

POST-MORTEM FINDINGS IN A CASE OF PSEUDO-HYPERTROPHIC PARALYSIS; ARTEFACT OF SPINAL CORD.

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IN this communication I propose briefly to record the appearances found in the spinal cord of a typical case of pseudo-hypertrophic paralysis.

CLINICAL HISTORY.

G. R., aged twelve, was first admitted to the Edinburgh Royal Infirmary on 9th June 1894, suffering from pseudo-hypertrophic paralysis; he died on 22nd October 1901.

History.—His parents stated that he was a backward child; he did not walk until he was two and did not speak until he was five years of age. The disease seemed to have commenced at the age of eight; up to that time he was strong and active.

Family History.—The case was an isolated one, no other members of his family being affected; the father, mother and four other children, all younger than the patient, were, and have remained, healthy. There was no definite hereditary history, but a maternal aunt had weakness in the legs, the exact cause of which is uncertain.

Condition when first seen.—The attitude and gait were highly characteristic (see Figs. 1 and 2); the arching of the back, the throwing forward of the thorax, and the marked enlargement of some of the muscles, more particularly of the infraspinati, the triceps, the calf and gluteal muscles, was very striking. The gait was typically waddling. In rising from the floor the patient climbed up his thighs, but he could not completely raise himself to the erect position; he was unable to fix the shoulders to the thorax. The knee-jerks and Achilles-jerks were absent; the bladder and rectum were unaffected. There were no sensory derangements. The mental condition was quite normal.

I need not describe the clinical features and course of the case in detail; suffice it to say the disease slowly but steadily progressed; the patient was from time to time readmitted to the Infirmary.

The photograph reproduced in Fig. 3 shows his condition on the 25th January 1899. He was then unable to stand, walk, or

sit up in bed; he could not raise his head from the pillow; the tendo-Achillis on each side was contracted, and a condition of talipes equinus varus was present.

One of the most striking features of the case at this stage was the extraordinary difference between the size of the thorax and the abdomen; it is well brought out in the photograph. At the level of the nipples the thorax measured $28\frac{1}{4}$ inches; at the level of the umbilicus the abdomen measured $20\frac{1}{2}$ inches. The head was relatively very large, the circumference being $21\frac{1}{2}$ inches.

The muscles, which when the patient first came under my observation were markedly enlarged, gradually became atrophied; in the final stage of the case the muscles of the whole body, with the exception of the masseters and tongue muscles, presented an extreme degree of atrophy; up to the end the face looked full—the masseter muscles and the tongue appeared to be enlarged.

Portions of the muscles were removed during life and also examined post-mortem; the typical features of pseudo-hypertrophic paralysis were present (see Figs. 4 and 5).

The patient died on 22nd October 1901, the total duration of the disease being eleven years.

POST-MORTEM EXAMINATION.

This was hurriedly made three days after death at the patient's home under great difficulties, the weather being somewhat warm. I, unfortunately, myself could not be present at the examination.

The spinal cord was hardened in the ordinary way. The lower part of the cervical, the dorsal, the lumbar and the sacral regions were subsequently examined.

In the lower cervical and the dorsal region of the spinal cord remarkable alterations were found; the grey matter was split up and distributed through the cord in an extraordinary way, and the affected sections curiously malformed. The appearances are shown in Figs. 6 to 12. In one of the cervical sections a double central canal was present—situated on opposite sides of the cord, the two canals were separated by a large interval of grey matter.

Interpretation of the Alterations.—When I first examined the sections I was inclined to think that the appearances were due to a malformation of the spinal cord, but on subsequent examination and consideration I came to the conclusion that they were post-mortem lesions (artefacts).

