

## II The morbid anatomy and pathogenesis of the disease.

Having now considered a few points in the anatomy of that portion of the human frame which is most obviously affected by the disease under consideration, we shall pass on to consider the position and nature of the pathological changes which are revealed in it after death. Having described these we can then take a more comprehensive view of the lesion as a whole and consider whether the morbid changes in question are primary in their nature or dependant upon some preceding disturbance of the general economy.

A post-mortem examination of a case of tabes shows the dura mater to be commonly normal, the arachnoid occasionally opaque, and the pia mater congested and thickened in the region of the posterior columns. The spinal fluid has been observed in some cases to be increased, and the trabeculae which traverse the subarachnoid space thicker and more numerous.

The tissue of the posterior columns is ~~decreased~~<sup>decreased</sup> in quantity and tougher than in health. In colour it is greyish, yellowish or pink. The morbid changes are most marked in those lower portions of the cord which we have endeavoured to show are most susceptible to vascular influences. As a rule morbid changes do not spread beyond the medulla, but corresponding alterations have been made out in the superficial layers of the pons Varolii and the Corpora quadrigemina.

For microscopic examination the cord may be hardened in Muller's fluid (one part of sodium sulphate and two and a half of potass. bichromate in a hundred of water), or in osmic acid which stains the myeline sheath of the central nerve fibre, or in ammoniacal solution of carmine as used by Vulpian and Charcot which stains the connective tissue and not the nerve tubules. It is perhaps better simply to freeze the cord in a microtome so as to avoid the possibility of all chemical alterations.

The microscopic examination of an

advanced case of tabes shows that the medullary sheaths as well as the nerve tubes of the posterior columns have disappeared, their places being taken by a loose areolar tissue, which may be simply the neuroglia rendered coarser and more fibrous by irritation. Any nerve fibres which remain appear granular, varicose and narrow. Numerous amyloid bodies, the product of nerve disintegration are scattered about.

The arterioles of the posterior columns have undergone a change also. There is a thickening of the adventitia and all the coats even in the smallest capillaries are infiltrated with oil globules and compound granular corpuscles, more particularly the lymphatic space between the outer and middle coats.

A good deal of attention has been directed towards the point as to which area of the cord is primarily affected. Pierret held that the disease began in two different portions of Burdach's column, while Goll's column became affected later. This view was endorsed by Charcot and by Vulpian. It has been

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modified however by the researches of Strümpell of Leipzig, who has shown that the area primarily affected differs in the various regions of the cord. In the cervical region Goll's columns appear to be involved early in the lesion, while in the dorsal a similar degeneration appears in them early in the disease, but limited to their internal margin. The anterior zone of Burdach's columns remains healthy longest and in typical cases of tabes escapes altogether.

The posterior nerve roots are found to be much wasted, showing a grey or dark discolouration. The fibres may be seen to be destroyed having undergone fatty and granular degeneration, the process being most marked in the axis cylinder while the medullary sheath holds out longer.

The spinal ganglia are not as a rule affected although Luy's and Pierret have reported exceptional cases in which there has been some atrophy in them. The central grey matter of the cord however, more especially

the posterior cornua are frequently found affected, the sclerosis affecting the nerve tubes and the neuroglia rather than the cells. The anterior cornua are usually healthy although Lyden has drawn attention to a peculiar tough hard condition of the anterior ganglionic cells which may be connected with those atrophies of muscle which occur in the course of the disease.

The lateral columns are occasionally affected in tabes, but only when the disease is far advanced. It occurs probably not by an extension from the posterior columns but through an entirely separate nerve lesion. Flechsig's direct cerebellar tract is the portion which usually suffers.

Much attention has not been given to the histology of the sympathetic system in cases of tabes. The researches of Raymond and Arthaud show that there are considerable changes in it, thickening of the vessels, proliferation of connective tissue, atrophy of ganglionic cells and fatty degeneration of the nuclei of Remak's fibres.

Besides the preceding changes the nuclei of the cranial nerves more particularly of the optic, the third and the fifth are frequently more or less affected.

Having considered the lesion as seen in the cord it remains to be considered whether this lesion is essential to the production of the disease. Modern research has shown that it is not so. Westphal in 1878 was the first to call attention to the peripheral changes occasionally visible in tabes. He found atrophy of the cutaneous branches of the cranial nerve in one of his cases. Pierret attributed lightning pains and anaesthesia to a peripheral neuritis of sensory nerves. Pitres and Vaillard have also described peripheral changes resembling Wallerian degeneration which tends to spread centripetally but usually leave the main trunk intact. Dujardin records that in two of his cases the symptoms were equally pronounced. In one after death extensive disease was found

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of the posterior columns, while in the other no central changes were found at all, but extensive neuritis in the peripheral expansion of the nerves.

An interesting case was quoted at a meeting of the Clinical Society of London on March 7<sup>th</sup> 1885 by Mr A Hughes Bennett which is worth transcribing here in greater detail. The patient presented during life all the prominent symptoms of tabes, including incoordination of movement, an ataxic gait, Romberg's symptom, impaired sensibility of the lower limbs, lancinating pains and loss of the knee-jerk. After death the cord itself was found to be perfectly healthy. Outside the meninges however the spinal nerves in the dorsal and lumbar regions were involved in a sarcomatous mass. This was a pathological confirmation of the experiments of Van Dersm and Claude Bernard who by dividing the posterior roots induced incoordination without motor paralysis. A similar conclusion was pointed at by the researches of Varrodt and Heyd.

who showed that by freezing the soles of the feet all power of coordination was lost.

