COMMUNICATION FROM THE CRIPPLES' CLINIC AT GÖTEBORG
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AN EXTRAORDINARY CASE OF DEFORMITY OF THE WRIST (SYMPTOMATIC FORM OF MADELUNG'S DEFORMITY)

BY

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I shall report the case of a young man, a farm-hand, aged 17, seeking advice for a deformity of the right wrist. He has always been in good health previously, states that he has not had rachitis and that he was able to walk before he had completed his first year. He has eight brothers and sisters, who are all in good health; malformations or deformities have not occurred in any person of his family.

In November 1930, about 7 months before he appeared at this clinic for the first time, he began after heavy work with threshing to feel pain in his right wrist, gradually radiating throughout the forearm. To begin with the pain was rather severe and troublesome, forcing him to cease working. Thereafter the pain decreased gradually and recently he has not had any pain worth mentioning, provided he does not use the hand for too heavy work. But he believes that the strength of the hand has diminished and in conclusion he states that since he began to notice discomfort in the wrist the latter has assumed an increasingly deformed and abnormal appearance. This is the main reason why he seeks medical advice.

Present state of the patient:— A rather brawny youth of ordinary stature. No signs of a passed rachitis. The ulnar part of the right hand is in a marked volar position of subluxation so that the ulnar head, which is considerably enlarged — nearly the size of a walnut —, is very prominent on the dorsal aspect of the wrist; the ulnar part of the wrist gets a typical bayonet-

shaped appearance (Fig. 1). The skin is firmly attached to the bones and it is easy to palpate the whole of the distal articular surface of the capitulum and the inner side of the styloid process of the ulna, which nowhere is in contact with adjacent carpal bones. In its lower half the radius forms a faint curvature with the concavity in a volar and to some extent radial direction (Fig. 2).

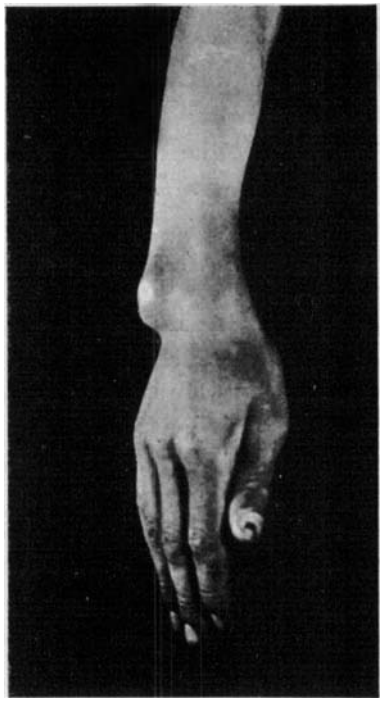

Fig. 1.

On the volar surface the flexor tendons are clearly visible as highly strained strings, especially the flex. carp. radial., running across the somewhat projecting multangulum majus, and the flex. carp. uln. inserted on the os pisiforme which is also very prominent (Fig. 3).

The right wrist is somewhat thicker than the left; the cir-

cumference, measured immediately below the process. styloid. ulnae, is 20 cm, the corresponding measure of the left wrist being 18 cm.

The mobility is as follows in the case of the right wrist:—		and in the case of the left wrist:—
Volar flexion	70°	65°
Dorsiflexion	20°	60°
Ulnar flexion	35°	30°
Radial flexion	0°	25°

There is thus a considerable restriction of the dorsiflexion and the radial flexion, and moreover the power of supination has decreased to about half the normal value reckoned from a position of complete pronation.

The gross muscular strength is distinctly diminished but has unfortunately not been measured by means of dynamometer.

X-ray examination (Dr. Renander) reveals a very marked defect, about 4 cm. long, in the distal end of the radius involving both epi- and metaphysis towards the ulna and forwards (osteitis fibrosa cystica?). Owing to the volar subluxation of the hand and the interference of the latter with the defect and owing to the ulnar and dorsal luxation of the ulna a deformity arises similar to that seen in *Madelung*'s disease (Fig. 4).

Treatment: The patient definitely refused to undergo any treatment whatever. The only thing that could be done was to enter him for a professional school at this city for training in his profession.

This is thus a case which can be said *clinically* in its broad features to afford a typical example of this rather rare and interesting occurrence that is termed *Madelung*'s deformity.

First and foremost there is the typical position of subluxation of the hand with the highly prominent and enlarged ulnar head; there is also the thickening of the carpus, however not so marked in this case as it may be in more advanced cases, in which it is said that the circumference can be found to measure double the normal measure of the wrist.

