NATURE OF OEDEMA IN PARALYSED LIMBS OF HEMIPLEGIC PATIENTS

BY

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The formation of oedema in the paralysed limbs of hemiplegic patients has been a well-recognized phenomenon for over half a century. Hare (1898) and Allen (1899) each described a case of "unusual oedema" in hemiplegia. In 1936 Luhan reviewed the literature and stated that 28 cases had been recorded; in addition he described in detail seven further cases. Although the mechanism of this type of oedema has not been clearly defined, it has usually been attributed to a disturbance of the blood vascular system in the affected limbs. Weiss and Ellis (1931) believe that the oedema results from an increase in capillary pressure which is in turn due to diminution in arteriolar tone in the paralysed limbs, while Bucy (1949) states that there is an increase in capillary permeability. We now produce new evidence that, in the upper limb at least, a major part in the formation of hemiplegic oedema is played by an impairment of lymphatic function. This evidence is based on a critical examination of the protein content of oedema fluids.

Clinical Features

Oedema in the paralysed limbs, occurring at some time during the clinical course, was noted in 21 patients in a series of 130 cases of hemiplegia. Thirteen of the oedematous patients had hemiplegia alone with no other important clinical condition, and in these cases it was noteworthy that the oedema was either confined to the arm or was more pronounced there than in the leg. In the remaining eight patients, who had other pathological processes besides hemiplegia contributing to oedema formation, the swelling was often more marked in the paralysed leg than in the arm. These clinical features are summarized in Table I.

TABLE I

Group	Clinical Condition	No. of Cases	Oedematous Limb		
			Arm Only	Leg Only	Arm and Leg
1	Hemiplegia alone ..	13	11	0	2
2	" with:				
	(a) Cardiac failure ..	4	0	2	2
	(b) Venous thrombosis of leg ..	4	0	2	2

In the 13 patients with hemiplegia alone the oedema was confined to the paralysed side; and in all but two of these only the upper limb was swollen. Where hemiplegia was associated with cardiac failure there was oedema of both legs, but it was more marked and more persistent on the paralysed side. The four patients in group 2b (see Table I) had a complete flaccid hemiplegia, and the phlebothrombosis developed while they were bedridden; two had oedema of the hand before the occurrence of venous thrombosis in the leg. In the absence of complicating factors oedema has been most easily detected on the dorsum of the hand; and volume measurements of the hands, using a water-displacement method (Eccles, 1956), have shown an increase of from 8 to 30% in cases tested. An 8% increase corresponded to the least detectable degree of pitting oedema.

The time of appearance of oedema after the onset of hemiplegia varied from 2 days to 10 weeks, but the most usual time was about two weeks after the paralysis. The duration of oedema was always more than two months (excluding patients who died in less than this period), and was sometimes as long as six months.

Although generally only patients with the most severe degree of paralysis developed oedema there were notable exceptions. In three patients oedema in the upper limbs appeared, and increased in amount at a time when the patients were making rapid recovery from hemiplegia with almost complete return of motor power. The duration of the oedema in these exceptional cases also appeared to be as long in the other patients, and in two female patients it persisted long after they had been discharged home able to perform their household tasks.

Results of Protein Estimations

Samples of oedema fluid were collected by miniature Southey-type needles inserted subcutaneously, and their protein content was estimated by an ultra-micro-Kjeldahl method (Crockett, 1956). In cases of uncomplicated hemiplegia the concentration of protein in fluids removed from oedematous hands varied from 1.2 to 4 g./100 ml., and the mean value for 13 patients was 2.8 g./100 ml. There was found to be a rough correlation between the protein content and the interval between the onset of hemiplegia and the occurrence of oedema. This is shown in the chart.

Relationship between protein content of oedema fluid removed from the hand and duration of paralysis in patients with uncomplicated hemiplegia.

