DORSOLUMBAR SPINE DUPLICATION

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We report a case of dorsolumbar spine duplication in a female patient. She had several vertebral anomalies, such as fused vertebrae, hemivertebrae and butterfly vertebrae, together with duplication of the spinal column in the lower thoracic and the whole lumbar tract. Clinically, she had no control of the sphincters, but her gait was only slightly affected. This is probably the third reported case of this rare anomaly.

Keywords : embryogenesis ; spinal duplication ; vertebral anomalies.
Mots-clés : embryogénèse ; duplication vertébrale ; malformations rachidiennes.

INTRODUCTION

Although duplication of the neural component of the spinal column is relatively common (1), duplication of its bony component has only rarely been reported.

A case of duplication of the cervicothoracic spine, associated with cervical meningocele, atrophy of the muscles of the left arm and hand, wide short neck, L5 spondylolisthesis and sacrum acutum has been studied (4). The multiple vertebral anomalies could have been produced by a mutant gene (4).

Recently, Keller *et al.* (2) reported a female infant with an anterior keel-shaped calcified mass, extending from the sacral promontory up to the diaphragm. The mass consisted of mature bone, cartilage and joint-like spaces. The abnormality was associated with multiple gastrointestinal and genito-urinary tract anomalies.

We report a case of vertebral column duplication. The dorso-lumbar spine anomaly was associated with long-term survival and remarkably few symptoms.

CASE REPORT

The patient came to our observation at the age of 22 years. She was born at term, after an uneventful pregnancy to nonconsanguineous parents. Maternal history was unremarkable.

Fifteen days after birth, a right paramedian mass of about 8 cm in diameter had been surgically removed from the lumbar region. Its relationship with the spinal cord was difficult to ascertain. Histological examination revealed a lipoma and a cyst lined with a membrane of unclassifiable origin. The mass was embedded in smooth and skeletal muscle and peripheral nerve bundles.

The girl showed delayed motor development and started to walk autonomously at 4 years. She had to wear orthotics, as she was not able to stand erect. At age 10 she could walk without aids quite satisfactorily. At 22, she gave a one-year history of increased fatigability of the lower limbs, with some difficulties in performing fine voluntary movements.

On examination, she had very long lower limbs when compared to the trunk. The right leg was hypotrophic, and she had a right flat foot, which was smaller than the left. Mobility of the right leg was normal, while mobility of the right foot

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was only possible in flexion-extension. The right lower limb was 2.5 cm shorter than the left one. The left leg was hyposthenic, with increased muscle tone, left foot clonus and patellar hyperreflexia. No sensory impairment was evident. Motility of the lumbar spine was grossly restricted.

Ambulation was possible only for about 100 meters, owing to easy fatigability. The patient was using two sticks to walk, but no calipers or orthotics. There was no voluntary control of the sphincters.

Radiographs at 3.5 (fig. 1) and at 22 years (fig. 2 and 3) showed multiple malformations, consisting of fused, hemi- and butterfly vertebrae. The vertebral column of the lower thoracic and of the whole lumbar tract was fully duplicated, and L5