Moreover we find the characteristic highly strained tendons on the volar surface and a distinct, incipient protrusion of the carpal bones in a volar direction, in this case the os pisiforme and the multangulum majus; instead of the latter it is, however, more frequently seen that the os hamatum protrudes on the radial side. — There is also the typical curvature of the radius, even if it is not very marked. There is no measurable

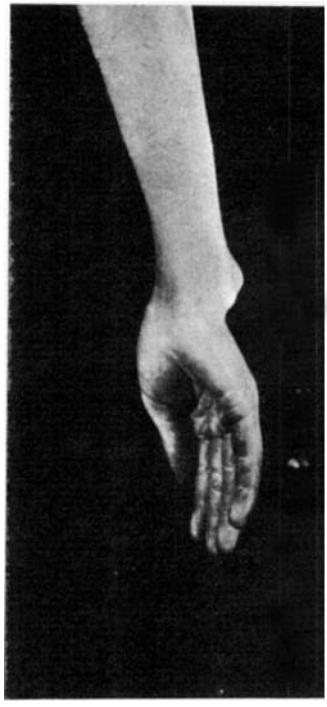

Fig. 2.

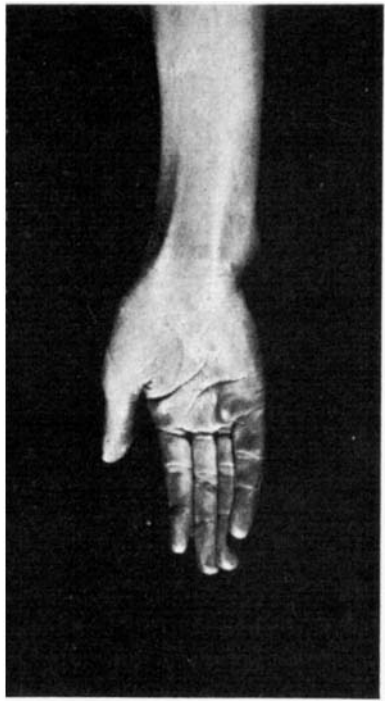

Fig. 3.

shortening of the radius, which, however, can be explained by the relatively short time during which the disease in all probability has lasted. According to *Kirmisson* at least two years pass until the disease has reached its full development, and in cases that have advanced so far a considerable shortening of the radius may be found, even up to 5 cm.

With regard to the mobility of the wrist we find in this

case the typical decrease of the power of dorsiflexion, the volar flexion being unimpaired or even somewhat increased. Of the movements from side to side the ulnar flexion is said to be more or less discontinued, whilst the radial flexion is said not to be influenced. In the present case the condition is, however, quite the reverse. The decrease of the power of supination is also a constant symptom.

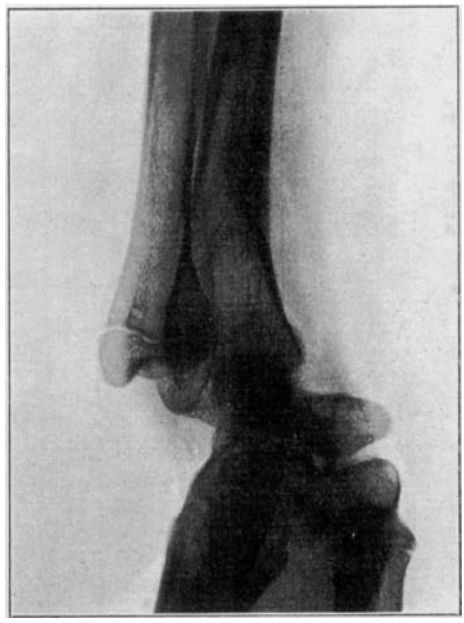

Fig. 4.

Lastly there is the decrease of the gross muscular strength, which is an almost constant phenomenon. This decrease of the strength is, however, not so much dependent on the disease itself, but is rather a direct consequence of the fact that the hand is in a position of slight volar flexion and cannot be dorsiflexed. Under these circumstances the strength of the hand will always decrease, and the greater the volar flexion is the greater will the decrease of the strength be owing to the shortening of the distance between the origin and the insertion of the flexors.