Patients of group 2 had specimens taken from the paralysed oedematous leg and at the same time from the hand when it was oedematous. The results of these protein estimations are shown in Table II. In all cases the oedema

TABLE II.—Protein Content of Oedema Fluids Removed From Hand or Foot in Hemiplegic Patients with Either Congestive Cardiac Failure or Femoral Venous Thrombosis

Group	Clinical Condition	Case No.	Protein Content (g./100 ml.)	
			Hand	Foot
2a	Hemiplegia with congestive cardiac failure	4	2.2	1.3
		8	—	0.7
		9	—	1.3
		17	2.1	0.6
2b	Hemiplegia with femoral venous thrombosis	1	—	0.8
		13	1.8	1.4
		14	1.2	0.7
		20	—	1.7

fluid from the leg contained a lower concentration of protein than that from the arm. This may be attributed to the fact that oedema of the leg was in part due either to congestive cardiac failure or to femoral venous thrombosis, and these conditions produce an oedema fluid of low protein content.

Interpretation of Results

Normal capillary walls allow the escape of only a small amount of protein from the blood stream. Landis (1934)

calculated that normal capillary filtrate has a protein content of about 5% of the plasma protein level. It is probable that the protein of tissue fluid re-enters the blood vascular system only via the lymphatics (Drinker and Yoffey, 1941; Taylor *et al.*, 1957).

In a previous report (Crockett, 1956) it has been shown that an estimation of the protein content of oedema fluid is often of value in determining the cause of oedema. As the result of the investigation of fluids in a variety of clinical states it was found possible to distinguish three classes of oedema: the mechanical filtration oedemas with a low protein level (0.1 to 0.9 g./100 ml.) occurring in cardiac failure, hypoproteinaemia, and venous obstruction; and those in which high protein levels are found attributable either to a failure of drainage of tissue-fluid protein—namely, lymph-oedema and paralytic oedema (1 to 4 g./100 ml.)—or to an increase in the permeability of capillary walls as in burns and allergy (4 to 6 g./100 ml.).

In every patient with oedema due to hemiplegia alone without any other major pathological process, the oedema fluid has been found to contain a high concentration of protein. Similar high protein fluids have also been found in cases of post-traumatic stiffness and in patients with paralysis due to lower motor neurone lesions.

Impairment of Lymphatic Function

It has been suggested that oedema in hemiplegia is due to an increase in the permeability of the capillaries in the paralysed limbs (Bucy, 1949); our evidence does not support this view. We believe that the most important factor responsible for the development of oedema (with its high protein content) in "uncomplicated" hemiplegia is a relative lymphatic insufficiency. Some of the observations supporting this belief are:

1. The oedemas of hemiplegia and of lymphatic obstruction resemble each other in that they show a similar relationship between protein concentration and time. The increase in protein content with duration contrasts with the high protein content which is found *immediately* after the onset of such conditions as angioneurotic oedema and burns. In these conditions the oedema is due to increase in capillary permeability. (In fluid from one case of angioneurotic oedema the protein level was 4.2 g./100 ml. and in a burn of the hand it was 4.7 g./100 ml. In each case the sample was collected within 24 hours of the onset of oedema.)

2. The electrophoretic pattern of the oedema fluid in hemiplegia resembles that found in the low-protein oedemas—for example, cardiac oedema. A characteristic feature of this pattern is absent or greatly reduced α_2 -globulin. We have found that all types of oedema (including lymph-oedema) in which the capillary permeability is believed to be normal show this pattern. On the other hand, where the capillary permeability is increased, as in burns and allergic oedema, the electrophoretic pattern of the oedema fluid resembles that of serum and the α_2 -globulin is present in normal amounts.

3. In a number of cases of hemiplegic oedema we have determined the protein concentration of the capillary filtrate by an indirect method. The procedure was as follows:

The volume of the hand on the hemiplegic side was measured by means of a water-displacement tank (Eccles, 1956). A specimen of oedema fluid was taken for estimation of its protein content. A cuff was applied to the forearm and inflated to 40 mm. Hg. After four hours the cuff was removed and the hand volume and the oedema protein concentration were again estimated. The experiment was repeated on another two occasions, and further sets of protein and volume estimations were made. The capillary transudate protein concentration (p) is given by the formula

$$p(V_2 - V_1) = v(P_1 - P_2) + (P_2V_2 - P_1V_1)$$

When V_1 =initial volume of oedematous hand; V_2 =final volume of oedematous hand; v =hand volume less the volume of extracellular fluid in which the protein is dissolved; P_1 =initial oedema fluid protein; P_2 =final oedema fluid protein.