On the other hand Langerhans has examined the touch corpuscles in several cases of anaesthesia without detecting any alteration in them. The evidence however is conclusive that cases resembling tabes dorsalis in every particular may occur without their being any central lesion to account for the symptoms.

Duchenne, the original investigator of the disease and more recently Nefel of New York have asserted that tabes is a lesion of the brain, or rather of the cerebellum. All pathological research and clinical experience is against this view however so that it may be put to one side.

Another theory of the disease which has been hazarded is that its starting point lies in the sympathetic system. This view

is supported by no great name, and is somewhat disparagingly alluded to by Dr Julius Althaus in his recent work upon diseases of the spinal cord. "Even supposing" he remarks "that there should be a constant and well demonstrated change in the sympathetic system, it would be impossible to explain why this should lead only to <sup>changes in</sup> the posterior columns of the cord and no other portion of it." The object of this essay is to attempt this 'impossibility' and to show that given a certain condition of the sympathetic system it is just exactly in the posterior columns of the cord that we should expect to find the very changes which we do find there.

Tacácz and others have thought the primary lesion was a degeneration of the posterior nerve roots which ascends until it implicates the cord itself. Many observers however have observed well marked disease of the columns before any degeneration of the roots, which negatives this view.

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has endeavoured to trace a connection between corns and tabes. In this view he thinks he is supported by the researches of Pitres and Vaillard already mentioned. It is a fact that corns are often conspicuous in tabid patients and that the seat of a corn is frequently the starting point of a perforating ulcer. It is more probable however that the corns are the result of the tabes than that the tabes springs from the corn. In any case in the majority of cases of tabes no such exciting cause can be shown, so that even granting that in an exceptional case the peripheral irritation of a corn might give rise to central disturbance, it cannot be quoted as a theory of the pathogenesis of tabes.

Some observers have thought that the original lesion might be lepto-meningitis, the inflammation spreading from the posterior part of the pia spinalis to the posterior columns. This has been disproved by careful investigations of Strümpell and Tuzgik who have shown conclusively that in early stages of the disease the columns are affected and not

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the membrane. No relation has been proved either between the amount of inflammation in the membranes and that in the columns.

On the supposition that tabes is primarily a disease of the posterior columns of the spinal cord, considerable differences of opinion have arisen as to which element of the cord is first affected.

Ordóñez has put forward a theory that the bloodvessels are the starting point of the disease. He pointed out that in tabes it was common to find the arterioles thickened and blocked up with oil globules and granular corpuscles in such a manner as must interfere with the nutritive functions of the vessels. On the other hand Vaillard and others have asserted that these changes are by no means constant and that considering the effect upon vessels of the experimental section of nerves as done by Waller, the changes in the vessels are more likely to be an effect than a cause.

Bevan Lewis has recently published an account of his microscopical examination of one of Buzzard's cases which appears to bear out the views of Ordóñez. In this case he found ample evidence of periarteritis in the spinal vessels. These vessels were numerous and dilated, with thickened and diseased coats, each one of them being the centre of a patch of sclerosis. In vertical sections ampullar enlargements were seen along the course of the vessels, and groups of large nucleated cells seen in their perivascular sheaths. In this case it was evident that arterial changes had played a prominent part in the disease.

Another theory with which Adamkiewicz has identified himself considers that the primary lesion is a degeneration of the interstitial connective tissue and subsequent atrophy of the nerve tissue <sup>fibres.</sup> In this case the lesion would be analogous to cirrhosis of the liver, and the supposition would accord with the syphilitic theory of the

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disease.

An objection to this theory is that in certain forms of interstitial inflammation of peripheral nerves where the connective tissue is undoubtedly affected there is no wholesale destruction of nerve fibres as we find in the cord in tabes. The balance of pathological evidence is also opposed to the theory of an interstitial origin of tabes.

The theory usually accepted is that the primary lesion occurs in the nerve fibres themselves. The axis cylinders appear to be the first to suffer. It atrophies, the nuclei of the nerve fibres swell and multiply while the myelin sheath becomes segmented and eventually degenerates entirely. The irritation extends to the surrounding connective tissue which exhibits a multiplication of its cells and a proliferation of its fibres. It is at this stage that the coats of the bloodvessels are supposed to participate in the disease.

This theory of the initial lesion in the disease may be applicable to the changes which

occur in the spinal cord and which interrupt the current of sensation and by doing so ~~interrupt~~ throw the reflex mechanism out of gear and do away with the co-ordination of movement. It may also explain, by the irritating changes occurring around the posterior roots, those terrible pains which are such a distinctive feature of the disease. No light is thrown however upon the strange evanescent paralyses occurring often years before the appearance of any spinal disease. Neither does it help us to understand the lesions in the nerves of special sensation, nor the gastric and other crises which also occur frequently in the malady. Above all it does not help us to explain why the posterior columns of the cord, especially in its lower segments should be the principal seat of morbid changes. <sup>On</sup> All these points we are as ignorant as when Bluchenne a quarter of a century ago first picked the disease out of the chaos when then existed among all things nervous and gave it an individuality of its own.