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BRACHYMESOPHALANGY AND LOOSE BODIES IN THE METACARPOPHALANGEAL JOINTS

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A family with autosomal inherited brachymesophalangy is presented. Some of the family members also had loose bodies in the metacarpophalangeal joints. This condition is similar to osteochondritis in other joints. New loose bodies may be formed after operative removal and arthrotic changes may occur. The patients were not able to perform hard physical work with their hands.

Key words: brachydactyly; brachymesophalangy; osteochondritis dissecans

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Short fingers or brachydactyly is due to anomalous development of any of the contributing phalanges or metacarpals and is the most thoroughly studied congenital abnormality of the hand (Kelikian 1974). Brachydactyly has been conveniently classified into five main groups but there is considerable variation of the anomaly (Chen 1978). The most common type of brachydactyly is brachymesophalangy, which exists when the middle digital phalanges have the same length or are shorter than the terminal phalanges (Witt et al. 1966).

Osteochondritis dissecans is very rare in the hand. A few cases in the scaphoid bone have been reported (Meves & Schneider-Sickert 1975). Recently Andrén et al. (1978) described a combination of brachymesophalangy and osteochondritis dissecans in the metacarpophalangeal joints. This report presents the second family to be described in the literature with this rare syndrome.

PATIENTS

For some years we have had the opportunity to examine several members of a family with brachymesophalangy. The family members can be easily recognized by their fingers even by laymen. It has therefore been possible, by interviews with some members of the family, to set up a pedigree (Figure 1). All members of the first three generations, except two (III-6 and III-7), are dead, while all members of the following generations are alive. However, the family members are very scattered geographically and it has not been possible to examine all members. Five of the examined family members had, in addition to brachymesophalangy, loose bodies in the metacarpophalangeal joints and these patients will be referred to below.

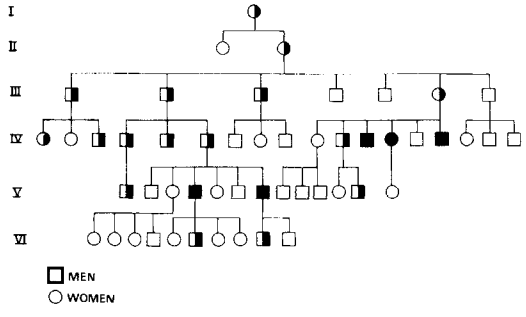

Figure 1. Pedigree showing a six-generation kindred; the half filled symbols represent members with brachymesophalangy but without recognized loose bodies in the finger joints, while filled symbols include members with brachymesophalangy and loose bodies.

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Table 1. Abnormalities of the hands

Number in pedigree		IV-12	IV-13	IV-15	V-4	V-7
Sex		M	F	M	M	M
Age		52	48	41	30	25
Brachymesophalangy	II	+	+	+	+	+
	III	−	+	+	−	−
	IV	+	+	+	+	−
	V	+	+	+	+	+
Deformity of metacarpal heads		+	+	+	+	+
Metacarpal joints with loose bodies: Right hand	II	+	+	+	+	−
	III	+	−	+	+	+
	IV	−	−	−	−	−
Left hand	II	+	−	+	+	−
	III	−	−	+	−	−
	IV	−	−	−	+	−
Short styloid ulnar process		+	+	+	+	+

RESULTS

The main abnormalities of the hands are summarized in Table 1. All five patients had brachymesophalangy of the second and the fifth fingers whereas the third and the fourth fingers were not affected in all cases (Figures 2 and 3). The length of the metacarpals was normal but the metacarpal heads were flattened and occasionally irregular. This deformity was most pronounced in the second and third fingers and to a lesser degree in the fourth and fifth fingers. As seen in the Table the majority of loose bodies were located in the second and third metacarpal joints. The loose bodies were usually discoid (Figure 4). In one case the loose body was examined histologically and consisted of normal bone surrounded by cartilage. All the patients had a short and blunt ulnar styloid process.

Clinically the patients had pain in the

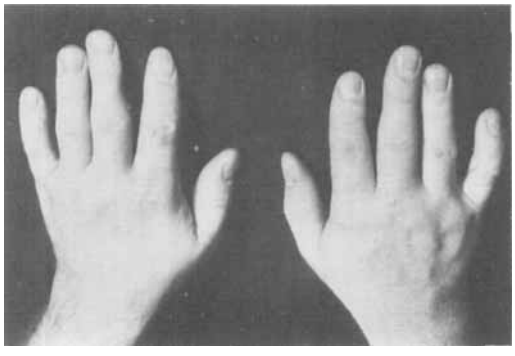

Figure 2. Patient with short index and little fingers.

