Forearm deformities in multiple cartilaginous exostoses

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Abstract. Sixteen patients with 20 forearm deformities were reviewed. The deformities were classified into three types. The degree of ulnar tilt of the radius, ulnar displacement of the carpus, and relative ulnar shortening were determined. The severity of the deformity correlated with these measurements. Metacarpal lengths were also measured. Significant metacarpal shortening without exostoses was seen in 10 of 11 patients and with exostoses remote from the metaphysis in 10 of 13 patients. Metacarpal shortening correlated with the type and severity of deformity.

Key words: Multiple cartilaginous exostoses – Forearm deformities

Multiple cartilaginous exostoses is a well-known disorder of enchondral bone manifested by bony prominences capped with cartilage and often complicated by defective modeling and retardation of bone growth. The forearm is affected in as many as 60% of patients with this disorder, but abnormalities in this region have received little attention in the radiologic literature. This study examined the location of the exostoses and the associated deformities in the forearm and also found significant metacarpal shortening in these patients. These observations provide an insight into the pathogenesis of the disorder.

Materials and methods

We reviewed the radiographs of 33 patients with multiple cartilaginous exostoses from the Children's Hospital and the Rehabilitation Centre for Children in Winnipeg. Sixteen patients had complete radiographs of the forearm available for study.

Deformities were classified into three types [9]. Type I involved ulnar shortening and bowing of the radius (Fig. 1). In Type II, ulnar shortening was associated with radial head dislocation (Fig. 2). Type III displayed relative radial shortening (Fig. 3). The degree of deformity was assessed by determining the radial articular angle, carpal slip, and relative ulnar or radial shortening. The radial articular angle is the angle between a line drawn along the radial articular surface and another one drawn perpendicular to a line that bisects the head of the radius and passes through the radial edge of the distal radial epiphysis. This angle is normally between 15° and 30° (Fig. 4A) [2]. Carpal slip is determined by the percentage of contact of the lunate with the radius. A line is drawn from the center of the olecranon towards the ulnar edge of the distal radial epiphysis. This line normally bisects the lunate (Fig. 4B) [2]. The degree of radial or ulnar shortening was assessed by determining the distance from the distal metaphysis of the radius to the distal metaphysis of the ulna and comparing this measurement with normal standards [4].

In all patients in whom metacarpals were radiographed, the metacarpal lengths were also measured and compared with normal standards [3]. Shortening was considered significant if the length of the metacarpal was more than 2 standard deviations below the mean.

Results

The patients ranged in age from 3 to 17 years at the time of their radiographs. Five patients underwent serial examinations. Ten were male. Fourteen of the 16 patients had forearm deformities, which were bilateral in 6. Type I was the most commonly seen deformity, observed in 16 forearms. Three deformities with a type II deformity were identified. Only one type III deformity was seen.

In patients with a type I deformity, the more severe the radial bowing, the greater were the measured carpal slip, radial articular angle, and ulnar shortening (Figs. 1, 2B). In the five cases in which serial radiographs were available, the deformity progressed until epiphyseal closure (Fig. 1).

There were not enough type II deformities to establish any significant correlation of carpal slip, radial articular angle, or ulnar shortening with the amount of radial bowing in these patients. All three of our examples of type II deformities were associated with exostoses of the proximal ulna (Fig. 2A).
Fig. 1A, B. Type I. Patient 12, left forearm at 9 years of age (A) and at 14 years of age (B). There is shortening of the ulna with progressive bowing of the radius.

Fig. 2A, B. Patient 11, 9 years of age. Right forearm (A) shows type II deformity with ulnar shortening, proximal ulnar exostoses, and lateral dislocation of the radius. Left forearm (B) shows mild type I deformity.

Fig. 3. Type III deformity in an 11-year-old boy. Note the shortened radius.

Fig. 4A, B. Methods of measurement of (A) radial articular angle (RAA) (see text) and (B) carpal slip (CS) (see text). From [2] with permission.