatic paralysis. This child suffered acute cardiac dilatation for some three weeks before death. She had hydrothorax, which as Dr. Globus said, sometimes occurs. The acute cardiac dilatation was not the primary lesion, but consequent upon the overstrain upon the heart. That view of the situation may have been entirely mistaken. Perhaps Dr. Globus will comment upon that view.

DR. GLOBUS said: I agree with Dr. Timme's suggestion that the disease of progressive muscular dystrophy is a more generalized pathological condition than we are apt to consider. Bunting found definite lesions in the musculature of the gastrointestinal tract, typical of the lesions found in the skeletal muscle. In one case that came to autopsy, I have found marked fatty infiltration in the liver, with no other pathological changes in the organ. The fatty infiltration was diffuse and bore no definite relation to the vascular or hepatic system of vessels and ducts. There was no inflammatory lesion or degenerative process, but some dystrophic change. The same worker reported changes in the musculature of the large blood vessels. The changes in the liver point to a more generalized pathological condition in progressive muscular dystrophy. I noted hydrothorax and hydropericardium in three cases. The finding of excessive fluid in the pleural and the pericardial cavities led me to the study of the heart muscle. The excess in pericardial and pleural fluids is best explained by the changes in the heart muscle. Dr. Kennedy adds one more to the limited number of cases studied.

DR. FOSTER KENNEDY (continuing) said: It has been said to be common to have marked changes in the spinal cord. I feel inclined to differ from this viewpoint. My experience has been that changes in the anterior horn and Clarke's column were not common. I do not think it is common in the literature. The neural changes occurring

in paralysis are identical with those occurring after amputation, brought about by disuse. I think the changes in the spinal cord are secondary to those in the muscle. If we are to consider this as primarily an anterior horn lesion, and a minor muscular disease, we become extraordinarily fogged in our classification of the pathology. I would like to ask the other men for some expression of opinion as to Dr. Timme's viewpoint. His experience has not been mine. I should like to discuss that point.

DR. WALTER TIMME said: Three cases were reported by Kollarits and two by Schultz. These autopsy findings showed changes in the anterior horn cells in some cases, but not comparable to those in poliomyelitis. The cells are simply smaller than normal, not degenerated. There was some disturbance of the fibers of the anterior cord substance. There were smaller cells in Clarke's column than normal. These findings are not universal. In cases of widespread muscular dystrophy there was not widespread diminution of the cells—often very few small cells. There was no concordance of the two conditions. They appeared to be absolutely independent of each other. It seemed to be a general atrophic condition involving the cord cells, as part of the general process and not secondary.

DR. GLOBUS (in closing) said: Modern neuropathologists have two types of lesions to account for: degeneration and inflammatory conditions. Poliomyelitis is an example of a purely inflammatory lesion. It may have secondary degenerative processes. The primary inflammatory process is characterized, as is well known, mainly by perivascular infiltration, as in any inflammation. You certainly can't think of the cord changes in progressive muscular dystrophy as inflammatory. Are they to be considered as degenerative? Every degeneration in the central nervous system is accompanied by changes in the blood vessels and in immediate vicinity. The modern pathologist in such instances need not study the parenchyma; he studies mainly the functions and relations of the glial elements. In degenerative lesions of the central nervous system one must find cells carrying away the products of destruction in the cord. The modification in size or shape of a nerve cell is not necessarily a pathological phenomenon—it may be due to technical errors in preparation of material for study. But the glial reaction, its accumulation of gitter cells, and other phagocytic cells in the adventitial spaces is evidence of a degenerative process. Such changes are not encountered in the spinal cords of progressive muscular dystrophy.

A NEUROPSYCHIATRIC PILGRIMAGE

DR. SMITH ELY JELLIFFE, NEW YORK CITY

DR. JELLIFFE said: As I hope to bring to you this evening some observations of personalities and of conditions concerning the work going on in Europe in neuropsychiatry following the war, I hope to be able to avoid that criticism once expressed by one of delicate sensibilities who, on observing a certain prima donna, remarked "There were too little clothes and too much Mary." So if in my narrative of my pilgrimage to the intellectual shrines of Europe there is too little shrine and too much Jelliffe, I feel assured I have