Fig. *Angiogram showing carotid-basilar anastomosis (long arrow) and dural fistula (small arrow)*

consistent with the hypothesis that dural arteriovenous fistula can be due to a malformation, as may be the case in the present observation. Indeed, the persistence of a trigeminal artery provides some indirect evidence that a structural defect may have predisposed to the disruption of the middle meningeal artery following a traumatic accident, which would have had little effect on a normal artery.

ANDREA SPISSU
*Clinica delle Malattie Nervose e Mentali,
Ospedale Civile,
Via Ospedale, 54,
09100 Cagliari*

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Pronator syndrome due to thickened bicipital aponeurosis

Sir: The entrapment of the median nerve at the elbow gives rise to a compression neuropathy commonly called the pronator syndrome.[1-5] We studied a patient presenting this symptom-complex caused by a thickened bicipital aponeurosis compressing the median nerve.

A 55-year-old brick-layer noted weakness of his left hand and numbness of the thumb, second and third finger, associated with pain at the left wrist, forearm and arm. These symptoms had an acute onset after a day in which the patient had carried heavy building material supported on his left forearm. When admitted, two months later, examination revealed a slight wasting of the thenar eminence and of the volar aspect of the forearm. Tinel's sign over the median nerve at the elbow was positive. There was weakness of the pronator teres, flexor digitorum sublimis, flexor digitorum profundus to the second and third finger, flexor pollicis longus, opponens pollicis, abductor pollicis brevis. The hand displayed a "benediction attitude" when the patient attempted to make a fist. Reduction of sensation was demonstrated over the median nerve distribution in the hand. Neurological examination failed to reveal any other abnormality. The electrophysiological study showed denervation and neurogenic atrophy in the flexor digitorum sublimis, flexor digitorum profundus, abductor pollicis brevis. Distal motor latency of the median nerve was 3·6 ms in the left side and 3·7 ms in the right side: motor conduction velocity across the elbow was respectively 48 ms^{-1} and 67 ms^{-1}. Radiography of the chest and of the left upper extremity was normal, as were a complete blood count and the sedimentation rate. At operation, the median nerve, explored in the antecubital fossa, was found to be entrapped beneath a thickened bicipital aponeurosis. Section of the structure exposed a flattened portion of the nerve in the site of the compression, and a swelling of the nerve trunk just above it. The recovery of the patient was complete in few months. The most common causes of the median nerve compression at the elbow are an hypertrophied pronator teres, the passage of the nerve under both its heads, and the kinking against the sublimis bridge.[2 4 5 7] In our patient, however, the weakness of the pronator teres muscle suggested that the entrapment of the nerve was indeed above the elbow. Besides acute traumatic incidents, some anatomical anomalies, such as a supracondylar process of the humerus, the Struthers ligament or a thickened bicipital aponeurosis can be causative factors of this syndrome.[5 7 8] Radiographs excluded the presence of a bony supracondylar process. Clinically it is not always possible to assess if an entrapment is caused by the Struthers ligament or by the bicipital aponeurosis. Our patient's symptoms represented the common features of the entrapment of the median nerve above the elbow, but did not give any suggestion about the structure restraining it.

Some manoeuvres are described which are supposed to be suggestive of the exact site of the compression:[2 7] the entrapment by the Struthers ligament is described as usually associated with forearm pain elicited or increased during forceful extension of the wrist, while reproduction of pain by resistence to forearm supination and elbow flexion is considered a positive sign for entrapment at the bicipital aponeurosis. Both these manoeuvres failed to demonstrate an increase of the forearm pain in our patient. The electrophysiological findings were consistent with a compression of the nerve in the region of the elbow[3 5-7] but they could not suggest the localisation of it.

Only the surgical exploration of the nerve in the antecubital fossa located the structure responsible for the entrapment, showing a thickened bicipital aponeurosis compressing the nervous trunk. Thus, thickened bicipital aponeurosis must be regarded as a rare cause of the uncommon syndrome of median nerve entrapment above the elbow.

P MARTINELLI
AS GABELLINI
M POPPI
R GALLASSI
E POZZATI
*Institute of Neurology,
University of Bologna
Via U. Foscolo 7-40123 Bologna,
and 2nd Neurosurgery Division
Bellaria Hospital,
Bologna, Italy*

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Matters arising

Should Charcot-Marie-Tooth disease be genetically subgrouped on motor conduction velocity?