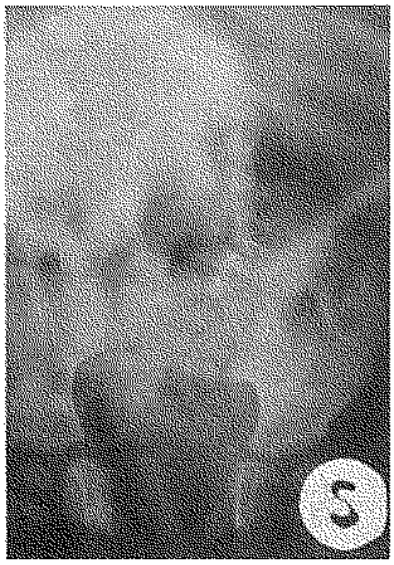

Fig. 1

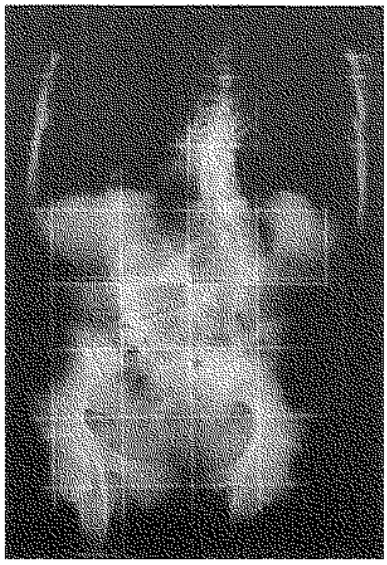

Fig. 2

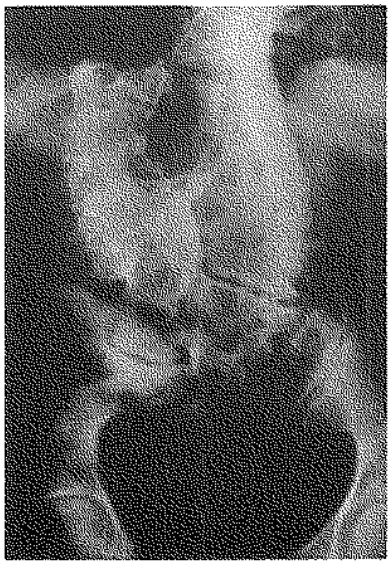

Fig. 3

Fig. 1. — Anteroposterior view at 3.5 years. The lower dorsal and the lumbar vertebrae are duplicated, and a bony ring is thus constituted.
Fig. 2. — Anteroposterior view of hips, chest and dorsal, lumbar and sacral spine at 22 years.
Fig. 3. — Detail of fig. 2. An ossified bony ring in the lumbar region is evident. A tentative identification of the vertebrae forming the ring is given in the diagram.

articulated on the left with the sacrum. On the right side, L5 articulated with the iliac crest via a neo-articulation.

The left iliac bone and the right half of the sacrum were hypoplastic. The later roentgenograms show that practically all the lower dorsal and lumbar vertebrae were fused in a huge mass, constituting an irregular bony ring.

DISCUSSION

Vertebral anomalies can be explained by the embryology of the vertebral column. *In utero*, between the third and sixth week, somite formation occurs. This is followed by the appearance of chondrification centers between the sixth and the seventh week. Ossification starts at week eight (3). If any of these processes is disrupted, vertebral anomalies can arise. By the 18th day of gestation, the definitive notochord is established as an independent structure. The notochord acts as an organizer, inducing the formation of the neural tube (3). The notochord is also a focus for the sclerotomes which migrate to enclose both the notochord and the neural tube, giving rise to the spinal column.

Unfortunately, our patient refused to undergo further investigations, such as sacculoradiculography, CT or MRI scans, which could have clarified whether the primary anomaly arose from a notochordal or a neural tube defect.

If a double meningeal space had been revealed, a neural tube defect could have been hypothesized. In this case, there would have been segmental doubling, and each segment would have influenced the development of a part of the vertebral canal.

If no double meningeal space had been found, the etiopathogenic insult would have been to the notochord. There could have been a fissuration, giving rise to a rachischisis, which would ultimately produce a spine duplication.

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SAMENVATTING

G. CAPASSO en N. MAFFULLI. Dorsolumbale duplicatie.

De auteurs rapporteren een geval van duplicatie van de dorsolumbale wervelkolom bij een vrouw. Zij had meerdere andere wervelanomalieën, zoals wervelblokken, hemivertebrae en vlindervormige wervels, samen met een ontdubbeling van de wervelkolom, laag thoracaal en over het ganse lumbaal segment. Klinisch had zij geen controle over de sfincters, maar de gang was praktisch normaal. Deze beschrijving van een zeldzame congenitale afwijking is hoogstwaarschijnlijk de derde in de literatuur.

RÉSUMÉ

G. CAPASSO et N. MAFFULLI. Duplication vertébrale dorso-lombaire.

Les auteurs présentent un cas de duplication vertébrale dorso-lombaire chez une femme. Elle présentait plusieurs anomalies rachidiennes, telles que blocs vertébraux, hémivertèbres et vertèbres "en papillon" ; il existait aussi une duplication de la colonne vertébrale au segment thoracique inférieur et sur toute l'étendue de la colonne lombaire. Cliniquement elle était incontinente mais la marche était pratiquement normale. La description de cette rare anomalie est très probablement la troisième dans la littérature.