Example.—Values from three experiments in a patient with hemiplegic oedema of the hand.

A. $V_1=654$ ml.	B. $V_1=574$ ml.	C. $V_1=625$ ml.
$V_2=684$ ml.	$V_2=650$ ml.	$V_2=676$ ml.
$P_1=2.28$ g.%	$P_1=4.88$ g.%	$P_1=3.95$ g.%
$P_2=2.08$ g.%	$P_2=3.33$ g.%	$P_2=3.25$ g.%

Solution for v as simultaneous equations:

A & B, $v=450$ ml.	} Mean value for $v=445$ ml.
A & C, $v=434$ ml.	
B & C, $v=455$ ml.	

Solution for p assuming $v=445$ ml.

A, $p=0.70$ g.%	} Mean value for $p=0.72$ g./100 ml.
B, $p=0.69$ g.%	
C, $p=0.77$ g.%	

Normal hand volume was 500 ml.

This value for the protein content of the capillary filtrate ($p=0.7\%$) accords closely with that obtained by Stead and Warren (1944), who measured the protein content of the oedema fluid obtained in normal subjects by compressing the forearm at 30 mm. Hg for 12 hours. Their results showed that at a venous pressure of 30 mm. Hg the protein content of the transudate for normal capillaries was within the range of 0.4 to 1.3 g./100 ml.; the average value was 0.8 g./100 ml. Comparison with our results indicates that the permeability of the capillaries is not increased in the limbs of hemiplegic patients.

Immediate Cause of Oedema

Thus it seems probable that the fluid which enters the tissue spaces has the composition of normal capillary filtrate and contains only a very small amount of protein. In the course of time, owing to defective removal of protein by the lymphatics, the concentration of protein in the tissue fluid rises. The high extravascular protein concentration reduces the effective colloid osmotic pressure of the plasma, which is one of the chief factors preventing the loss of fluid from the blood stream. Landis (1934) has shown that a fall in plasma colloid osmotic pressure has quantitatively the same effect in increasing the rate of capillary filtration as a rise in capillary pressure. In all but two of the uncomplicated hemiplegic cases in the present series, the hand oedema fluid had a protein concentration greater than 2 g./100 ml. Electrophoresis shows the fluid protein to be proportionately richer in albumin than plasma, and therefore osmotically more active. It is probable that a protein level in excess of 2 g./100 ml. is sufficient in itself to cause oedema. There are, however, in nearly all cases other factors which increase capillary filtration rate and promote oedema formation.

Ancillary Factors Contributing to Oedema Formation

1. *Arteriolar Dilatation.*—Weiss and Ellis (1931) found that the surface temperature over the paralysed side in cases of hemiplegic oedema was increased, especially in the arm. Moreover, the difference between the oxygen content of the blood in the brachial artery and cubital vein was diminished on the paralysed side compared with the normal. They conclude that there is a disturbance of cerebral vasomotor regulation which leads to diffuse arteriolar dilatation, increased blood flow and capillary pressure, and a greater capillary filtration rate on the paralysed side. In two of our patients whose oedema appeared early there was a clinically detectable increase in skin temperature in the paralysed arm, and it is likely that an increase in the rate of transudation of fluid from the capillaries has been the result of arteriolar dilatation in these cases. Many observers have drawn attention to the difference in temperature on the two sides of the body in hemiplegic patients. Bucy (1935) states that early in cases of hemiplegia the temperature of the skin is commonly higher on the affected side than on the other; whereas in chronic cases the hemiplegic side is cooler. However, the proportion of patients showing these vasomotor disturbances is unknown.

2. *Local Increase in Venous and Capillary Pressure Due to External Causes.*—(a) *Dependency of paralysed arm:* In two cases oedema appeared while the patient was sitting in a

chair, and the most likely explanation is that the paralysed arm had been allowed to hang in a dependent position. (b) *Lying on the paralysed side*: Hemiplegic patients who are kept in bed often lie on the paralysed side in order to give more freedom of movement to the normal arm. In several cases this was thought to be a factor in causing oedema. (c) *Effect of splinting*: In one case oedema in the paralysed arm directly followed the application of a light "polythene" splint which was used to correct flexion deformity of the fingers.

