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FAMILIAL OSTEOCHONDRITIS DISSECANS AND CARPAL TUNNEL SYNDROME

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A short-statured family – mother, son and daughter – each with osteochondritis dissecans (O. D.) of the elbow and two with O.D. of the knee is described. The mother and her siblings had, in addition, carpal tunnel syndrome.

Key words: familial–carpal tunnel syndrome; osteochondritis dissecans; short stature

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Osteochondritis Dissecans (O.D.) is a condition in which osteocartilagenous fragments become separated from the articular surface of the ends of the long bones. Trauma appears to be the most important aetiological factor, but in certain susceptible patients it may appear without predisposing injury (Hanley et al. 1967).

Several reports refer to a familial form of the condition (Hanley et al. 1967, Pick 1955, Stougaard 1964). It has also been described in association with developmental abnormalities including abnormally short stature (White 1957). This association with dwarfism has occasionally been noted to be familial (Pick 1955). Carpal tunnel syndrome is another condition which occasionally shows a familial tendency (Tanzer 1959).

This report describes the occurrence, in the same family, of O.D. affecting members of short stature who show, in addition, the features of carpal tunnel syndrome.

CASE REPORTS

Three members of a family are described, the mother (MG), son (AG) and daughter (SG).

Case 1

AG was a 16-year-old male (138 cm in height, below 3rd percentile; 60 kg in weight, above 97th percentile). At the age of 9 years he was investigated for short stature. His weight age (97th percentile) was found to be twice his height age (3rd percentile). Radiological survey showed normal bone development, the serum protein bound iodine measurement was 6.9 μg per cent and the fasting blood sugar 82 mg per cent. No specific cause was found for his stunted growth. AG was initially referred to the Orthopaedic Department aged 14 years in October 1974 with a complaint of pain in his right elbow after a fall 1 year previously. Radiographs showed the presence of O.D. of the capitellum. Treatment was conservative until November 1977 when operation for removal of a loose body was performed, after recurrence of symptoms. He had no other joint complaints but radiographs also revealed O.D. affecting the left elbow and the knee.

Investigations included renal function evaluation, serum calcium, phosphate, alkaline phosphatase, total proteins and thyroxine estimations, oral glucose tolerance test and urinary amino acid chromatography. They were all normal. Chromosome studies showed a normal 46, XY karyotype. Bone age was normal as measured by the Tanner-Whitehouse score index.

Case 2

SG was a 12-year-old female (133 cm in height, below 3rd percentile and 64 kg in weight, above 97th

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percentile). She was initially seen in October 1975, aged 10 years, with pain in the right elbow after a fall 10 days previously. Radiographs showed O.D. of the capitellum. Her symptoms subsided and she was discharged in September 1976. She was referred to Orthopaedic Out-patients in August 1977, this time with intermittent pain affecting the left elbow and left knee. Radiographs showed that she had O.D. affecting the capitellum of both elbows and the medial femoral condyle of both knees. There were no symptoms associated with the right knee. Apart from her height and weight, there were no other abnormal findings. Her periods had started at the age of 11 years and breast development was normal. All the investigations listed under AG were carried out and found to be normal.

Case 3

MG was a 39-year-old female (138 cm in height and weighing 68.2 kg) she presented in 1974 with symptoms of bilateral carpal tunnel syndrome and surgical decompression was carried out. In November 1977, she returned with pain and increasing stiffness of the right elbow. She had a history of pain in both knees when in her teens and at that time her family doctor diagnosed Osgood-Schlätter's disease. Her knee symptoms had subsided spontaneously. Radiographs revealed O.D. of both elbows with loose body formation and mild osteoarthrosis (Figure 1). Although the typical defects of O.D. were absent from the knees, there were changes of osteoarthrosis in both knee joints, mainly affecting the patello-femoral compartment. Radiographs of the hip joints were normal, but there was considerable irregularity and increased density around the lower lumbar vertebrae and early calcification in the spinal ligaments. Further investigations including biochemical, chromosomal and bone age studies, as for Cases 1 and 2, revealed no abnormality. Her husband declined to attend for interview but he was said to have no joint complaints.

