This ignorance of the cardinal principles of health on the part of the poorer portion of the community is not to be wondered at, when an almost equal ignorance exists among those who are by mere fortuitous circumstances differently situated. While the first class are oblivious of the real causes of their present condition, and do not regard the results that ensue in the light of cause and effect, the latter have yet to learn, that the moral and material condition of the whole community is involved in the subject.

PATHOLOGY OF PROGRESSIVE MUSCULAR SCLEROSIS.

By WILLIAM PEPPER, M.D., Philadelphia.

THE fragment removed from the left deltoid of the patient under examination was of a slightly pale-reddish color. When examined microscopically, a large majority of the fibrils showed distinct, though often fine and delicate, transverse striation. In a few instances, striation was entirely absent, the fibrils looking homogeneous and much like ground-glass cylinders. In a very few fibrils, also, distinct longitudinal striation was visible, and in others there was multiplication of the nuclei in the sarcolemma. In not a single fibril was there any trace of fatty degeneration. The fibrils varied in size from $\frac{1}{4\,0\,0}''$ to $\frac{1}{2\,5\,0}''$, or even, in a few cases, $\frac{1}{1\,6\,0}''$. The striation was particularly faint, or at times even absent, in the largest fibrils. There was a large amount of interstitial white fibrous tissue, with abundant granular matter containing many oval nuclei. In places there were small collections of minute fat-globules or refracting granules.

The fragments removed from the gastrocnemii presented closely analogous conditions. The muscular tissue was merely rather paler red than normal. The muscular fibrils varied greatly in appearance and in size. The transverse striation was in some fibrils perfectly healthy, but in a majority it was altered, though in various ways. Thus, in some it was very faint and difficult to distinguish; in others, it was wholly absent, the fibrils presenting the appearance of fine ground glass. In other fibrils there was a marked appearance of longitudinal striation, due to delicate fibres or very fine fusiform cells arranged in the long axis of the muscular fibril. In many fibrils there was distinct excess of the nuclei of the sarcolemma, which appeared as large oval nuclei with a punctiform nucleus. A few fibrils presented streaks of minute fatty granules along their centres, and a very small number were decidedly fatty. The muscular fibrils varied greatly in size also. Many were about $\frac{1}{2\,5\,0}''$ to $\frac{1}{3\,5\,0}''$ in diameter; but a number were $\frac{1}{4\,5\,0}''$ to $\frac{1}{4\,0\,0}''$, while others were as much as $\frac{1}{1\,2\,5}''$, $\frac{1}{1\,4\,0}''$, $\frac{1}{2\,0\,0}''$ in width. There was a large excess of interstitial tissue, in places taking the form of long, narrow, wavy bands of pure white fibrous tissue; in others, appearing as abundant granular stroma, thickly strewn with oval nuclei. There was also some curly, elastic fibrous tissue. There was a considerable amount of interstitial fat, existing as scattered globules, or arranged in patches of large, closely aggregated fat-globules. In places isolated muscular fibrils lay imbedded in this fibroid tissue so as to be scarcely visible. But in other places a number of fibrils lay directly in contact with each other, forming a little bundle, around which the excessive growth of interstitial tissue had occurred. The arterioles and capillaries appeared healthy. No nerve-fibrils were detected.

Similar examinations have, as before stated, been made in a number of cases of this disease, and have yielded results agreeing in all essentials with those I have found in the muscles of this patient. The points which I desire to dwell upon as of capital importance in their bearing on the pathology of the disease are, in the first place, that the primary fundamental change in the affected muscles is an excessive growth or hyperplasia of the interstitial connective tissue. This is found to have taken place even in those muscles which have not undergone any increase in bulk, or which are even reduced in size (for instance, the left deltoid in this case). In the latter case it is evident that the process cannot have advanced far, and it also appears probable that there is a certain amount of simple atrophy of the muscular fibrils developed simultaneously. This, however, does not appear to be the only change in the muscular fibrils, which are also found to begin to lose their transverse striation, and to present increase in the nuclei of their sarcolemma, or distinct longitudinal striation. Observe, however, that there is not the slightest tendency to a primary fatty degeneration of the muscular fibril. It may happen that this interstitial growth never reaches such an extent as to cause apparent enlargement of the muscle, so that only certain muscles may thus enlarge. Thus, as a rule, the muscles of the calves are the first to undergo this subsequent change,

and they may be the only ones in which it appears, although numerous other muscles may present the first stage of the process.

