Congenital Pseudarthrosis of the Clavicle: Report of Three Cases

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Congenital pseudarthrosis of the clavicle is a rare benign condition with only one report in the radiologic literature [1]. There have been 107 case reports of this entity, although it is probably much more common than the literature suggests. Three additional cases are reported here, and the clinical, radiologic, and surgical spectrum of congenital pseudarthrosis is reviewed.

Case Reports

Case 1

A newborn boy, with no history of birth trauma and a product of a normal pregnancy and delivery, was referred to the x-ray department for chest examination because of a grade II out of VI systolic ejection murmur. Physical examination was normal except for the cardiac murmur and a palpable bump over the right clavicle. There was no evidence of tenderness, swelling, or restriction of motion in the right clavicle.

Chest radiography (fig. 1) revealed findings typical of congenital pseudarthrosis of the clavicle with overriding of clavicular fragments. The sternal segment was seen above the clavicular segment.

Case 2

A 2-year-old girl, product of a normal pregnancy and delivery, had a visible bump at the midportion of her right clavicle and a diagnosis of fracture was made. The parents stated that the child had never been uncomfortable on palpation over this bump, and there was full unrestricted range of motion of the right upper extremity. The child continued to have motion at the midportion of her clavicle, although the area was very prominent on abduction of the right shoulder. Right shoulder radiography revealed changes typical of congenital pseudarthrosis. Diagnosis of congenital pseudarthrosis of the clavicle was later made when the child was seen by her physician for a totally unrelated orthopedic problem (fig. 2). No treatment was necessary.

Case 3

A 10-day-old infant was seen for multiple tetramelic deformities consisting of syndactyly and clefts. Family history was negative for similar anomalies, and there was no history of teratogenic agents during pregnancy nor other abnormalities of pregnancy or delivery. The referring orthopedist’s diagnosis of congenital pseudarthrosis of the right clavicle was based on a palpable mass at the midportion of the clavicle; the child was asymptomatic on shoulder motion. Subsequent radiography showed the characteristic findings (fig. 3).

Discussion

Congenital pseudarthrosis of the clavicle was first described and classified as an entity distinct from cleidocranial dysostosis by Fitzwilliams in 1910 [2]. Since then, most of the case reports have been in the orthopedic literature, with only one reference in the radiographic literature [3-5].

The entity is extremely interesting in that it almost always occurs on the right side of the patient. In 10%, involvement is bilateral [3]. There have been three cases of left-side congenital pseudarthrosis, but one of these...
had associated dextrocardia [3]. The reason for the predominant right-side involvement is unclear, but several theories have been advanced. Lloyd-Roberts et al. [6] believe there is a relation between high right subclavian artery and pressure changes on the right side of the clavicle due to either cervical ribs on the right side or a high right subclavian artery itself.

The etiology of congenital pseudarthrosis of the clavicle is not clear. Fracture is an unlikely possibility because redness, tenderness, and a history of birth trauma are almost always absent. Usually there is no family history elicited, although one-third of Gibson’s and Carroll’s [5] 27 patients did have a family history. If in fact a family history can be elicited, the disorder is believed to be inherited as a Mendelian recessive gene.

In examining the embryology of the clavicle, some clues as to a possible etiology are found. The clavicle is the first bone to ossify in the human body at 5–6 weeks [7]. Some authors [8, 9] suggest that there are two centers of primary ossification of the clavicle, and that the failure of these two centers to coalesce gives rise to this condition. Other authors [5] claim that there is only one center of ossification of the clavicle, and the formation of pseudarthrosis is due to improper bony reabsorption of this center.

Regardless of the etiology of congenital pseudarthrosis of the clavicle, clinical diagnoses are usually quite similar. Diagnosis is usually made at birth or several months later. The patient’s family notices a lump of increasing size, which is described as a prominent spike under the skin in the midportion of the clavicle. The most significant clinical feature is the absence of pain, which is critical to differentiation from traumatic nonunion. In traumatic nonunion, pain is the predominant feature [3]. Occasionally, there is some discomfort from activity, which is another reason the patient seeks medical help. A constant physical finding in congenital pseudarthrosis is that the sternal segment always overlies and is above the acromial segment. Café au lait spots are not a feature of congenital pseudarthrosis, although one case report indicated several spots with no other stigmata of neurofibromatosis. The absence of other congenital anomalies is also a constant finding in clinical evaluation of these patients. Case 3 in our series is quite atypical in that there were several teratogenic defects.

Radiographic diagnosis of pseudarthrosis of the clavicle is easy. The middle one-third of the clavicle is always involved, and the condition is usually close to the junction of the clavicle and the lateral third. The segments are not in continuity and the ends of the bone are enlarged. There is no callus present and no reactive bone. As mentioned before, the medial segment always overlies the lateral segment.

Differential diagnoses of congenital pseudarthrosis of the clavicle are primarily cleidocranial dysostosis and posttraumatic pseudarthrosis. Cleidocranial dysostosis usually affects other bones and midline structures. There are skull defects, bilateral clavicular defects, pelvic defects, and a very strong family history. Posttraumatic pseudarthrosis is a less distinct differential diagnosis. This entity is always painful; there is always tenderness on pressure and on motion of opposing fracture fragments [3]. Radiography of posttraumatic pseudarthrosis usually shows exuberant callus and the ends of the bone are quite bulbous.

The gross pathologic findings of congenital pseudarthrosis are constant. The ends of the two bone segments are covered by cartilage and often separated by fibrous tissue. There are no pathologic features of neurofibromatosis or fibrous dysplasia.

Surgical treatment of congenital pseudarthrosis is straightforward, consisting of excision of the cartilaginous cap, alignment of the fragments, and occasionally bone grafting or internal fixation. At least one-half of the cases of congenital pseudarthrosis of the clavicle are asymptomatic and need no further treatment.

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