Reverting to the epicritical examination of the case I draw the attention to the fact that the disease apparently began spontaneously — the statement about overstrain in threshing is probably the usual attempt at finding a causal connection — with an initial painful stage and advanced slowly but steadily and that it began at the age that is said to be the typical age,

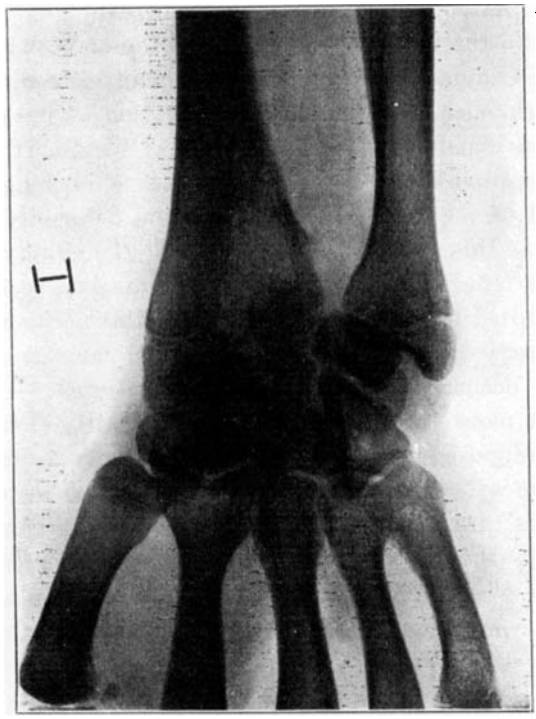

Fig. 5.

viz. late adolescence. So far it corresponds very well to the *true Madelung*'s deformity. The result of the X-ray examination, the destructive osseous process in the radius, however, sets aside the idea of this diagnosis and involves that we can only consider the case a symptomatic form of *Madelung*'s deformity.

According to *Ewald* all the cases are included under this group in which some cause of the occurrence of the deformity can be demonstrated, e. g. inflammatory (osteomyelitic) pro-

cesses in the bone, or fractures. As to the *true Madelung*'s deformity its etiology has scarcely been elucidated yet. The cause is, however, now considered to be dependent on a too early partial synostosis of the distal epiphyseal cartilage of the radius.

So far the case reported above may possibly be of interest, as it may be considered a support to the correctness of that view. For the epiphyseal cartilage is here almost completely destroyed by the destructive process in the bone, which in the case of the radius as a matter of course had to bring about the same consequence as a too early ossification of the epiphyseal line, viz. an impairment of the increase of length. It must thus be the disproportion between the increase of length of the radius and of the ulna that engenders the deformity.

As far as this too early ossification of the epiphyseal cartilage in the true *Madelung*'s deformity is concerned our knowledge is limited to suppositions. A congenital predisposition is, however, regarded as the most probable cause — cases of hereditary occurrence have thus been observed — moreover rachitis, or more exactly a kind of late rachitis (*Peter Berg*), has been supposed to be the cause, and lastly I would state that certain oft-repeated movements, i. e. one-sided overstrain of the arms, are supposed to play a part. It is for instance stated that the disease is comparatively prevalent among laundresses who wring linen all through the day; pianists, too, have been considered to be predisposed. The disease is much more frequent in females than in males.

Therapy: As already referred to the patient was unwilling to undergo any treatment »as he felt no pain now«, which unfortunately makes the case lose some interest.

As, however, an eventual treatment perhaps would coincide in its main features with the principles of the ordinary methods of treatment of *Madelung*'s deformity a short reference to these principles may be considered justified here.

During the first painful stage of development of the disease the hand should be allowed to rest. Among other things a supporting bandage is recommended which, when applied at a

comparatively early stage, is said to be able to prevent the deformity from becoming too marked. Moreover massage and electricity of the extensors is recommended in order possibly to increase their strength, thus letting them counteract the flexors. *Hoffa* has tried to supply the flexors with an artificial supplement of strength by means of application of an elastic bandage.

In the case of a fully developed deformity the chances of obtaining any improvement by means of conservative methods are, however, small, not to say non-existent. As a last resort surgical interference has, therefore, been recommended. Already in 1885 osteotomy was performed by *Duplay,* but the results were not so good, especially not from a cosmetic viewpoint. Later on *Streissler* introduced the curved osteotomy. In order to place the hand in a position of complete supination the pronator is even cut through now, and lastly resection of the distal part of the ulna is recommended. In this manner good results can be obtained, both with regard to function and to the cosmetic appearance.

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SUMMARY

It is the case of a young man clinically showing all typical signs of a true *Madelung*'s deformity of his right wrist. On X-ray examination it appeared, however, that the patient suffered from a destructive osseous process, an osteitis fibrosa cystica, localized to the lower end of the radius and even partly destroying the epiphyseal cartilage. The case must, there-

fore, be perceived as a symptomatic form of *Madelung*'s deformity.

It is considered worth while to publish it, as it possibly may be said to give support to the present perception of the etiology of the so-called genuine or true *Madelung*'s deformity.

