ARTHROGRYPOSIS CONGENITA: REPORT OF A CASE

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Arthrogryposis congenita is a medical curiosity. It is rare and authors who have written on it count with their fingers the cases they have seen. We are reporting a case which was confined in the nursery ward of the Philippine General Hospital.

CASE REPORT

S.J., a 35 yr. old G3P2, Filipino housekeeper from Malate, Manila was admitted in the Obstetrical ward of the Philippine General Hospital on June 20, 1957. She had moderately severe preeclampsia but with no signs of hydramnios or oligohydramnios. After 24 hours of labor, she delivered spontaneously a baby boy weighing 2740 grams with a head and chest circumference of 33 cm. and 30.5 cm. respectively. His cry was like the soft crow of a rooster. He had slightly narrowed fontanelles but the suture lines in the skull were not fused.

The deformities presented in the upper extremities were dorsiflexion of the wrists, flexion contracture of the elbows and slight bowing of the right arm due to fracture of the humeral shaft. The lower extremities showed characteristic bilaterial hip abduction and outward rotation, flexion contracture of the knees and the heels almost touched the buttocks. The extremities had a firm muscular tone.

The x-ray skeletal survey revealed a thin right humerus fractured at the middle and slightly underdeveloped right radius and ulna. The rest of the skeleton was within normal limits except for the malposition of the lower extremities.

Corrective casting of the lower extremities with gradual wedging at the knee and ankle was started on the 4th week. The upper extremities were placed in corrective splints, changed weekly to produce extension of the elbow and the wrist.

However, the course of the baby in the ward was stormy. Feeding was always by gavage as the baby never sucked. He frequently became cyanotic and febrile. There was no tracheoesophageal fistula on barium examination of the upper gut.

On the 4th month, he developed persistent rises of temperature and died ten days after its onset. Autopsy revealed acute bilateral pneumonia.

DISCUSSION

There has been much conjecture about the etiology of this condition. The theory of direct intrauterine compression has been advocated because some had a history of hydramnios or oligohydramnios. On the other hand, there is also a good reason to accept primary germ variation as the immediate cause.

It was Otto in 1841 who first described this entity and termed it congenital myodystrophy. Since then, it has acquired many synonyms, namely: amyoplasia congenita, myodystrophia fetalis, multiple congenital articular rigidity and arthrogryposis congenita. The most widely used name is the last, meaning arthros-joint and grypos-crooked.

The synonyms have been given on account of the involvement of both the skeleton and soft tissues, but primarily the latter. There are multiple joint contractures and periarticular changes but there is no bony aplasia nor any evidence of primary malformation of the bones from errors of suppression or differentiation.

The significant pathological feature is the aggregation of fat and degeneration of muscle fibers, resembling myodystrophies of later life. This muscle atrophy is not due to neurogenic involvement and seems to occur rather late in embryonal life when the muscle fibers are already fully differentiated. Some, however, may present degeneration of cells of the anterior and posterior horns of the cord and changes in the white matter of the brain.

The outstanding clinical feature is the rigidity of one or more joints, usually a number of the larger joints. This contracture never gives the impression of an absolute bony block but is always stringy, no matter how greatly the motion may

be restricted. The contracture may be of the flexion type as in this case or extension type or a combination of the two. The extension of the knees may be of such severity sometimes that it resembles extreme genu recurvatum. Pain is characteristically absent. Sometimes the overlying skin is tense and shiny due to lack of motion producing lymphangiectasis.

Among the conditions to be differentiated from it but rarely with difficulty are congenital club foot, scleroderma and spasticity due to central nervous system disorders.

MANAGEMENT

There can be no generalization made as to the form of treatment as cases differ so much in degree and type. However, by far the best results are obtained by conservative treatment.

In general, the success of nonoperative management is much greater in the very young than in the older infant. The flexion contractures are more resistant than the extension contractures. The clubfoot deformity yields better to corrective measures than the ordinary congenital talipes because of the atrophy of the muscles and ligaments.

The conservative means employed are traction, splints and corrective casts. Gradual wedging of the cast is done till the desired result is obtained.

The obstacles in the correction of the flexion contracture as in the case discussed are the danger of subluxation of the knee; the tension produced by the corrective efforts upon the sciatic nerve and popliteal vessels should also be considered.

Operative treatment calls for operations upon the soft tissues as muscle lengthening, tenotomies and capsulotomies and on the bones as osteotomies. Postoperatively, traction devices are necessary and should be continued until the correction is absolutely stable. To prevent deformities from recurring as much as possible after operation, the position of correction must be secured by braces. Exercise should also be done to provide a powerful stimulus for the development of the greatly weakened muscular apparatus.

However, in conclusion, no treatment is satisfactory and when the deformities are corrected, they have a tendency to recur. Some improvement may be expected but even after the deformities are lessened, the muscle power is more often so slight that joint function cannot be anticipated.

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