DURAL ECTASIA IN VON RECKLINGHAUSEN’S DISEASE OF THE LUMBAR SPINE: A CASE REPORT

C. EICHHORN, G. WENDT, H.-W. STAUDTE, J.-M. GILSBACH

Dural ectasia is known to occur in von Recklinghausen’s disease and to cause bony erosion which may be severe enough to destroy spinal stability (Winter 1991). We report a case in which fractures of lumbar pedicles were treated by internal fixation.

Case report. A 20-year-old woman known to have type-I von Recklinghausen’s disease involving the lumbar spine was seen for the first time in 1985. She complained of back and leg pain. Radiographs showed dural ectasia and we performed an endoscopy of the lumbar dural sac to exclude local neurofibroma as the cause of its width. Symptoms were not severe, there were no neurological deficits and the patient later had three normal pregnancies and deliveries.

In 1992 and 1993 she was reviewed several times with increasingly severe back pain, and was eventually unable to walk. Neurological examination had remained normal. At this stage MRI, CT, myelography and plain radiographs showed multiple fractures of the pedicles of L1 to L4 with dural ectasia penetrating the body of L2 (Figs 1 and 2). The transverse diameter of the dura was twice that of the vertebral body at this level, reaching and lifting the psoas.

Operation was planned to graft the fractured pedicles in an attempt to restore stability. After autologous blood dona-

Figure 1 – Lumbar spine radiographs in different years showing the fractured pedicle at L3. Figure 2 – MRI of lumbar spine in 1993 showing the gross dural ectasia.

C. Eichhorn, MD. Senior Registrar
H.-W. Staudte, MD. Professor of Orthopaedic Surgery
Department of Orthopaedic Surgery. Kreiskrankenhaus Marienhöhe, 52146 Würselen, Germany.

G. Wendt, MD. Assistant Professor
Department of Thoracic and Cardiovascular Surgery
J.-M. Gilsbach, MD. Professor of Neurosurgery
University Clinic, RWTH Aachen, D-52057 Aachen, Germany.

Correspondence to Dr C. Eichhorn.

©1995 British Editorial Society of Bone and Joint Surgery
0301-620X/95/$5.00
Received 31 August 1994; Accepted 2 November 1995
rest because of leg and back pain thought to be due to direct contact between the dura and the muscles. She still had no neurological deficits but the bone grafts were almost completely resorbed.

One screw at L3 was protruding anteriorly and was thought to be in danger of damaging the abdominal aorta. At a transabdominal operation in April 1994 with vascular surgeons the aorta was exposed and the end of the screw cut off. The aortic wall was intact. Since this series of operations the patient has improved and is able to live a normal life as a housewife.

**Discussion.** There are many reports of the spinal manifestations of von Recklinghausen’s disease (Heard, Holt and Naylor 1962; Isu et al 1983; Castelein and MacEwen 1984; Stone et al 1987; Pfeiffer and Gutsche 1988; Winter 1991; Bensaid et al 1992; Craig and Govender 1992). Dural ectasia has also been reported in Marfan’s disease (Stern 1988).

The typical scalloping of the vertebral bodies has been thought to be due to primary dysplasia, but we agree with Heard et al (1962) that this and the dural ectasia are a result of increased dural pulsation due to weaknesses in the dura, causing erosion of the vertebral bodies.

In the case which we report the pedicles slowly became thinner over a period of six years until they fractured. CSF pressure was always at a normal level. Our attempt to graft the pedicles failed, despite an apparently stable implant. Others have reported difficulties in obtaining solid bony fusion in patients with von Recklinghausen’s disease (Heard et al 1962; Stone et al 1987).

Should our patient deteriorate and again become unstable we plan to re-expose the fixation and add more bone as seems necessary. We consider that an anterior approach would be potentially dangerous (Winter, personal communication, 1994).

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

**REFERENCES**


