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A 36-year-old woman presented with an unusual deformity in her hands that began ten years ago. She noticed thermal sensory loss in her hands. On examination, her fingers were thickened, nails deformed, and there were slowly healing ulcers and fissures in her palms (Figure 1). On neurological examination, there was touch, thermal and pain sensory loss limited to her arms. The cranial nerves were normal. Tendon reflexes were normal in the arms, and exaggerated in the legs. Hoffmann and Babinski signs were elicited bilaterally.

Routine urine examination and blood counts were normal. Fasting blood glucose was 86 mg/dl, blood Venereal Disease Research Laboratory (VDRL) was non-reactive and the X-Ray of the chest was normal. She was not diabetic, and did not have any history of exposure to sexually transmitted diseases.

The X-ray of the hands showed resorption of the terminal phalanges (Figure 2).

Examination of the spinal cord by magnetic resonance imaging (MRI) confirmed Chiari malformation associated with syringomyelia (Figure 3). No other abnormalities were noted.

Treatment consisted of a suboccipital craniectomy and C1 laminectomy with duraplasty sufficient to decompress the cerebellar tonsils. The patient was discharged on the fourth postoperative day and recovered without any complications.



FIGURE 1: The clinical appearances of our patient's hands.



FIGURE 2: The plain radiographs of the fingers demonstrated the flexion contractures of distal interphalangeal joints and the resorption of the distal phalanges of the left hand. There were no irregularities of internal structures or periosteal reactions in the distal phalanges.

## SURGICAL TECHNIQUE

The patient was placed in prone position with her head moderately flexed. A linear skin incision was performed in the midline, from the inion down to the C3, that followed by suboccipital craniectomy with total C1 laminectomy. After durotomy in a Y-shaped fashion, cerebellar tonsils were exposed. The arachnoid was opened and obvious adhesions were cleared. Duraplasty was performed with allograft.

**Question:** Based on the patient history, physical and radiologic examination, which of the following is the most likely diagnosis?

- A. Multifocal osteomyelitis
- B. Leprosy
- C. Morvan's disease
- D. Raynaud's disease



**FIGURE 3:** MRI of cervical spine. A) Sagittal T2 image of the cervical and upper thoracic spine shows syrinx in the spinal cord.