3. *Congestive Cardiac Failure*.—Several authors have noted that hemiplegic oedema is more common in those patients who have previously experienced episodes of congestive cardiac failure (Weiss and Ellis, 1931; Luhan, 1936; Bucy, 1949). In the present series two cardiac patients (well controlled by digitalis and mercurials) developed hemiplegia and became oedematous on the paralysed side. In two other hemiplegic patients oedema appeared only after the development of congestive cardiac failure. In each case the oedema was more marked in the paralysed leg. When the cardiac failure was treated oedema was more persistent on the paralysed side than on the other; moreover, the protein content of the oedema fluid gradually increased—for example, in one case from 0.7 to 2.0 g./100 ml. Thus the oedema which initially has a low protein content owing to the predominant effect of cardiac failure may become converted to a high-protein oedema; the improvement in cardiac efficiency and the use of mercurial diuretics lead to a removal of water and electrolytes from the tissues, but the protein remains owing to the relative lymphatic insufficiency on the paralysed side.

4. *Thrombophlebitis*.—Four of the hemiplegic patients developed venous thrombosis in the paralysed leg. In two cases the patients had oedema of the affected arm prior to the venous thrombosis; subsequently, the affected leg became very much more oedematous than the arm. The protein levels of the oedema fluids were 1.2 and 0.7 g./100 ml. for the upper and lower limbs respectively in one patient and 1.8 and 1.4 g./100 ml. in the other. In the other two patients no oedema was present until the occurrence of the venous thrombosis, after which the leg alone became oedematous (protein levels of 0.8 and 1.7 g./100 ml.). In the absence of cardiac failure oedema of the paralysed leg and not of the arm, in the hemiplegic patient who is nursed in bed, is rare. Any patient who has remained recumbent following the cerebrovascular accident and who subsequently develops oedema of the paralysed leg alone should be suspected of having phlebothrombosis, and treatment should be instituted accordingly.

Discussion

The removal of the tissue-fluid protein requires effective lymphatic function, which in turn depends very largely on the activity of skeletal muscle. Lymphatic flow also varies with the position of the limb (McMaster, 1937); elevation considerably increases flow, and dependency will stop the movement of lymph in the main lymphatic trunks. In most cases of hemiplegia lymphatic drainage is sufficient to prevent the accumulation of protein in the tissues, provided that the rate of transudation of fluid from the capillaries is not increased. All the ancillary factors which have been considered above lead to an increase in the rate of capillary transudation, and in such circumstances the accumulation of fluid and protein in the tissue spaces occurs. The greater the extent to which these factors are concerned in the causation of oedema the lower will be the protein content of the oedema fluid. This we find to be the case; patients with congestive cardiac failure and hemiplegia have a lower protein content of their oedema fluid than those with hemiplegia alone. On the other hand, when the ancillary factors are operative to a minimal degree oedema will be long in appearing, but its protein content will be high by the time enough fluid has accumulated to be detected clinically.

In all patients in our series the oedematous limbs showed a soft swelling which pitted readily on pressure. Moreover,

there was no alteration in skin colour. Gilbert and Garnier (1897), however, described a case of "tumescence" of the hand and "un aspect éléphantiasique" in the leg of a patient 20 years after she had sustained hemiplegia at the age of 31. The hand was violaceous red in colour and it resembled "la main succulente" of syringomyelia, although there was no muscle-wasting and the motor power in the arm was comparatively good. They concluded that this condition may appear in long-standing hemiplegia and that it is due to vasomotor disturbances.

Raymond and Courtellemont (1904) consider that "la main succulente" with its firm non-pitting swelling is rare in hemiplegia, but a condition of "white oedema" occurs very much more commonly. Their patient developed swelling of the hand one hour after the onset of hemiplegia. On account of this very early onset, the good general condition of the patient, and the return of almost full movement in the arm they attribute the formation of oedema to vasomotor disturbances.

