OSTEOCHONDRITIS DISSECANS IN ASSOCIATION WITH DWARFISM

JOHN WHITE, GLASGOW, SCOTLAND

From the Royal Infirmery, Glasgow

The cause of osteochondritis dissecans is still disputed. Axhausen (1912) suggested that the lesion could be embolic in origin, but Fairbank (1933) and Smillie (1951) believed that trauma was more likely to be responsible. The occurrence of multiple joint lesions, with a hereditary tendency, was demonstrated by Bernstein (1925), by Wagoner and Cohn (1931), by Gardiner (1955), and by Pick (1955) who all described families in which two or more members were suffering from the disease. The occasional occurrence of multiple joint lesions in association with dwarfism has been reported, and Roberts and Hughes (1950) noted that one of their patients was a dwarf. Pick mentioned that the members of the family he described were under average height. No biochemical investigations have been recorded except in the one case described by Hay (1950).

The existence of a constitutional factor underlying certain types of osteochondritis dissecans is now accepted, but no evidence has been produced to identify the nature of such a factor, although Roberts and Hughes suggested that it might be an endocrine disorder.

Three patients with osteochondritis dissecans, all well under average height, in each of whom several joints were affected, are reported here as further evidence of a constitutional upset in this condition.

Case 1—Woman aged twenty-eight years, only four feet four inches tall. The general appearance of the patient (Fig. 1) was that of a much younger person, and the secondary sex characteristics were poorly developed. Her hair was scanty and fine, the spine showed a
mild kyphoscoliosis, and there were numerous pigmented striae on the abdomen. The family history was of no significance, and her parents and two sisters were of average height. At puberty the patient complained of aching in her knees, hips and elbows, but it was not until the age of nineteen that she first complained of locking and instability of the right knee. This was due to a loose body in the joint, which was demonstrated radiographically and subsequently removed (Fig. 2). At operation the articular surface of the patella was found fragmented and an area of osteochondritis dissecans was noted in the medial femoral condyle. Two years later similar symptoms occurred in the same knee which necessitated another arthrotomy. Recently the patient complained of pain in the right hip, radiating down the inner side of the thigh to the knee, and a radiograph of the hip showed a loose body near the lower surface of the femoral head (Fig. 3). Radiographs of the thoracic spine revealed evidence of previous osteochondritis juvenilis (Fig. 4).

Investigations—Because of the large number of biochemical tests undertaken, only abnormal results are recorded. Total serum protein was 7.9 grammes per 100 millilitres. Serum albumin was 2.8 grammes per 100 millilitres, serum globulin 5.1 per 100 millilitres, serum potassium 5.8 m.eq. per litre, serum calcium 3.4 m.eq. per litre, plasma inorganic phosphorus 5.2 milligrams per 100 millilitres. Follicular stimulating hormone was 40 mouse units per diem. The basal metabolic rate was 4 per cent below standard. A radioactive iodine test for thyroid activity showed a mild impairment of thyroid function and there was also a low flat glucose tolerance curve. All tests for renal function were normal. Gynaecological examination under anaesthesia showed a hypoplastic uterus, and microscopy of the endometrial mucosa revealed that the stromal changes were more fully developed than glandular ones for the phase of the cycle. These tests suggested an impairment in thyroid and parathyroid function with underdevelopment of the sexual glands.
Case 2—Man aged twenty-one years, four feet six inches tall. The patient (Fig. 5) had a high-pitched voice and the secondary sex characters were poorly developed. The family history was irrelevant, and his parents and three sisters were above average height. There was no previous illness of note, except for pains in the large joints and thoracic spine at puberty. At the age of sixteen he complained of pain in the right elbow, and radiographic examination showed an area of osteochondritis dissecans in the capitulum. Later the elbow locked and a loose body was removed from the joint, the site of origin being the capitulum (Fig. 6). Within a year similar symptoms occurred in both knees; loose bodies were demonstrated radiologically and removed at operation (Figs. 7 to 9). Recently a second operation was performed on his left knee, from which three loose bodies were removed. Although the articular surface of the patella was rough and fragmented the loose bodies appeared to arise from an area of osteochondritis dissecans in the lateral condyle of the femur (Fig. 7). A radiograph of the thoracic spine showed evidence of previous osteochondritis juvenilis (Fig. 10).

