PAINFUL SPINA BIFIDA OCCulta
WITH REVIEW OF THE LITERATURE
FRANKLIN JELSMA, M.D. AND E. J. PLOETNER, M.D.,
Department of Surgery, University of Louisville School of Medicine, and
St. Joseph Infirmary, Louisville, Kentucky
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In a review of the literature from 1930 to 1950, 14 references to painful spina bifida occulta were found.1–3,5–7,9,11,12,14,21,24,32,34 Usually it is thought that spina bifida occulta is of no clinical significance and that it produces no symptoms, but actually a certain percentage of cases of low-back pain and even of pain that extends into the extremities are attributable to this congenital defect. It is known that myelodysplasia and filum terminale traction may cause organic changes in the lower extremities.26

We were able to find 48, or 0.8 per cent, out of a group of approximately 6,000 cases of low-back pain, in which we thought the pain was produced by spina bifida occulta. This conclusion was arrived at mainly by a process of exclusion of the usual entities known to produce low-back pain, and then only after a series of examinations and re-examinations and a review of these cases over and over again. We have operated upon 18 patients in this group and have obtained very gratifying results. Therefore, we thought this lesion should be more thoroughly studied and evaluated and brought to the attention of those dealing with low-back pain.

HISTORICAL CONSIDERATION

In 1641 Nicholas Tulp, a Dutch physician, first described spina bifida and by 1652 he had reported 6 cases.29 Virchow26 in 1875 was first to describe spina bifida occulta. Then followed a group of clinical reports by von Recklinghausen25 in 1886, Denec64 in 1906, Hesse17 in 1918, Sternberg27 in 1929 and many others.

Up until 1915, 85 cases of spina bifida occulta had been reported. With the development of better X-ray equipment and its more general usage, it was soon found that the lesion was quite common. At the present time it is thought to occur in the low back in about 3 per cent of the adult population.

Next followed many papers explaining the faulty embryological development.8,10,19,20,22,25,33 The earlier theories of causation are of interest. Lebedeff22 in 1881 felt that the lesion was caused by the bending backward of the embryo in utero. This idea of stress on the neural plate preventing closure of the neural tube was also supported by Ernst8 in 1909. von Recklinghausen25 in 1886 thought the defect was the result of incomplete development of mesodermal tissue. Frazier10 in 1918 suggested that increased spinal fluid pressure, from delayed absorption, was the causative mechanism. Sachs19 in 1934 reported that hyperactivity of the choroid plexus in utero might cause the condition.
Let us consider some underlying physiological or biochemical disturbance as the causative mechanism for spina bifida occulta. Levine\textsuperscript{23} in 1942 presented the isosensitization theory as being associated with congenital defects. Further elaboration on this theory was made by Wiener\textsuperscript{31} in 1947. Many have accepted the present-day concept of the antigen-antibody reaction as a primary cause of developmental defect. In this concept the antibodies from the mother pass through the placenta into the fetus, producing certain types of physiological reactions, since there is an isosensitization of the mother to the antigens from the fetus. This may produce (1) intravascular hemolysis, or (2) intravascular coagulation (clumping of fetal erythrocytes). We wish to suggest that this process may produce the embryological developmental defect known as spina bifida occulta.

**PHYSIOPATHOLOGY**

The relationship of low-back pain to spina bifida occulta is clear when the physiopathological nature of the lesion is understood. With the incomplete development of the neural arch, nature fills the defect with dense fibrous tissue, which sometimes encases the dura mater snugly. The ligaments in the area are not attached to a true osseous structure but to this fibrous-tissue replacement. This fibrous mass, being closely associated with the dura and the nerve roots, can effect a stretching or compression syndrome producing local or referred pain.

Our interpretation of the gross pathological lesion in spina bifida occulta producing pain, agrees with observations made elsewhere in the literature.\textsuperscript{3,5,8,9,11} There is an inelastic, unyielding fixed mass of fibrous tissue filling the space normally occupied by the midline structures of the low back. The usual mechanics of motion are altered. Normally, the dura is free and protected by the epidural fat with ample space about it. The laminae and the posterior spinous processes serve as additional protection to the dural canal and spinal nerves. Traction of ligaments, muscle pull, stress and strain, are absorbed by these rigid structures. But when the latter are absent, this force is transmitted to the dural canal and spinal nerves of the area involved. A mass of fixed and even hard fibrous tissue extends from the superficial fascia to the dura. This mass of tissue may be quite large, depending upon the size of the defect. Sometimes it practically encases the thin poorly developed dura mater and the nerve roots emerging. This sort of pathological structure does not serve to protect the dura and nerves as well as does the normal bony canal with epidural fat in place. It does not permit motion without traction. Instead, bending and twisting cause direct dural and nerve pull and this in turn produces pain. This same condition is often produced by postoperative scar tissue, as has been demonstrated at re-operation for discs or after a laminectomy for other spinal cord pathology.

In 3 instances we have also observed impingement upon the canal by nubbins of misshapen bone representing the rudimentary lamina. These pieces of bone may project into the canal from their lateral attachment and
produce dural compression with indirect nerve involvement. Or, as in the 3 cases noted, the rudimentary lamina may directly impinge upon the nerve root extradurally and cause pain.