Many authors have postulated the existence of vasomotor centres which are involved in the cerebral lesion in cases of hemiplegia with oedema, and in the descriptions of the post-mortem examinations of the recorded cases particular attention is paid to the situation of the pathological changes in the brain. Parhon and Goldstein (1899) concluded that the vasomotor centres are contained in the corpus striatum, especially the head of the caudate nucleus. Luhan (1936), however, states that thus far results of clinical studies which have been checked with post-mortem observations have not furnished any critical evidence for localizing a cerebral vasomotor centre. In our series necropsy was performed on seven of the eight patients who died. Although the cerebral lesions were usually extensive the situation of the pathological changes was not essentially different from that generally found in cases of hemiplegia without oedema.

Summary and Conclusions

Oedema in the paralysed limb is noted in about 16% of cases of hemiplegia. It is usually more marked in the upper limb than in the lower, and it may be confined to the hand.

The oedema fluid has a high protein content, and it is believed that the most important factor in its causation is impairment of drainage of protein from the tissues by the lymphatics consequent upon the loss of muscular activity. The retained protein reduces the "effective colloid osmotic pressure" of the plasma and leads to accumulation of fluid in the extravascular extracellular compartment. Removal of oedema from the paralysed limb is favoured by encouragement of lymphatic drainage.

Contributory factors responsible for the development of oedema in hemiplegia include increased venous pressure caused by lying on the affected arm, a dependent position of the paralysed arm when it is flaccid, splinting, venous thrombosis, and congestive cardiac failure. Arteriolar dilatation due to a disturbance of vasomotor regulation in the brain may also play a part.

In the cases investigated we have been unable to find evidence of any increased permeability of the capillaries to protein in the hemiplegic side.

Hemiplegic patients who are confined to bed, and who subsequently develop oedema in the paralysed leg and not of the arm, should be suspected of having thrombophlebitis in the leg.

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REFERENCES

- Allen, C. L. (1899). *J. nerv. ment. Dis.*, 26, 471.
 Bucy, P. C. (1935). *Arch. Neurol. Psychiat.*, 33, 30.
 — (1949). *The Precentral Motor Cortex*, p. 29. Urbana, Illinois.
 Crockett, D. J. (1956). *Lancet*, 2, 1179.
 Drinker, C. K., and Yoffey, J. M. (1941). *Lymphatics, Lymph, and Lymphoid Tissue*. Cambridge, Mass.
 Eccles, M. V. (1956). *Brit. J. phys. Med.*, 19, 5.
 Gilbert, A., and Garnier, M. (1897). *C.R. Soc. Biol. (Paris)*, 4, 553.
 Hare, H. A. (1898). *J. nerv. ment. Dis.*, 25, 189.
 Landis, E. M. (1934). *Physiol. Rev.*, 14, 404.
 Luhan, J. A. (1936). *Arch. Neurol. Psychiat.*, 36, 42.
 McMaster, P. D. (1937). *J. exp. Med.*, 65, 347.
 Parhon, C., and Goldstein, M. (1899). *Roumanie med.*, No. 3. Abstract in *Rev. neurol.*, 1902, 10, 865.
 Raymond, F., and Courtellemont, V. (1904). *Rev. neurol.*, 12, 397.
 Stead, E. A., and Warren, J. V. (1944). *J. clin. Invest.*, 23, 283.
 Taylor, G. W., Kinmonth, J. B., Rollinson, E., Rotblat, J., and Francis, G. E. (1957). *Brit. med. J.*, 1, 133.
 Weiss, S., and Ellis, L. B. (1931). *J. clin. Invest.*, 9, 17.

TRICUSPID ATRESIA

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Among congenital heart diseases, tricuspid atresia is by comparison a rare complaint (White, 1951) and by itself is incompatible with life. Gibson and Clifton (1938) reported one case in a series of 105 congenital cardiac cases, Abbott (1936) had 16 out of 1,000 cases, and we had only one case out of 106 cases (Gwee, 1956, unpublished). However, compensatory mechanisms in the form of other coexisting congenital cardiac malformations sometimes enable a patient to live for many years. Taussig (1947) pointed out that the method of compensation may be either a communication of the two atrial chambers by an interatrial septal defect, the vestigial right ventricle receiving its blood supply through a ventricular septal defect, or persistent ductus arteriosus with interatrial defect and non-functioning right ventricle. White (1951) stated that tricuspid atresia is almost always attended by compensatory septal defects. Blackford and Hoppe (1931) regarded it as a functionally two-chambered heart.