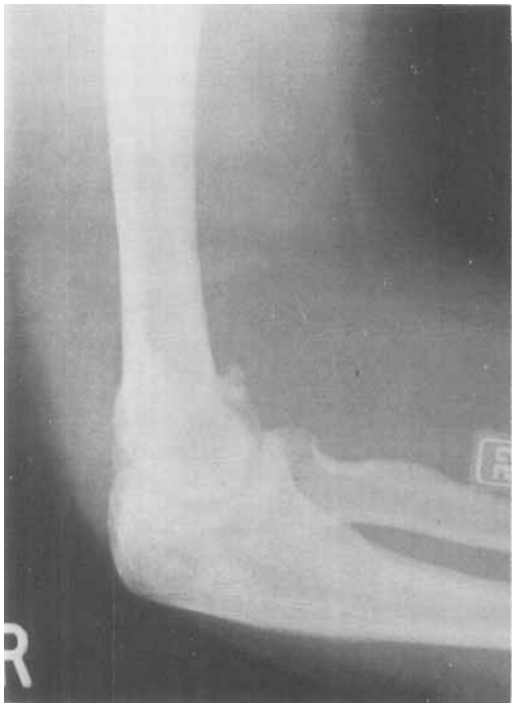

Figure 1. Osteochondritis dissecans of the right elbow in patient MG.

The relatives of MG were scattered throughout the country; a questionnaire was sent to the surviving nine of MG's ten siblings (one male, eight female, age range 27–50 years). All were of normal height and weight and of the seven who replied, four complained of pain and/or stiffness affecting either the knees or elbows. Radiographs were obtained from only one of the four symptomatic siblings and these were normal; radiographs were obtained from only one of the three asymptomatic siblings and these were also normal (Figure 2). Two of the siblings, both female, had had surgery for carpal tunnel syndrome (one bilateral, one unilateral) and two others (both female) had symptoms typical of this condition (Figure 2).

DISCUSSION

This family consisting of mother and two children all had O.D. affecting the capitellum. In addition, the daughter was shown to have O.D. affecting both knees, and the son, O.D. of one knee. The mother had osteoarthrosis of both knees, but no obvious radiological evidence of old O.D. of the medial femoral condyles. It is likely that the members of this family fall into the category of juvenile osteochondritis in which the lesions are often multiple and the condition may be familial (Smith 1960). Smillie believes that there is a dysostotic constitutional background in such families leading to the development of an accessory ossification centre which is susceptible to trauma (Smillie 1960).

White described three unrelated cases who were of short stature and in whom there was evidence of endocrine imbalance (White