Since the above physiopathological processes have occurred repeatedly in our cases, we feel they are a reasonable explanation of the low-back pain in spina bifida occulta.

SYMPTOMS

In most cases symptoms and signs are completely absent. Occasionally low-back pain will be the only complaint. Hadley reported that 4.1 per cent of individuals with spina bifida occulta have low-back pain. This pain may vary in degree, location and extent. It depends, however, on the anatomical structures or tissues involved and the manner in which the pain is produced. Sensations of heaviness, and pulling of the back muscles on moving or bending may occur with local back pain. With involvement of the lumbar nerves there may also be radiation of pain into the buttocks and hamstring muscles.

Local inspection often reveals a subcutaneous lipoma, a telangiectasis or a dimple over the lumbar or sacral region, that may suggest the underlying defect. There may be an ironed-out lumbar lordosis associated with local tenderness over the back muscles. Reflexes in the lower extremities are sometimes diminished. There may or may not be a positive Lasègue’s sign. Other and more marked findings, such as motor and sensory changes, with deformity of extremities, hypertrichosis, etc., occur with the more extensive types of spina bifida occulta.

Dittrich reported 5 cases in which the patients required absolute bed rest for 3 weeks or more because of the intense low-back pain. A laminectomy with removal of the pathological structures resulted in complete relief of the pain.

CASE REPORTS

The following cases may serve to illustrate the syndrome of painful spina bifida occulta.

Case 1. F., a woman 41 years of age, was first seen by us in December 1940. She had been disabled for the preceding 3 weeks because of pain in the low back, extending into the buttocks on both sides but never into either lower extremity. During the past 4 years, pain had been intermittent and caused considerable suffering. Bending and twisting would aggravate it. A pelvic operation for malposition of the uterus had been performed in 1936. The low-back pain continued.

Examination. Reflexes in the lower extremities were present and equal. There were no sensory or motor changes. The sphincters were normal. A fat pad was palpable over the sacrum and a defect in the spine could be felt beneath it. X-rays showed that the lamina of the 5th lumbar vertebra was not fused. There was a defect in the posterior arches of the 1st and 2nd sacral segments (Fig. 1).

Operation. On Jan. 14, 1941 a large fibrolipomatous pad extending from the superficial fascia to the dura was disclosed (Fig. 2). The dura was firmly encased by this structure while the external portion of the fibrolipomatous pad became continu-
ous with the lumbar fascia. It was necessary to dissect this fibrous-tissue encasement from the dura throughout the extent of the long defect. The dura was much thinner than normal.

*Course.* Convalescence was normal. The patient has remained free of pain to date, 11 years later.

*Case 2.* R.K., a male aged 40 years, was seen first in February 1946. This man was a tense business executive and had been suffering from pain in the low back for 12 years. At times the pain was in one leg and then the other. He had had numerous examinations in various clinics and had been studied thoroughly from all angles. X-rays and other studies were repeated over and over again and always the patient was told that nothing could be found. The low-back pain continued to cause considerable suffering that tended to incapacitate him about half the time. He became discouraged as the years slowly passed without relief. Every now and then he made a new effort to secure relief and would go to another medical center for another complete study. Fortunately he was well able to afford these numerous and extensive medical studies. Finally he resorted to osteopathic treatment, a low-back brace, and empirin compound as a daily routine.

*Examination.* He was a large, well developed individual. BP was 160/115. His back was straight. There was tenderness on pressure over L5 and in the midline, and over the upper part of the sacrum. All reflexes were normal. There was no hypesthesia over the lower extremities and there were no sphincteric changes. Bending would cause pain in the back. Lying down would give relief.
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X-rays showed an incomplete fusion of the posterior arch of the 1st sacral segment and a very small L5 lamina. There was considerable lack of bony development in the S1 and L5 segments (Fig. 3).

Clinical Diagnosis. Considering this patient from an objective standpoint, there was very little with which one could hope to explain his troubles. Organically, the congenital defect in the lower spine was all one could find. Since the defect was in the exact site of his pain, and in view of our Case 1, we talked the matter over with the patient. We explained that again we found nothing organically wrong with him except the spinal defect which it had been our custom in previous patients to discount entirely, with one exception. This woman (Case 1), with a similar history and similar findings, had been operated upon and had done well. The patient, being an astute business man, thought the matter over carefully and reported back for surgery.

Operation. On April 8, 1946, under local anesthesia, an incision was made from L4 to S2. A midline fascial septum extended down to a fibrous-tissue mass which filled the space ordinarily occupied by the posterior spinous process and the lamina of the 1st sacral segment (Fig. 4). All the scar tissue and fascia were dissected free from the dura. The 4th and 5th interspaces were explored with a nerve hook and nothing could be found in the way of a disc protrusion; in fact the operator could put his finger in the defect of the lamina and palpate the anterior surface of the canal.

Course. Convalescence was normal, and the patient has had no pain subsequently. He is very gratified over the relief he has obtained, and has asked repeatedly why he had not been operated upon long before.