In the year 1892 Dr. Ira van Gieson published an important

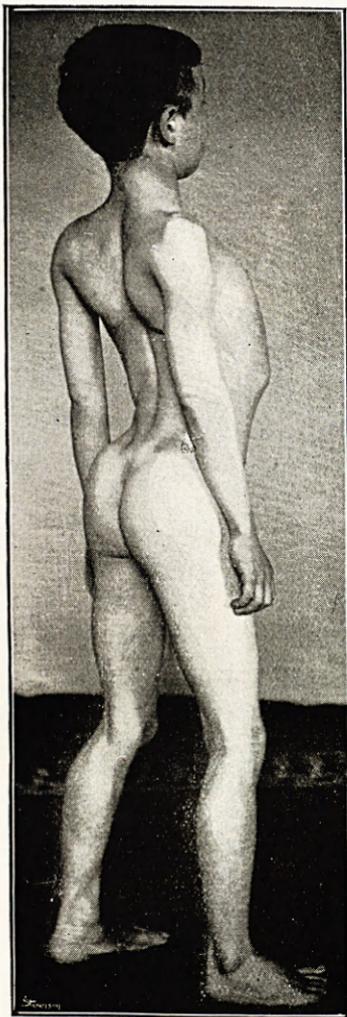


FIG. 1.—The case of pseudo-hypertrophic paralysis described in the text, showing the attitude in the erect position. The curvature of the back, the forward projection of the thorax, and the enlargement of the calves, buttocks and infraspinati muscles are well seen.

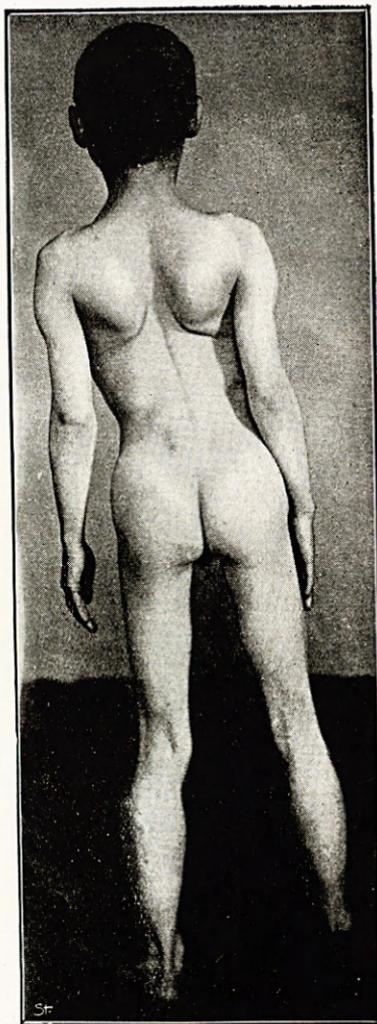


FIG. 2.—The case of pseudo-hypertrophic paralysis described in the text, showing the waddling gait. The great enlargement of the infraspinati muscles is well shown.

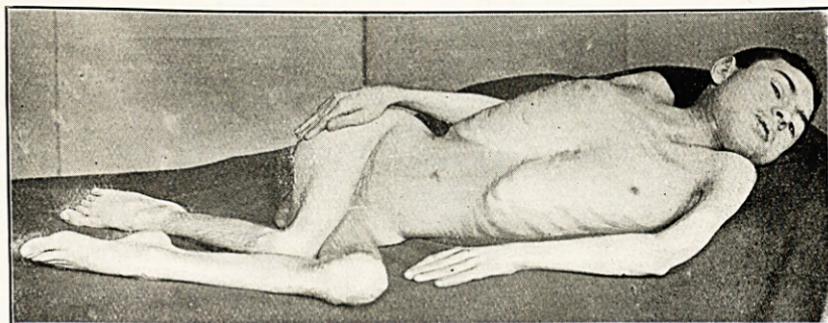


FIG. 3.—The case of pseudo-hypertrophic paralysis described in the text, showing the last stage of the disease—extreme muscular atrophy, contractures, extreme disproportion between the size of the thorax and the abdomen.