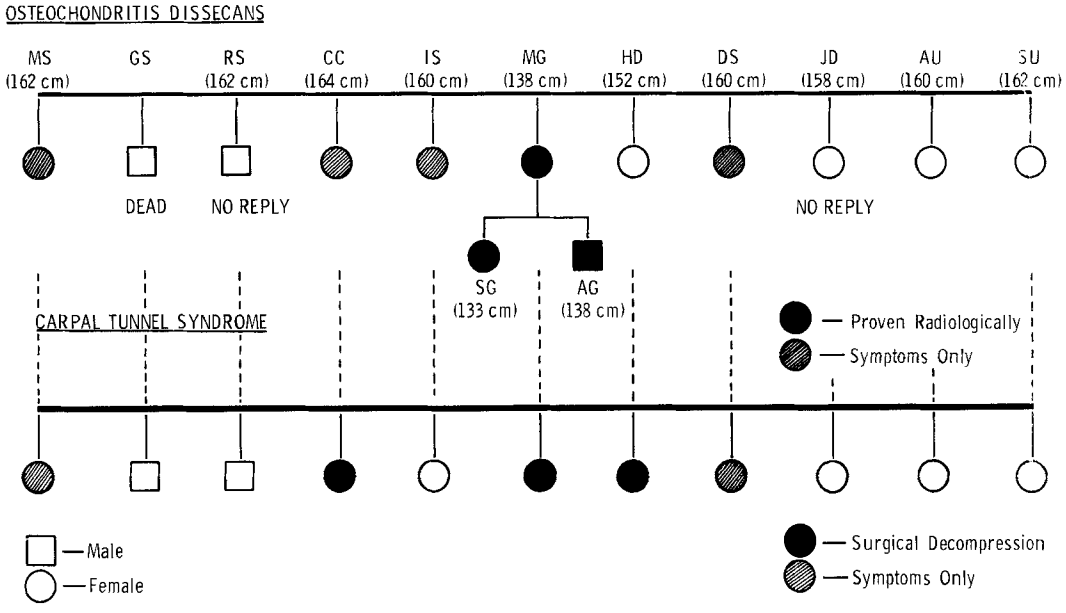

Figure 2. Familial incidence of osteochondritis dissecans and carpal tunnel syndrome.

1957). Hanley described two brothers affected by the condition who also demonstrated several developmental abnormalities and suggested that endocrine factors might play a role in the aetiology (Hanley et al. 1967).

The association with short stature was reported by Pick in a family who were otherwise normally developed, although no endocrine investigations were carried out (Pick 1955). None of the endocrine abnormalities reported by others were found in the patients reported here. It is likely that the condition is incidental to some undetermined hereditary disturbance of epiphyseal growth as suggested by Smillie (1960), although bone development was normal in all three patients as measured by the Tanner-Whitehouse index. Apart from the members of this family, four of the mother's siblings complained of pain or stiffness of elbows or knees. Radiographs were obtained in only one case and were normal. The siblings were of normal height and weight.

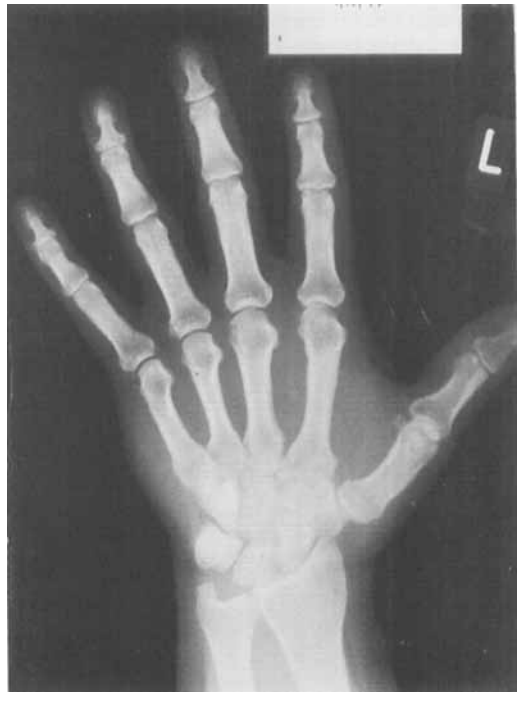

Figure 3. Radiograph of the left radio-carpal joint in patient MG.

Carpal tunnel syndrome was noted in the mother and four of her siblings. Tanzer described four cases of carpal tunnel syndrome, each of whom had one affected parent; in one case, a grandparent was also affected (Tanzer 1959). A familial incidence has been noted by others (Zabriskie et al. 1935, Stephens & Welch 1956, Phalen & Kendrick 1957).

Both O.D. and carpal tunnel syndrome are well recognised entities but the aetiology of these conditions is uncertain. Although both may be associated with an underlying anatomical abnormality, subsequently susceptible to trauma, there was no radiological abnormality of the radio-carpal joint in one such patient (MG) (Figure 3). Whether the two conditions are coincidental in members of this family or not is uncertain, but it is possible that a common constitutional factor may increase the possibility of their development.

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