In those muscles which do undergo this subsequent enlargement, the hyperplasia of interstitial connective tissue is found to have reached an extreme degree. The appearances presented indicate that the entire process has been one of sclerosis, in which there has been rapid growth of nucleated fibro-cellular tissue, with the development of bands of wavy, fibrous tissue, and even some curly elastic fibres. It is easy to recognize, therefore, the identity of this process with the other sclerotic inflammations, as of the connective tissue of the nervous centres (sclerosis of brain and spinal cord), of the lungs, liver and kidney (cirrhosis), and of the subcutaneous tissue (scleroderma). There is, however, one point in which this affection of the muscles appears to differ from the other sclerotic conditions mentioned. In the latter, we constantly observe that, with the progress of the change, a tendency to organization and contraction of the newly-formed fibrous tissue soon manifests itself, while the essential elements of the part (nerve tubules, liver-cells, or uriniferous tubules) are compressed and undergo atrophic degeneration. In the disease we are now considering, however, there is a simultaneous change in the muscular fibrils, even in the early stage; but this does not appear to depend wholly on the hypertrophy of the interstitial connective tissue, nor does it maintain any definite or constant relation with this latter change throughout the course of the disease.* Indeed, as is seen in this patient, those very muscles which present the greatest degree of sclerotic enlargement may be the strongest of the whole series which are implicated in the disease. It is true that the muscular fibrils of such muscles present a further stage of the change begun in the first period of the disease. Their transverse striation is still more delicate and faint, or is even, in a number of fibrils, entirely lost. But they do not seem to have undergone any further atrophy; indeed, the measurements I have made, and the increased strength in the muscles of the patient's calves, would point to the belief that the muscular fibrils may temporarily share the exaggerated nutrition of the surrounding connective tissue, and undergo a delusive increase in size and power.* Thus, I find that the fibrils in the gastrocnemii muscles of this patient are fully one-third wider than those in his left deltoid, and that some of the former have acquired the enormous size of $\frac{1}{125}''$ in transverse diameter.

Whether this transitory stage usually exists or not, the sclerotic change does not depart from its inevitable law of development. Already in this second stage we have seen patches of fat-globules appearing in the interfibrillar spaces, and as the disease passes into the final stage, this fatty degeneration of the muscle advances with varying rapidity, even leading in some cases to such an accumulation of fat as to be visible to the unaided eye as yellowish streaks. This extreme condition is, however, very rare, and much more frequently the accumulation of fat is moderate. Even when very great, however, it is found to be far more due to the increase of the interstitial fat than to a true fatty degeneration of the muscular fibrils. Their nutrition must be, however, very gravely impaired, so that they undergo atrophy, and in many places entirely disappear, leaving their sheaths empty. This change coincides with the rapid extension of paralysis which characterizes the final stage of the disease.

This detailed account of the muscular lesion naturally leads to the question of the pathology of this curious affection. It cannot be held that the disease depends upon or is essentially connected with any cerebral lesion. It is true that in a number of cases the patients have been idiotic, or at least exhibited marked impairment in intellectual development: in the present case, also, epileptic convulsions are present as a complication. It must, however, be remembered that cerebral disturbances of any kind whatsoever are not uniformly present, that disorders of special sense are rare, and that some cases of the disease have been observed associated with a normal state of the intellectual faculties; and, finally, that in the one case where the nervous centres have been examined, the brain was found healthy. The peculiar character of the muscular lesion and its symmetrical distribution are additional proof, if more were wanting, of the absence of all connection between the disease we are discussing and any cerebral lesion.