Quite often the coexisting deformities produce no pathognomonic clinical signs, and in fact some cases may even be silent. Their presence is nevertheless suspected from the fact that the circulation must be able to complete its circuit to be compatible with life. The diagnosis is being made with increasing frequency in life, and in more and more cases knowledge of the coexisting defects is obtained pre-mortem by means of special investigations, of which angiocardiology and cardiac catheterization are probably the most informative.

Clinically, the patient is a cyanotic child with polycythaemia and gross clubbing of fingers and toes, and, cursorily inspected, may well pass for a case of Fallot's tetralogy (Alexander and White, 1947). A closer examination, however, may reveal in some cases a left ventricular apical impulse to suggest the true nature of the anomaly. Electrocardiography typically, though not necessarily, shows a left axis deviation and suggests left hypertrophy (Rihl, Terplan, and Weiss, 1929; Burchell, 1949). With the clinical appearance of Fallot's tetralogy, this finding is almost diagnostic of tricuspid atresia

(Astley, Oldham, and Parsons, 1953)—one of the few occasions when the electrocardiograph is of value in the elucidation of cardiac malformations. Radioscopy usually shows a large left ventricle, and the appearance may resemble that of truncus arteriosus, which, however, produced more cyanosis and is seldom seen beyond infancy. Also, the plateau-like appearance in truncus arteriosus is usually more marked, and the vascular pedicle remains the same size on rotation during radioscopy (Taussig, 1947). The diagnostic triad therefore consists of clubbing with cyanosis, left axis deviation in the electrocardiogram, and a characteristic appearance on radioscopy (Taussig, 1936). However, the shape may vary considerably (Astley *et al.*, 1953), and the pulmonary vascularization may be normal or markedly oligoemic. An exact diagnosis of all the coexisting anomalies may be impossible even after very exhaustive studies.

The salient differences of these cyanotic types of congenital heart disease are shown in the Table.

The life expectancy is usually short, although Abbott's (1936) series of 16 cases included a case of Hedinger's (1915) surviving up to 56 years. The review of Edwards and Burchell (1949) shows that the longevity of the patient depends to a large extent on the presence or absence of a transposition of great vessels. In their review, 45 cases were classified into four groups, thus: (1) No transposition of great vessels, (a) pulmonary

Signs and Symptoms

	Cyanosis	Clubbing	Polycythaemia	Stunting of Growth	E.C.G.	Pul. Vasc.	Death
Fallot's tetralogy	++	++	++	++	R.V.H.	Sparse	Often reach adult life
Eisenmenger's complex	Later	Later	++	May be little	R.V.H.	Normal	" "
Tricuspid atresia	++	++	++	++	L.V.H.	Sparse	Usually infancy and childhood
Truncus arteriosus	++++	++	++	++	L.V.H. or normal	"	" "

atresia, closed ventricular septum; (b) subpulmonary stenosis. (2) Transposition of great vessels, (a) pulmonary and subpulmonary stenosis; (b) no pulmonary or subpulmonary stenosis.

Of these, 28 cases belong to group 1b with subpulmonary stenosis and 4 to group 1a with pulmonary atresia. The life expectancy of groups 1a and 1b is stated to be quite short, the oldest being four years (Bellet and Stewart, 1933) and the causes of death being cardiac failure, bronchopneumonia, intracranial venous thrombosis, brain abscess, and bacterial endocarditis, in that order of frequency (Edwards and Burchell, 1949). The presence or absence of persistent ductus arteriosus and the adequacy of the pulmonary circulation do not seem to influence the length of survival.

Case Report

A male Chinese aged 20 was first seen on September 26, 1955, with a history that he had been breathless and blue as far back as he remembered, and that at about 5 years of age his fingers and toes began to get bulbous at the tips. Three weeks before admission he was short of breath and febrile. His legs and body became swollen.

On examination he was very cyanosed, with gross clubbing of fingers and toes. He was on the small side, height 4 ft. 8 in. (142 cm.), with no adult secondary sex