Investigations—Total serum proteins were 6.8 grammes per 100 millilitres. Serum albumin was 3.1 grammes per 100 millilitres, serum globulin 3.7 grammes per 100 millilitres, serum calcium 5.4 m.eq. per litre, plasma inorganic phosphorus 5.6 milligrams per 100 millilitres; 17-ketosteroids were 7.3 milligrams per diem. Lipid phosphorus was 1.65 milligrams per 100 millilitres. Fasting serum total fatty acids were 2,340 milligrams per 100 millilitres as stearic acid. All tests for thyroid and renal function were normal. A flat glucose tolerance curve was recorded. There was no gross abnormality in the external genitals, but he
admitted diminished libido. These tests suggested hypofunction of the sex glands with a mild metabolic upset.

Case 2. Figure 7—Left knee with osteochondritic changes in the lateral condyle of the femur. Figure 8—Right knee showing deformity of the patella from osteochondritis of the articular surface.

Case 2. Figure 9—Skyline view of left patella showing osteochondritic changes and loose body formation. Figure 10—Osteochondritis juvenilis of the thoracic spine.

Case 3—Woman aged twenty-one years, four feet five inches tall. When the patient was first seen her appearance was that of a child about eight years old (Fig. 11). Although late in
appearing, the secondary sex characters were normal, but her menstrual periods were scanty and irregular. The rest of the family were of average height, but a cousin suffered from loose bodies in both knees, and was of diminished stature. Initially the patient was referred from a rheumatic clinic because of pain in both elbows, and radiographs showed osteochondritic changes in the capitula of both joints (Fig. 12). Some months later the left elbow locked and radiographs showed a loose body in the joint (Fig. 13). Later she noticed similar symptoms in the right elbow, and a loose body was demonstrated radiographically. The elbows were explored and the loose bodies removed. A year later loose body formation necessitated a second operation on the left elbow. Radiographs of the right knee and ankle were taken after she had had symptoms in these joints, and further evidence of osteochondritis dissecans was found, but no loose bodies were seen (Figs. 14 and 15). This patient also had osteochondritis juvenilis in the thoracic spine (Fig. 16).

Investigations—Only the plasma inorganic phosphorus (46 milligrams per 100 millilitres) and the total fasting serum fatty acid (568 milligrams per 100 millilitres) showed abnormal figures. All tests for renal and thyroid function were normal. Gynaecological examination under anaesthesia revealed nothing abnormal, but microscopic examination of the uterine mucosa showed proliferative endometrium with a tendency to a cystic pattern. No conclusions were drawn from this patient’s biochemical tests, but it is interesting to note that the plasma inorganic phosphorus was elevated in all three cases.
Case 3—Right knee illustrating roughness of the articular surface of the patella and osteochondritis dissecans of the medial femoral condyle.

Fig. 14

Case 3. Figure 15—Right ankle showing an osteochondritic lesion in the lateral aspect of the articular surface of the tibia. Figure 16—Mild osteochondritis juvenilis of the thoracic spine.

DISCUSSION

During their studies of endocrine dwarfism, Schaefer, Strickroot and Purcell (1939) were struck by the frequency of single and multiple chondro-epiphysial disturbances associated with hypothyroidism. Among the epiphysial disturbances that they listed was osteochondritis juvenilis of the thoracic spine, and this was a feature common to the three cases described above. Schaefer et al. also maintained that in a number of instances this endocrine imbalance adjusted itself as the patients reached maturity.
OSTEOCHONDritis DISSECANS IN ASSOCIATION WITH DwarFISM

Although the first of my three patients alone showed impairment of thyroid and parathyroid function, there was some evidence to support the diagnosis of a mild endocrine imbalance in the second patient. There was no biochemical evidence of a glandular disturbance in the third case, but a careful study of the course of the disease suggested that she had passed through phases like the other two patients. It is notable that the condition was heralded in each case at puberty by a generalised aching in the joints in which loose bodies subsequently occurred, and that all the patients had a raised inorganic plasma phosphorus.

I would like to suggest that in these cases the underlying constitutional disturbance was an endocrine imbalance at puberty, which adjusted itself in varying degrees as the patients matured. This adjustment possibly accounts for the variation in the biochemical investigations in each patient in later life.

SUMMARY
1. Three patients suffering from osteochondritis dissecans in several joints, and all below average height, are described.
2. There was evidence of a constitutional upset in each case.
3. It is suggested that there was an underlying endocrine imbalance at puberty.

I wish to thank Mr J. Patrick for permission to report these cases, and Dr J. C. Eaton, in charge of the Biochemistry Department, for his help in the biochemical investigations.

REFERENCES