Case 3. M.H., a farmer aged 38 years, complained of low-back pain extending into both hips, for the past 20 years. He had been disabled for 4 to 5 weeks at a time

Fig. 3. Case 2. Roentgenogram showing bony defect in S1 and L5 segments.

Fig. 4. Case 2. Fibrous tissue extending from lumbar fascia to dura mater.
on several occasions. Coughing and sneezing did not aggravate the pain, and lying down relieved it. Bending, twisting and turning caused pain.

Examination. There was tenderness on pressure over L5 on both sides and over the sacrum. There was no visible change in this area, but a bony defect could be palpated. Roentgenograms showed a spina bifida of the 5th lumbar and 1st sacral segments (Fig. 5). The lumbosacral angle was increased.

Course. We followed this patient for almost a year. Palliative measures, such as physiotherapy, medication and a brace did not give satisfactory relief.

Operation was performed on Oct. 30, 1945. The fascia extended into the midline all the way down to the lamina and about the edges of the bony defect. What seemed to be dura was actually a fibrous encasement of the dura. When this fibrous encasement was opened, epidural fat was found beneath it. The dura was dissected free. The dura and cauda equina were definitely compressed and constricted by this tissue (Fig. 6). It was noted that the undersurface of the 5th lamina remnant compressed S1 nerve root. This was rongeured away.

Histological diagnosis: Hyalinized connective tissue.

Course. Convalescence was normal, and the patient returned to his duties as a farmer. He has not complained of any more trouble.

Case 4. A.K., a male aged 47 years, complained of intermittent low-back pain extending into the buttocks but not extending into either lower extremity for the past 20 years. He had been incapacitated at times, but for 2 weeks prior to admission in August 1948, the pain had been constant.

Examination. There was tenderness on pressure over L5 and the upper sacral region, more in the midline than elsewhere. Reflexes were normal. There were no sensory or motor changes, and no sphincteric changes. Roentgenograms of the

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[Fig. 5. Case 3. Bony defect of 5th lumbar and 1st sacral segments, with curved laminal remnant compressing S1 root.]
lumbosacral spine and pelvis showed slight reduction in the normal lumbar lordosis, possibly associated with some muscle spasm. The intervertebral joint spaces were normal. There was a large defect in the posterior arches of the 5th lumbar and 1st sacral segments (Fig. 7), a developmental variation that frequently is not associated with clinical symptoms.

Operation, Sept. 4, 1948. It was found that the nubbin of the right L5 lamina was freely movable, and this was removed. The fibrous lipomatous mass filling the bony defect was carefully resected, including that part encasing the dura.

Course. Recovery was normal. The patient has had no more complaints and has continued his work as a dairy salesman.

COMMENT

The above cases would indicate that low-back pain, with extension of pain into the hips, is rather characteristic of this condition. Sometimes, however, the pain may extend into one or both lower extremities. The fact that pain is on both sides is certainly significant. Also, there is a history of recurrent and often continuous pain over a number of years, aggravated particularly by bending. Usually there are no reflex changes or sensory changes.

These people seem to represent a group of spina bifida occulta who have not had the additional cord changes as occur in the more severe cases, or when myelodysplasia is present. We have not noted any cord involvement through traction by the filum terminale.

It seems that the mechanism producing the pain is caused by nature’s effort to fill in the bony defect with a fibrous or fibrolipomatous tissue, and there is often found a constricting mass of fibrous tissue involving the dura mater and at times the sheaths of the emerging roots in the area.

Our interpretation of the gross pathological lesion in this condition is based on the premise that painful spina bifida occulta occurs usually in those individuals with a minimal bony defect who have no myelodysplasia or signs of cord involvement. In the cases in which we have performed operation, there was a traction mechanism produced by dense fibrous tissue, which many times encased and constricted the dura mater, and sometimes even produced traction on the nerve roots. The fibrous tissue extended from the dura all the way up through the spaces where ordinarily the posterior spinous processes would have developed, and attached itself to the lumbar fascia.
of the back. In fact, the lumbar fascia invaginated in the midline and actually became part of the lesion, so that when the back was bent, traction was produced by the non-elastic fibrous-tissue structures. Sometimes small bands or cords of fibrous tissue could be found extending from the defect in the lamina to the dura mater and to the sheaths of the nerve roots. Occasionally lipomatous pads were interposed between the dura and the dense fibrous tissue encircling it, but this was not a usual finding.

Microscopically, local swelling with minimal hemorrhages of the tissue was noted. There were some calcified flakes in the connective tissue, indicating old hemorrhagic areas. Some lymphocytic infiltration was noted. In one case there was hyalinization of the connective tissue.

SUMMARY

1. A review of the literature on spina bifida occulta is presented.
2. The possibility of isosensitization between mother and fetus is suggested as a causative mechanism for this developmental defect.
3. Low-back pain and pain into the lower extremities may occur with spina bifida occulta. The mechanics of this pain are explained.
4. The symptomatology of painful spina bifida occulta is given, with illustrative case reports.

REFERENCES

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