Open quiz

Answers must be received by Dr. Jacobson no later than June 15, 1987. The solution will be published in Volume 16, Number 5, July 1987. Individuals from the department or practice from which the case originated will not participate in the quiz. Thus, cases chosen for the quiz will not come from any of the cases presented at the closed meetings of the Society.

Case report 422

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Photographic and radiological features

Fig. 1. A A frontal photograph of the patient at the age of 19 as a soldier is presented. B The patient is shown at the age of 35 years after the loss of 31 cm in body height

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Fig. 2. A A lateral plain film of the thoracic spine shows flattening of most of the vertebral bodies, severe kyphosis and virtually a horizontal course of the upper segment of the spine. Marked osteopenia (osteoporosis) is present. B Plain film tomography of the thoracic spine in the lateral view shows changes similar to those described in A. The flattening of most of the vertebral bodies is profound and the deformity of the spine is extremely severe.

Fig. 3. Another plain film lateral tomogram (this time of the lumbar spine) shows fish-bone-like indentations of the vertebral end plates – again with marked flattening of the vertebral bodies and extensive osteopenia. Because of the sharpness of the end plates the changes have the appearance of osteoporosis.

Clinical information

This 37-year-old man has been followed for 8 years. Fig. 1A is a photograph of the patient at the age of 19 years as a soldier and Fig. 1B at the age of 35 years. It is obvious that the patient had lost total body height to the extent of about 31 cm. From the clinical history it was retrospectively assumed that the disorder began at the age of 25 years. The shortening of the spine led to an almost 90 degree angulation of his sternum. The deformity of his thoracic spine caused his head to appear to be resting directly on his shoulders. The crutches led to fractures of both humeri. The loss of height was primarily due to shortening of the spine with flattening and wedging of the vertebral bodies of the thoracic spine, accompanied by increased kyphosis and fish-bone-like indentations of the end plates, particularly in the lumbar spine (Figs. 2 and 3).

Fig. 4 shows the thoracic spine at the age of 28 years (9 years before this admission), when the vertebrae still showed no gross changes, but the osteoporotic pattern was obviously beginning.

Fig. 5 showed that the skull was somewhat osteoporotic, but Wormian bones were not identified, militating against the diagnosis of osteogenesis imperfecta.

In addition to the fractures of both humeri, four more spontaneous fractures occurred. These were in the right scaphoid, the right second metatarsal and the right and left femoral necks. As a result of these fractures the patient became totally unable to walk and at the time of his most recent presentation, in order to move around his apartment, he had to use an office chair on wheels.

The two curves in Fig. 6 show the progressive loss of bone mineral from the radius over the course of eight years, measured by $^{125}$iodine photon absorptiometry. At the distal $\frac{1}{10}$ point the mineral content was reduced to less than forty percent of the normal age range. The three separate points indicate the percentage of the compact region of the total area of the right second metacarpal. The course is similar to the upper curve of the mineral content of the radius in the region of the $\frac{1}{3}$ point.