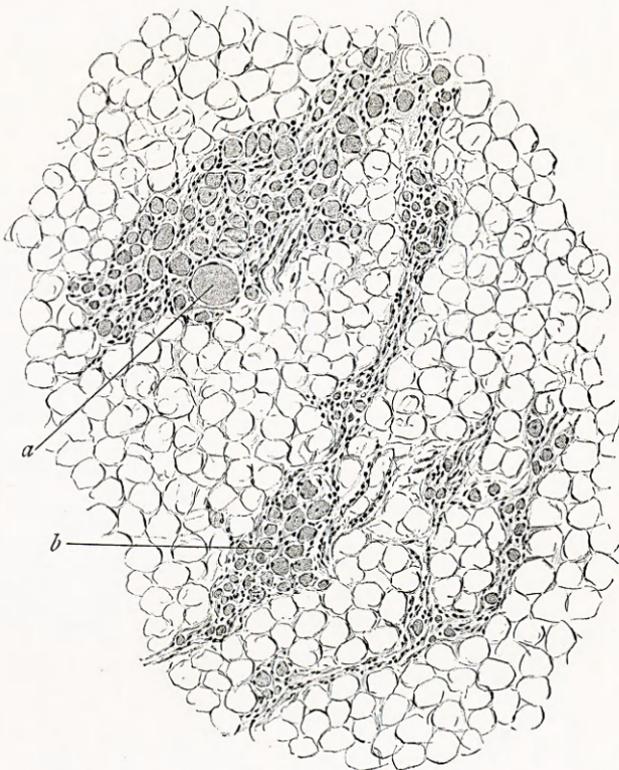


FIG. 4.—Camera lucida drawing of a microscopic section from a portion of the deltoid muscle removed during life in the case of pseudo-hypertrophic paralysis described in the text.

Large masses of fat cells are seen lying between the transversely divided muscular fibres. The letter *a* points to a greatly enlarged muscular fibre, the letter *b* to a group of transversely divided small muscular fibres.

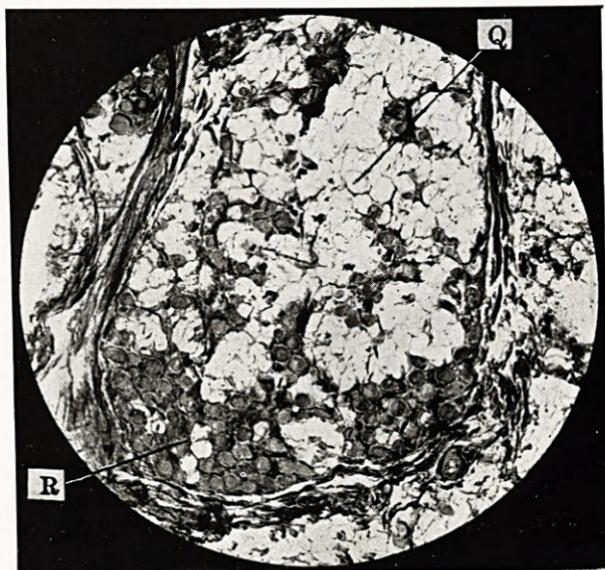


FIG. 5.—Micro-photograph of a transverse section of a portion of the deltoid muscle excised during life in the case of pseudo-hypertrophic paralysis described in the text, magnified 50 diameters.

Numerous darkly stained, transversely divided muscular fibres (to which the letter *R* points) are seen; the groups of muscular fibres are separated by fat and connective tissue (to which the letter *Q* points).

