

Hand Involvement in Primary Biliary Cirrhosis in an 85-Year-Old Woman

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An 85-year-old woman presented 16 years ago with a 6 month history of pain and swelling in proximal interphalangeal (PIP) joints and distal interphalangeal (DIP) joints of both hands. Laboratory investigation revealed an elevated sedimentation rate of 70 mm/h, positive antinuclear factor of 1:100, and elevated alkaline phosphatase 209 IU/l; rheumatoid factor was negative. Hand radiographs showed soft tissue swelling at the 5th PIP joint and a cystic swelling over the head of the proximal phalanx. Subsequent investigation of her liver function abnormalities led to a diagnosis of biopsy proven primary biliary cirrhosis (PBC).

Over the next 16 years she experienced episodic attacks of synovitis of her PIP and DIP joints. Her chronic PBC was also accompanied by xerostomia, pruritus, and deteriorating liver function.

On May 5, 2000, she presented again with acutely

swollen PIP and DIP joints. There was no clinical evidence of psoriasis, scleroderma, or hyperparathyroidism. Radiographs of the hands taken supine (Figure 1A) reveal predominant interphalangeal joint involvement with pencil and cup deformity noted at several joints (solid arrowhead). The 4th PIP joint of the left hand (Figure 1B) is enlarged to highlight the pencil and cup deformity. There is sparing of metacarpophalangeal joints in both hands. Chondrocalcinosis is present in both wrists (Figure 1A, open arrowhead). There is sparing of metacarpophalangeal joints in both hands. Figure 1C reveals marked erosions at the right ulnar styloid and right distal radioulnar joint (curved arrow), and large erosions with mild surrounding sclerosis within the lunate were also noted (arrow).

The changes at the PIP joints differ from those observed in rheumatoid arthritis by the presence of the pencil and cup



Figure 1A. Anteroposterior view of right and left hands.



Figure 1B. Left 4th PIP joint.



Figure 1C. Posteroanterior view of the right hand.

deformity. Erosive changes are asymmetrically distributed in the distal small joints of the hands, which is characteristic of this condition. The differential diagnosis for the DIP joint findings in a woman of this age is osteoarthritis, but the absence of any significant changes at the first metacarpophalangeal joint would be unusual if this were the case. In addition, other reports of a deforming arthropathy secondary to PBC describe similar PIP and DIP joint findings as in our

patient^{1,2}. In summary, these images show the advanced dramatic changes of hand involvement secondary to PBC.

REFERENCES

1. Clark M, Sack K. Deforming arthropathy complicating primary biliary cirrhosis. *J Rheumatol* 1991;18:619-21.
2. O'Connell DJ, Marx WJ. Hand changes in primary biliary cirrhosis. *Radiology* 1978;129:31-5.