Sir: Based on clinical, electrophysiological and morphological studies Dyck and Lambert divided patients with Charcot-Marie-Tooth disease into what they considered to be two different disorders, which were reported separately in two papers.[1][2] One was denoted hypertrophic because of the existence of onion bulbs (prominent segmental demyelination and remyelinations) whereas the other was designated neural and did not show onion bulbs. The dividing line in terms of motor conduction velocity (MCV) was set at 47, 45 and 40 m/s in the ulnar, median and lateral popliteal nerves respectively and individuals within any kinship would show similar velocities. A disorder of movement similar to that seen in patients with essential (familial) tremor was described in the hypertrophic variety only, as were plantar ulcers of the feet. Dyck and Lambert[1] also differentiated the hypertrophic Charcot-Marie-Tooth disease from Dejerine-Sottas disease. They reported two brothers with this latter disorder. One was chairbound and had a MCV in the ulnar and median nerves of 5 and 6 m/s respectively whereas the other, whose physical signs consisted only of absence of tendon jerks, showed a MCV of 42 and 44 m/s in the same nerves.

Credit and recognition should be given to Dyck and Lambert for drawing attention

to these important facts. However, it has become evident that (a) any clinical feature, including the disorder of movement similar to essential tremor,[3] and plantar ulcers of the feet,[4] could be seen in both types, (b) MCV in the lower limbs has proven to be of less value than in the upper limbs,[3-6] (c) internodeal length is of little value in clearly distinguishing subtypes,[5][6] (d) onion bulbs are also seen in the neural type.[6] Gradually the concept of two different Charcot-Marie-Tooth diseases has lost support. Patients having an hereditary motor and sensory neuropathy with the type of inheritance, natural history, symptoms and signs of Charcot-Marie-Tooth disease that Dyck and Lambert categorise into two different disorders were recognised to be a single entity.

Authors with the most experience with this disorder have put forward a genetic classification of Charcot-Marie-Tooth disease based on MCV[4][7] (concordance of conduction velocity in the upper limbs within each family). As with the original dividing line set by Dyck and Lambert[1][2] there were kinships with both types, Thomas and Calne[7] lowered the limit of MCV in the median nerve to 38 m/s, thus maintaining the division of the disorder into two genetically different types in terms of MCV. However, Harding and Thomas[4] have written that in their study there were examples where MCV in the affected individuals fell "out of the appropriate cluster." Other authors have found affected individuals of the same family with widely different MCV. Myrianthopoulos et al[8] (see table III of their paper) and Davis et al[9] (see kinship 27 in table II of their work) mentioned one family each and Brust et al[10] quoted six families. In a detailed study Salisachs et al[5] found that the MCV in the median nerves of two brothers with Charcot-Marie-Tooth disease were 48·3 and 28·3 m/s respectively. Teased fibre preparations (studied by Professor WG Bradley) showed similar changes in these two patients although segmental demyelination and remyelination were more marked in the latter. It is clear therefore that some kinships could not be categorised satisfactorily in the classification proposed by Dyck and Lambert[1][2] even after the alteration in the dividing MCV as made by Thomas.[4][7]

It is interesting that the limit set by Thomas and Calne[7] in the median nerve would qualify one of the two patients of the kinship with Dejerine-Sottas disease

reported by Dyck and Lambert[1] (see above) as having the neural type of Charcot-Marie-Tooth disease. Davis et al[9] studied many kinships and quoted others from the literature in which some affected members had MCV in the median nerve above 38 m/s whereas other cases of the same kinship had MCV values below this (see below the "intermediate" type, and fig 3 of their work). It is clear that in terms of classification, the lowering of MCV to 38 m/s in the median nerve has apparent advantages in some kinships but may not be helpful in others, since wherever this arbitrary dividing line is placed in terms of MCV there seem to be kinships with affected members above and below the line.

It has been claimed that although in Charcot-Marie-Tooth disease subgroups may exist, MCV is an inadequate means to define such subgroups.[10] Some authors, who are keen to maintain part of the division suggested by Dyck and Lambert and thus classify in terms of MCV alone, consider the existence of widely different MCV in affected individuals of the same kinships as "potential flaw" in the genetic classification of the disease.[4] Designating such findings as a "potential flaw" ignores the facts that (a) in few families have electrophysiological studies been made in several affected members, and (b) when such studies are available some have shown widely different MCV in the upper limbs in affected members. Bradley[6] grouped his cases according to MCV in the upper limbs into hypertrophic (<25 m/s), intermediate (25-45 m/s) and neural (>45 m/s). Harding and Thomas[4] found it impossible to accept such classification because there were kinships with both the intermediate and the hypertrophic type. In spite of the occurrence of both types in the same kinship,[5][8-10] surprisingly the same authors[4][7] accept the division between the hypertrophic and neural type made by Dyck and Lambert[1][2] at 45 m/s in the median nerve and now set at 38 m/s.

More evidence of the inadequacy of MCV as a means for the genetic classification of Charcot-Marie-Tooth disease may come from data on Refsum's disease. Indeed, in recessively inherited disorders it is common to find that the expression of the disease is similar between, and especially within, families; but minor differences, which do not warrant different classification, are not unusual. Thus, if Refsum's disease would ever present with widely different MCV in affected