

the interossei, thenar and hypo-thenar groups. The wasting is confined to these small muscles. In this patient the atrophy affects the trunk muscles principally, while the hand muscles are perfectly free from any form of wasting. They differ also as to the condition of the affected muscles. In the spinal case they are soft and flabby, while in our patient here they are firm, hard, and have a knotty feeling. In the man previously seen, the atrophied muscles are the seat of fibrillary twitching, while the muscles in this boy's case are free from these fibrillary movements. Another marked difference is that in the case of the spinal form there is neither true nor false hypertrophy of the muscles, while there is here, especially in the calf. Other points of difference are the ages at which they make their appearance. The spinal form is essentially a disease of advanced adult life, while the juvenile form is seldom or never seen after the twentieth year. They are both slowly progressive diseases; the juvenile is, however, much slower than the spinal variety. In the latter the periods of intermission are comparatively short and seldom, while in the former they are long and frequent. They differ also as to the complications that may arise during their course. Last week, when we were examining the patient affected with the spinal form, I pointed out to you that there was marked trembling of his tongue when he protruded it. This is sufficient evidence that there is commencing bulbar paralysis in his case, and is the beginning of a series of symptoms that will before very long lead to a fatal ending. In the patient before you no such complication exists. In all the cases of the juvenile form of progressive muscular atrophy described up to the present, no such complication has existed. Secondary sclerosis of the pyramidal columns is not infrequent as a result of the changes that take place in the spinal form. It does not occur in the juvenile form. When we come to discuss the pathology of the disease, it will then be clear to you why these complications are so frequently present in the one case and never present in the other. Another marked point of difference between these two forms of atrophy is the fact that one is much more amenable to treatment than the other, the juvenile form being much more likely to have a favorable ending than the spinal.

They differ also in their pathology. In speaking

last week of the appearances found post mortem in the spinal variety of the disease, I mentioned that the essential change was a slowly progressive obliteration of the multipolar cells in the anterior horns of grey matter of the spinal cord. The local muscular changes were simple atrophy of the muscular fibres. There is no increase of connective tissue, no deposition of fat, and no hypertrophy of the muscular fibres. Now in the juvenile form the changes are wholly seated in the muscles. The multipolar cells of the anterior horns of grey matter remain free, as do also the peripheral nerves. The muscular changes consists in atrophy of the muscular fibres, with here and there fibres which have undergone hypertrophy. In advanced cases hyperplasia of the connective tissue is very marked, and lying between the connective tissue fibres is seen only a small quantity of muscular fibres in an advanced state of atrophy, which, however, still retain their transverse striation. The most important change is the hyperplasia of the interstitial connective tissue, and next to this is the deposition of a more or less quantity of fat. It is probable that the increase in the muscular fibres is the first phase of the morbid change, and that the later appearing connective tissue hyperplasia gives rise to atrophy of the muscular fibres. These changes, as we will presently discuss, are essentially those found in cases of pseudo-hypertrophic muscular paralysis, and the so-called hereditary form of progressive muscular paralysis. This hereditary form of muscular atrophy has been described by Friedreich and others, but it is essentially the same disease as we are now considering. When the disease is hereditary and sets in about puberty, the muscles affected are those of the upper arms and trunk, while if it sets in during childhood the atrophy is principally confined to the muscles of the lower extremities.

The disease commonly called pseudo-hypertrophic muscular paralysis, differs but little, if at all, from the disease with which the patient before you is affected. Clinically, the only difference appears to be, that in the pseudo-hypertrophic paralysis, we have lipomatosis, while in the juvenile form of muscular atrophy, hypertrophy is not necessarily present, and if present it is true and not false. If this is the only difference it is quite plain that it would be better to describe the juvenile form of muscular atrophy as being sometimes attended