The latter is nowadays considered to be dependent on injury in the epiphyseal cartilage of the radius, more exactly on a too early partial synostosis of the distal epiphyseal cartilage. The cause of this too early ossification is unknown; it has been supposed to be due to a congenital predisposition, heredity, or rachitis, and also to overstrain of the arms (laundresses). In contradistinction to this form, the genuine form, the cases in which some primary cause of the occurrence of the deformity can be demonstrated, e. g. inflammatory processes in the bone, tumours, and traumata, have according to *Ewald* been referred to one group, the so-called symptomatic form.

With regard to therapy it has been tried — apart from the removal of possible primary causes — in early cases to prevent the progress of the deformity by means of conservative methods, bandaging etc. In the more advanced cases correction by means of operation must be resorted to.

RÉSUMÉ

Il s'agit d'un jeune homme qui présente par l'examen clinique tous les symptômes typiques d'une déformité vrai de *Madelung,* siégeant au poignet droit. L'exploration par radioscopie a cependant fait constater que l'homme est affecté d'un processus destructif dans le tissu osseux (ostitis fibrosa cystica), localisé dans l'extrémité inférieure du radius et s'attaquant même partiellement au cartilage de l'épiphyse. Il convient donc de caractériser ce cas comme une forme symptomatique de la déformité de *Madelung.*

Nous avons trouvé utile de publier ce cas, comme il pourrait être considéré comme un appui pour la théorie maintenant

généralement adoptée, sur l'étiologie de la lésion appelée *déformité vraie* (ou genuine) de *Madelung.*

Comme on le sait, cette lésion est aujourd'hui regardée comme tenant primitivement à une abnormité dans le cartilage de l'épiphyse du radius, plus étroitement définie comme une synostose partielle et précoce du cartilage distal de l'épiphyse. On ignore quelle est la cause de cette ossification précoce; on a voulu la trouver dans une disposition congénitale, dans l'hérédité ou dans un rachitisme, ou bien dans un surmenage des bras pendant le travail (blanchisseuses).

Séparément de cette forme vraie, on a, selon Ewald, classé ensemble les cas où l'on peut indiquer quelque cause primaire qui aurait déterminé la déformité, telle p. ex. que des processus inflammatoires dans le tissue osseux, des tumeurs et des traumas. Ce groupe est nommé la forme symptomatique.

Quant à la thérapeutique — outre la suppression d'éventuelles causes primaires — on a essayé, dans les cas frais, d'enrayer le développement de la déformité par des moyens conservateurs, traitement par appareils, etc. Dans les cas plus avancés la correction opérative est indiquée.

ZUSAMMENFASSUNG

Es handelt sich um einen jungen Mann, der klinisch alle typischen Zeichen von echter Madelungscher Deformität an seinem rechten Handgelenk aufwies. Bei der Röntgenuntersuchung zeigte es sich jedoch, dass der Mann an einem knochenzerstörenden Prozess litt, osteitis fibrosa cystica, der sich am unteren Ende des Radius lokalisierte und sogar teilweise den Epiphysenknorpel zerstörte. Der Fall darf also als eine symptomatische Form von Madelungscher Deformität rubriziert werden.

Diese Tatsache schien der Veröffentlichung wert, da sie möglicherweise als eine Stütze für die augenblickliche Auffassung bezüglich der Aetiologie der sogenannten genuinen oder echten Madelungschen Deformität genannt werden kann.

Diese sieht man ja heutzutage als letzten Endes auf einer Schädigung des Epiphysenknorpels beruhend an, näher bestimmt als eine vorzeitige patielle Synostose im distalen Epiphysenknorpel. Die Ursache dieser vorzeitigen Verknöcherung kennt man nicht; man hat auf kongenitale Disposition, Erblichkiet und Rachitis geraten, aber auch auf eine einseitige Ueberanstrengung der Arme (Wäscherinnen). Im Gegensatz zu dieser genuinen Form hat man, nach Ewald, solche Fälle zu Gruppen zusammengestellt, wo man die eine oder andere primäre Ursache für die Entstehung der Deformität nachweisen kann, wie z. B. Entzündungsprozesse im Knochen, Tumoren und Traumata; die sogenannten symptomatischen Formen.

Was die Therapie betrifft, so hat man, abgesehen von der Behebung ev. erfindlicher primärer Ursachen, in beginnenden Fällen versucht, auf konservativem Wege, durch Bandagebehandlung etc., ein Fortschreiten der Deformität zu verhindern. In weiter fortgeschrittenen Fällen muss operative Korrektion eingreifen.