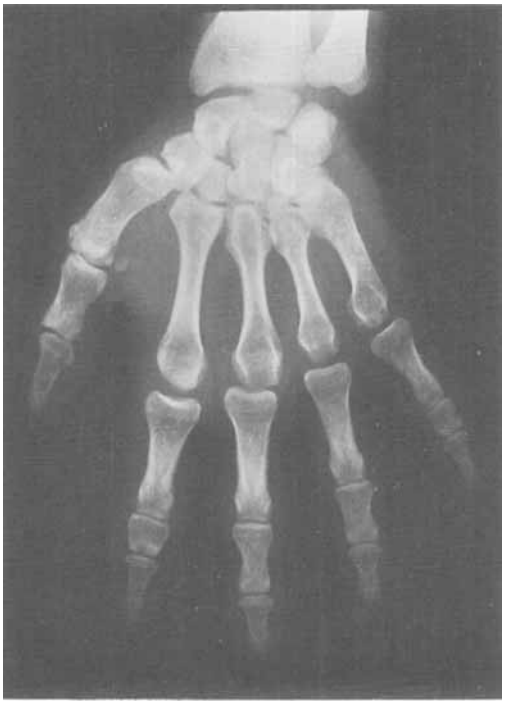

Figure 3. X-ray of the same patient as in Figure 2 demonstrating the short middle phalanges of the index and little fingers.

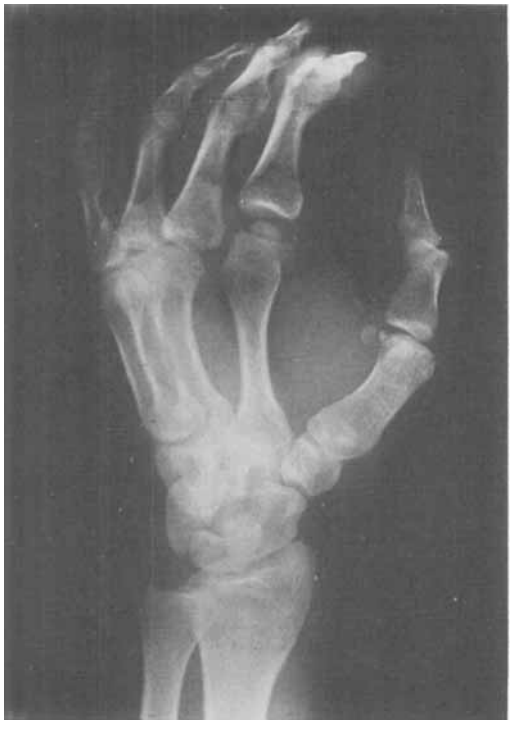

Figure 4. A loose body is present in the metacarpophalangeal joint of the index finger.

affected joints especially if they tried to perform hard work. There was episodic mechanical blocking of the joint motion. The examination revealed varying degrees of looseness and restricted movements of the joints with loose bodies.

The loose bodies were removed by operation except in one patient, who had only mild complaints. The operation resulted in disappearance of the mechanical blocking and the pain was reduced. However, in one patient (IV-15) three joints were reoperated on because of new symptoms. In one joint a loose body, formed since the last operation, was removed. In the two other joints no free bodies were found and the pain could be explained by arthrotic changes. Another patient (IV-12) was reoperated on in one joint because of pain but only arthrotic changes were found. Clinical examination of patient V-4 half a year after the first operation showed a new loose body in the second metacarpophalangeal joint but the symptoms were so mild that the patient did not want reoperation.

These five patients also had short or absent middle phalanges of the toes, but no loose bodies were found. The feet were without symptoms. The patients did not present other congenital abnormalities.

DISCUSSION

In the present family brachymesophalangy appears as an inherited autosomal dominant disorder and this is in agreement with other studies (Kelikian 1974, Andrén et al. 1978). In addition to being an isolated anomaly, brachymesophalangy may be part of a generalized disorder (Kelikian 1974). The predisposition to shortening of the phalanges is greater the later the enchondral ossification occurs (Witt et al. 1966), and therefore the middle phalanges of the index and the little fingers are most often affected (Kelikian 1974). The present syndrome which includes brachymesophalangy, deformed metacarpal heads, loose bodies in the metacarpal joints, hypoplasia of the ulnar styloid process and brachymesophalangy in the feet, in general, is consistent with the syndrome described by Andrén et al. (1978). However, in contrast to the latter description our patients did not have clinodactyly and some, besides the dwarfing of the index and little fingers, also had short middle phalanges of the third and the fourth fingers.

The aetiology of loose bodies in the finger joints is unclear, but the condition is rather similar to osteochondritis dissecans in the knee. This process is defined as separation of a cartilage-bone fragment from the joint surface (Smilie 1960). Accordingly, the microscopy of a loose body in the present study demonstrated that it consisted of bone and cartilage. It may be suggested that the deformity of the metacarpal heads plays an important role in the formation of the loose

bodies. This is consistent with the fact that the majority of the loose bodies were found in the second and third metacarpophalangeal joints, in which the greatest deformity of the metacarpal heads occurred.

Loose bodies were not seen in children in the sixth generation, but first occurred in adults, and were, in some cases, formed again after primary removal. Our patients did not present any history of trauma. Milgram (1977), however, suggests that osteochondritis dissecans is, in all cases, due to osteochondral fractures.

The prognosis of the present syndrome is rather dubious. In addition to recidivism of the loose bodies there is a tendency for the development of arthrosis, and none of our five patients were able to perform hard physical work with their hands.

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