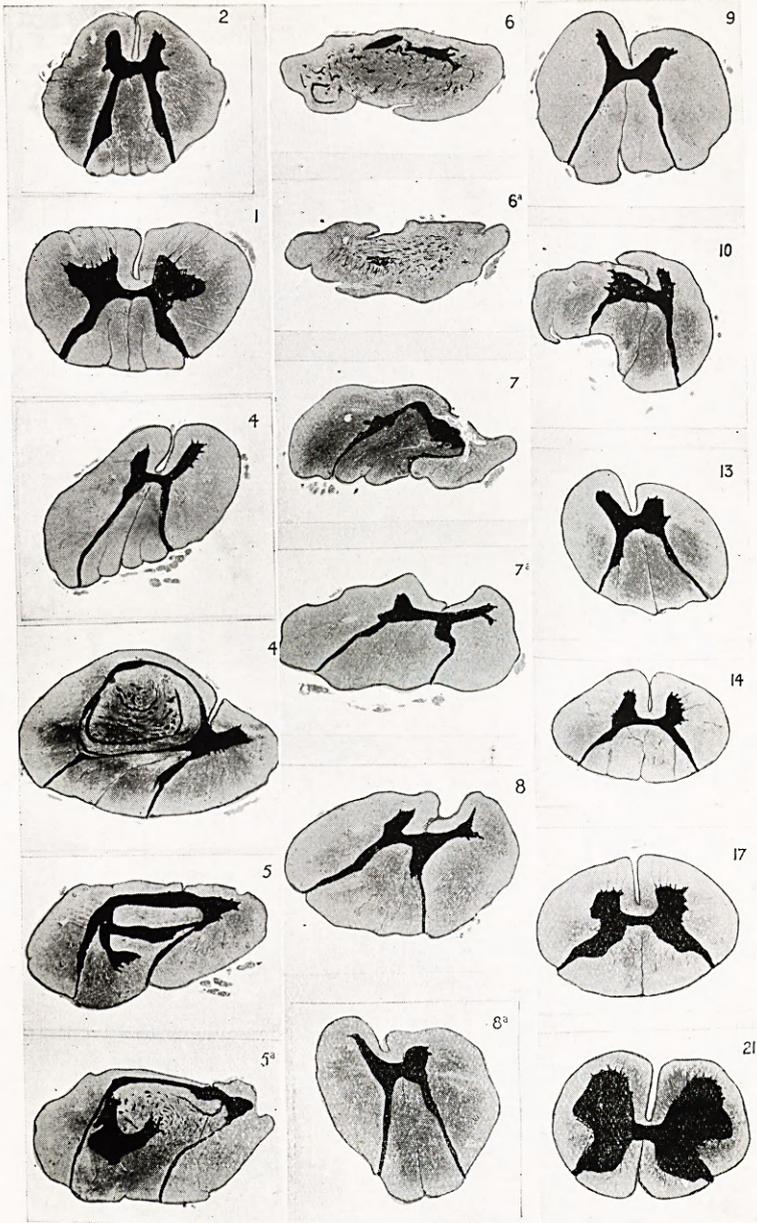


FIG. 6.—A series of sections through the lower cervical, dorsal and lumbar regions of the spinal cord in the case of pseudo-hypertrophic paralysis described in the text, stained by Weigert-Pal.

Figs. 4 and 4a, 5 and 5a, 6 and 6a, 7 and 7a, and 8 and 8a are sections through the upper and lower portions of the same block (segment).

The structure of the cord in Figs. 6 and 6a is completely disorganised; probably this was due to a severe bruise of the cord at the level of this segment. The segments immediately above the bruise (Figs. 5 and 4) and the segments immediately below the bruise (Figs. 7 and 8) are malformed, more particularly the segments above the lesion (bruise), the grey matter being forced upwards and downwards into the white matter.

The sections numbered 1, 2, 14, 17 and 21 are normal.

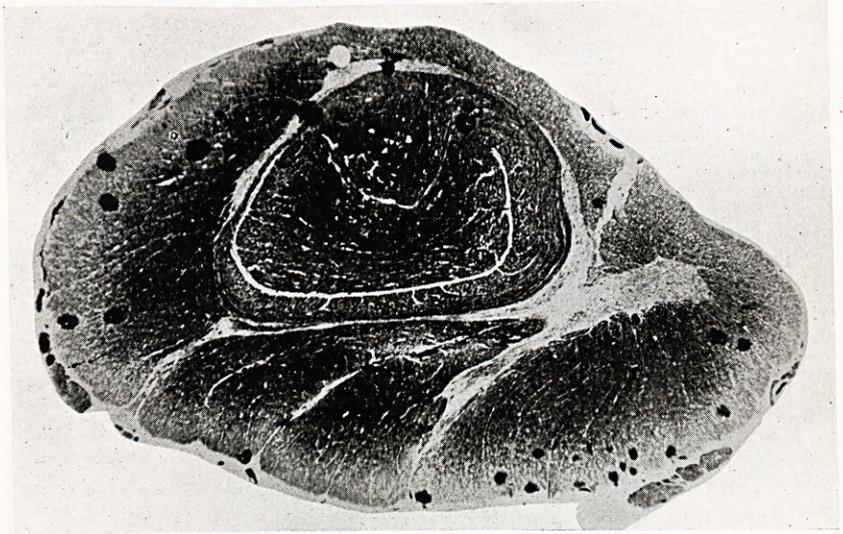


FIG. 7.—The segment represented in 4a, Fig. 6, more highly magnified.

In this section and in the sections represented in Figs. 8 and 9, the grey matter which is unstained is scattered about, in the midst of the darkly stained white matter, in the most curious way.

A fine horse-hair was passed through the left half of the spinal cord; the round hole seen in the sections reproduced in Figs. 7, 8, 9, 10, 11 and 12 represents the position of the horse-hair. It will be noted that in Fig. 9 the section has been reversed, the hole due to the horse-hair being on the right side of the section.

