PROGRESSIVE SCOLIOSIS AS THE FIRST PRESENTING SIGN OF SYRINGOMYELIA

REPORT OF A CASE

A. S. BAKER, JOHN DOVE

From the Stoke-on-Trent Spinal Service

We report the first recorded case in which syringomyelia presented purely as a progressive scoliosis in the absence of abnormal peripheral neurological signs.

There is an established relationship between syringomyelia and scoliosis (Woods and Pimenta 1944; McIlroy and Richardson 1965; Huebert and MacKinnon 1969; Logue and Edwards 1981). However, we report a case that we believe to be the first in which syringomyelia presented purely as a progressive scoliosis in the absence of peripheral neurological symptoms or signs.

CASE REPORT

A five-year-old girl was found to have a thoracic scoliosis during a routine medical examination at school. The child had been born at full-term by a spontaneous vertex delivery after a normal pregnancy. She had no relevant past history nor was there a family history. An erect anteroposterior radiograph of the spine showed her to

Fig. 1

Fig. 2

Figure 1—An anteroposterior erect radiograph of the spine showing a thoracic curve with a Cobb angle of 23 degrees. Figure 2—Erect anteroposterior radiograph taken three months later showing rapid progression of the curve to 42 degrees.

A. S. Baker, FRCS, Senior Orthopaedic Registrar
Mount Gould Hospital, Plymouth, Devon PL4 7QD, England.
J. Dove, FRCS, Consultant Orthopaedic Surgeon
Hartshill Orthopaedic Hospital, Hartshill, Stoke-on-Trent, Staffordshire ST4 7NZ, England.

Requests for reprints should be sent to Mr A. S. Baker.

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have a thoracic scoliosis convex to the left from T5 to L1 with a Cobb angle of 23 degrees (Fig. 1). When she presented to the Stoke-on-Trent Spinal Service three months later, she was found to have a clinically mobile thoracic scoliosis with a rib hump of 1.5 centimetres. The results of careful neurological examination, including

temperature testing, were entirely normal. An erect anteroposterior radiograph of the spine showed that the curve had progressed to 42 degrees in only three months (Fig. 2). A possible transposition of the large bowel was conjectured from the presence of a left subcostal liver shadow on the radiograph.

Radiographs of the cervical spine (Figs 3 and 4) showed marked widening of the spinal canal (Lee and Williams 1977). A cervical myelogram showed that the child had syringomyelia (Fig. 5). Hydrocephalus was excluded by means of a computerised tomographic scan of the head.

Fig. 3

Fig. 4

Figure 3—Anteroposterior radiograph of the cervical spine of a normal child of the same age as the patient. Figure 4—Anteroposterior radiograph of the cervical spine of the patient with syringomyelia showing a widened canal.

Decompressive surgery was performed because of the progressive scoliosis. The posterior margin of the foramen magnum and the arch of the atlas were removed. The dura was opened and by separating the cerebellar tonsils a free flow of cerebrospinal fluid was obtained

Fig. 5

Supine lateral myelogram of the cervical spine showing the widened cord (A), the Chiari malformation (B), and the fourth ventricle (C).

from the fourth ventricle into the spinal subarachnoid space thereby diverting flow away from the syrinx.

The child made an uneventful recovery from the operation and is now being managed in a Milwaukee brace. Twenty months after operation an erect anteroposterior radiograph of the spine in the brace showed that the curve was controlled at 10 degrees.

DISCUSSION

It is unusual for scoliosis to present and become rapidly progressive at the age of five years. This alerted our suspicions and made us look for a cause for the scoliosis. The only diagnostic clue was the widened cervical spinal canal.

Syringomyelia is most unusual in so young a patient and, although there is an established relationship between syringomyelia and scoliosis, it usually presents with peripheral neurological signs and symptoms (Williams 1979). This report emphasises the importance of seeking a cause if a disorder presents and behaves in an unusual manner and, as far as we can discover, is the first recorded case in which syringomyelia presented purely as a progressive scoliosis in the absence of any other neurological symptoms or signs.

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