Nor is the argument more strong in favor of a spinal lesion as the cause of this affection. It is true that the first idea which

* It may be that in some cases where there is marked loss of power, with general preservation of the size and striation of the muscular fibrils, the sclerotic change in the connective tissue compresses the branches of the motor nerves as they traverse the muscular tissue.

* This enlargement of the size of the fibrils of the gastrocnemii has also been observed by Leyden (loc. cit.).

will arise, on learning from a patient that his malady began with gradual loss of power of both legs, is that there is some disease of the anterior columns of the spinal cord. When, however, as in the present case, we further find that there has been no alteration of sensibility, no loss of coördination of muscular movements, no subjective sensations, such as of formication or of constriction, no implication of either bladder or rectum; when also we learn on careful examination that the loss of power was not in reality the primary symptom, but was preceded by and existed only in proportion to certain muscular changes—we must conclude that the disease is not dependent upon any affection of the spinal cord. The most important fact to be clearly apprehended here is that in reality there is no paraplegia, in the strict and only correct significance of the word, present in the disease we are studying. It is of course true that a paralyzed muscle will often undergo atrophy or fatty degeneration, but these changes are then dependent upon loss of function and of innervation, and are essentially secondary to the interruption of the transmission of motor power. Widely different, however, is the relation which here exists between the loss of power and the alterations in the muscles. There is throughout the course of the disease no real loss of motor nerve-power, nor any interference with its transmission, but merely an interference with its manifestation, owing to a progressive sclerotic change in certain muscles. This change has been slowly advancing for some time before the loss of power is so marked as to attract the notice of the patient; and just in proportion as it progresses, do the muscles become more and more weak, until, when the final stage of the sclerosis is attained, their contractile power is so completely lost that the patient is bedridden and almost motionless. It is on account of the radical difference between this process and any form of paraplegia that I object to the name "pseudo-hypertrophic paralysis," applied by Duchenne to this disease, and greatly prefer "progressive muscular sclerosis," as expressing accurately the pathological condition present.

Having, then, excluded the possibility of either a cerebral or spinal origin for this disease, I can only offer the somewhat unsatisfactory view of its pathology, that it consists essentially in a perverted nutrition of the muscles affected, probably dependent upon a lesion of the branches of the sympathetic nerve which are distributed to the tissues involved. This opinion that the sympathetic nerve is primarily affected, does not, it is true, rest on any positive or convincing evidence. In a certain number of cases, however, especially in those observed in Germany, there have been symptoms noticed, such as reddish or bluish discoloration of the skin of the affected parts and variations in their temperature, which would certainly indicate some marked disturbance in the vaso-motor supply of the cutaneous vessels, and have accordingly led many of the observers of this disease to adopt this view of its pathology. It must be confessed, however, that such symptoms are not constant, or at least are not present at all stages of every case: thus, in the present instance, the most careful examination fails to determine them. Still, for the present, in default of any exact knowledge or of any more satisfactory explanation, this view of the pathology of progressive muscular sclerosis may be accepted.

I may add that no additional light is to be derived from a study of its causes. It is eminently a disease of infancy and childhood, making its appearance, in the vast majority of cases, between the ages of five and thirteen years. It has, however, been observed to begin in one case at the age of fourteen years; in the present case it was first noticed at the age of fifteen years; and in two cases observed by Benedikt (loc. cit.), and in one by Laycock (loc. cit.), the disease seems to have originated in adult life. The disease is much more common in the male than in the female sex; and, finally, it frequently affords evidence of a hereditary tendency, two or even four cases having been more than once observed in a single family. It usually appears spontaneously, not appearing to depend upon any particular external causes. In single instances it has been attributed to such causes as the influence of cold and damp, or an attack of some eruptive fever. In my own case, the cause assigned by the patient—protracted over-exertion in doing work too heavy for his years—is far more likely to have really influenced the development of the disease. It must be conceded, however, that in its etiology, as well as in its pathology, this curious affection still presents an unsolved problem.—*Phil. Med. Times.*

SUPERNUMERARY LITTLE FINGERS.

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Extra digits are by no means a rarity. When removed, they sometimes show a wonderful pertinacity in growing again.