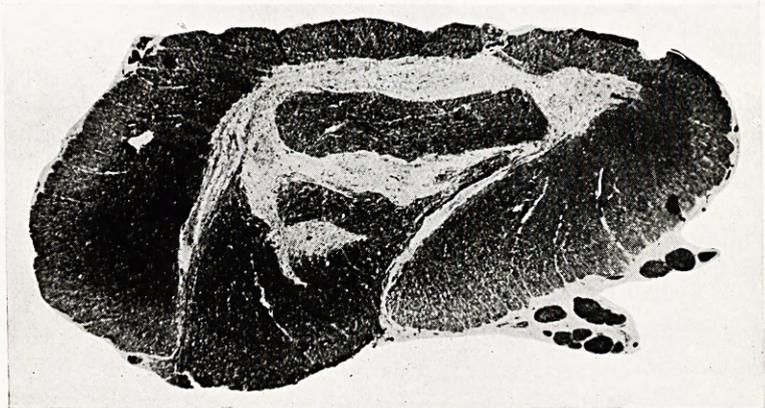


FIG. 8.—The segment represented in 5, Fig. 6, more highly magnified.



FIG. 9.—The segment represented in 5a, Fig. 6, more highly magnified.



FIG. 10.—The segment represented in 6, Fig. 6, more highly magnified.

In this section, and still more in the section represented in Fig. 6a, the grey matter has been almost completely squeezed out of the section and the direction of the fibres of the white matter very much altered.

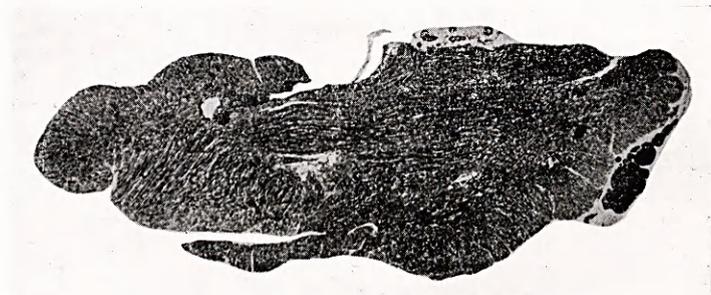


FIG. 11.—The segment represented in 6a, Fig. 6, more highly magnified.

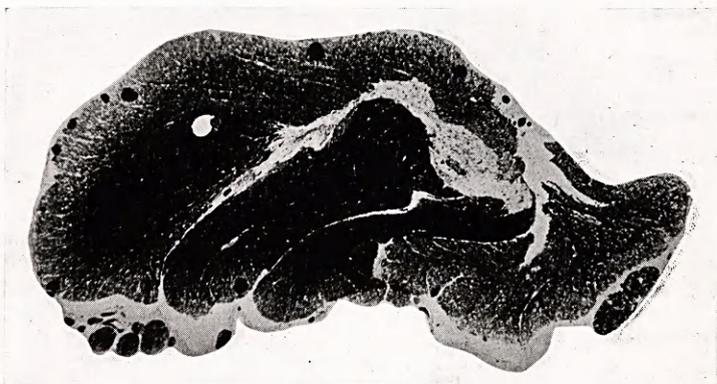


FIG. 12.—The segment represented in 7, Fig. 6, more highly magnified.

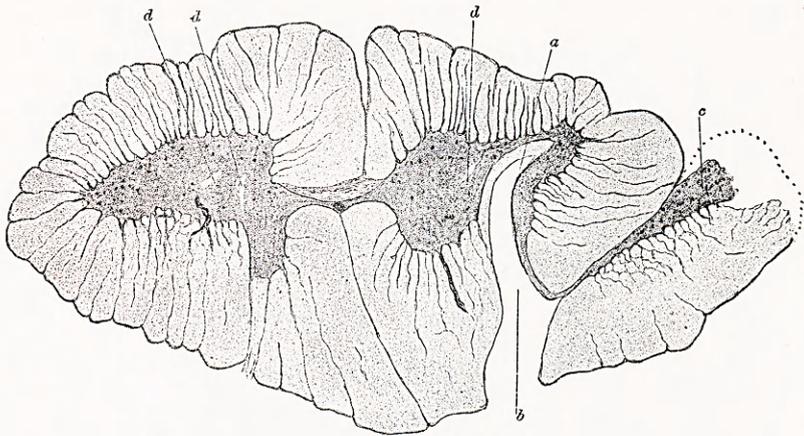


FIG. 13.—Transverse section through the middle of the cervical enlargement in a case of pseudo-hypertrophic paralysis (magnified about 10 diameters).
The grey matter of the right lateral half is split up by a band of white matter (*a*) which passes into it from the posterior column, and by a deep fissure (*b*); *c*, detached portion of grey matter running out to the surface of the lateral column; *d, d, d*, fissures or tears in the grey matter.

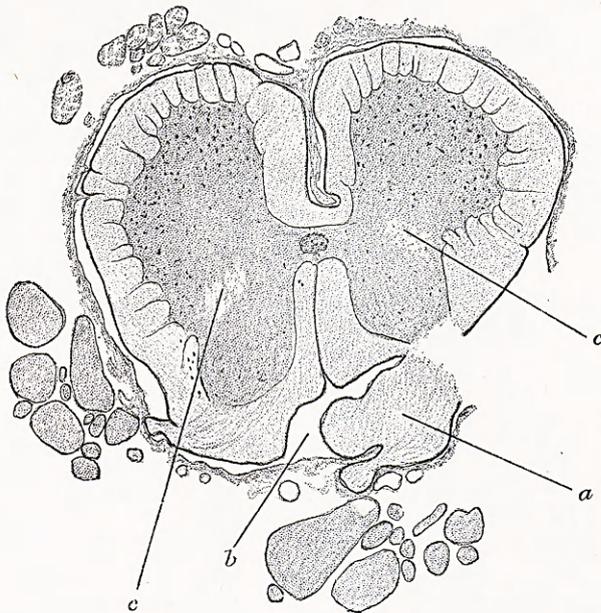


FIG. 14.—Transverse section through the lumbar enlargement of the spinal cord in a case of pseudo-hypertrophic paralysis.
The posterior column is peculiarly shaped; part of the posterior horn of grey matter and a knob-shaped mass of the posterior column, to which the letter *a* points, are separated from the rest of the cord by an artificial fissure. The intermediate grey matter on both sides of the cord, to which the letters *c, c* point, is disintegrated. Two nerve cells are seen in the left posterior column, and several in the left lateral column, just outside the posterior horn of grey matter.
Note.—In the process of mounting this section has been reversed; the right half of the drawing is really the left half of the cord.

paper¹ on the "Artefacts of Nervous System," in which he showed that alterations in the shape and form of the spinal cord and of the relative positions of the grey and white matter, quite as remarkable as those which are present in the case which I am recording, may result from mechanical lesions (bruises, &c.) made in the spinal cord after death. In that paper he emphasises the importance of care in removing the spinal cord from the body, a point which cannot be too strongly insisted upon, more especially when the cord tissues are softened either by ante-mortem or post-mortem changes.

In the first edition of my book on the Spinal Cord, published in the year 1882, I described and figured the post-mortem findings in a case of pseudo-hypertrophic paralysis. In that case a curious alteration in the spinal cord, which I thought was a malformation, but which van Gieson thinks was an artefact, was present (see Fig. 13). In that, my first, case, fissures or localised softenings were present in the intermediate grey matter between the anterior and posterior horns.

In a second case of pseudo-hypertrophic paralysis, which was published and figured in the second edition of my book on the Spinal Cord, softening of the intermediate grey matter (which was perhaps an ante-mortem lesion), and a post-mortem lesion of the posterior column (artefact) were present (see Fig. 14).

From these statements it will be seen that in three cases of pseudo-hypertrophic paralysis which I have examined microscopically, alterations in the spinal cord, all of which were apparently artefacts, were present. If this is so, it is a remarkable coincidence, and shows how extremely careful one should be in drawing conclusions not only from one but from several cases. It is needless to say that I have examined microscopically a very large number of spinal cords; in four only out of all of the cases were there appearances suggestive of artefacts. The remarkable point is that three of these four cases were cases of pseudo-hypertrophic paralysis. I should add that in only one of the four cases did I myself remove the spinal cord from the body.

In the fourth case, which I have described and figured in the second edition of my book on the Spinal Cord, Plate LXIV. p. 74, a detached portion of grey matter was present in the cervical region between the two anterior horns of grey matter. The condition in that case was, I thought, due to a malformation; van Gieson thinks it was an artefact.

¹ *The New York Medical Journal*, 1892.