NEONATAL OEDEMA*

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In the first 4 weeks of life so-called 'physiological' oedema of the newborn occurs in 13% of premature infants and 1% of babies born at term[1]—in some series even more frequently. The term 'oedema neonatorum' has had different meanings for different observers, but is now usually reserved for 4 categories, viz. (*a*) sclerema, (*b*) subcutaneous fat necrosis, (*c*) neonatal cold injury, and (*d*) 'physiological' oedema. Besides these syndromes there are other oedemata unique to the neonate, as in the oedematous infant of the diabetic mother or the hydrops foetalis syndrome due to several causes, among them haemolytic disease and cytomegalic inclusion disease. With a few exceptions, such as kwashiorkor or lipoedema, any cause of oedema in the older child or adult can operate in the newborn, even as apparently unlikely a cause as acute or chronic nephritis.

The following case is presented because the aetiology of the oedema remains obscure.

CASE REPORT

A male African infant born normally on 28 June 1958 after a normal pregnancy. His mother was a healthy African primipara aged 24. She was delivered at home in Alexandra in the early evening under the care of her mother and grandmother. It was a vertex delivery. Immediately the baby was born he cried, but he was not properly inspected until next morning, because all lights in the house were extinguished on account of noises in the street attributed to 'tsotsis'. Next morning he was seen to have grossly swollen feet. He had not been exposed to cold. When he was 2 days old and had not improved the clinic doctor was called in. He found a lusty newborn infant whose legs and feet below the knees showed gross pitting oedema. He found no other abnormality, and asked me to see the baby when 8 days old.

On examination I found a mature, well-looking neonate with gross pitting oedema of legs and feet (Fig. 1). The femoral pulses were palpable, and the extremities felt normally warm. The child was vigorous, and the mucous membranes were well coloured. The Moro and sucking reflexes were present. There was no ascites. Liver and spleen were not palpable. No signs were found of cardiac enlargement, congestive failure, or abnormal heart sounds or rhythm. The air entry was good, and no adventitious respiratory sounds were heard. The genitals appeared normal; both testicles were in the scrotum. The urine proved normal on chemical and microscopic examination.

Pitting oedema of a localized area of the scalp was first noticed at 2 weeks old, and persisted (Fig. 2).

The baby always sucked well at the breast, and had daily stools. He appeared to thrive at the breast for 6 weeks, at which time the breast milk diminished and he received gradually increasing amounts of sweetened full-cream dried-milk mixtures. He was smiling socially and looked vigorous and well at 7 weeks of age, when he was 22 inches long and weighed 10 lb. 4 oz., with a head circumference of 15½ inches. His oedema of the lower legs and feet and the occipital region of the scalp has remained unchanged (to age 3 months—the time of writing). He has never been dyspnoeic nor shown any signs of distress.

Cardiac and renal causes for the oedema were excluded. X-ray of lungs and heart was normal, except for an enlarged thymus. There was no cardiac enlargement. Repeated examinations of the urine revealed no abnormality. Blood urea and electrolytes were normal. Serological tests for syphilis were negative, though penicillin therapy was given while awaiting results. X-ray of bones were normal. There has never been any hypoproteinaemia; paper electrophoresis shows a normal pattern. A normal blood-count without evidence of haemolysis, on several occasions, has ruled out erythroblastosis foetalis in this case.

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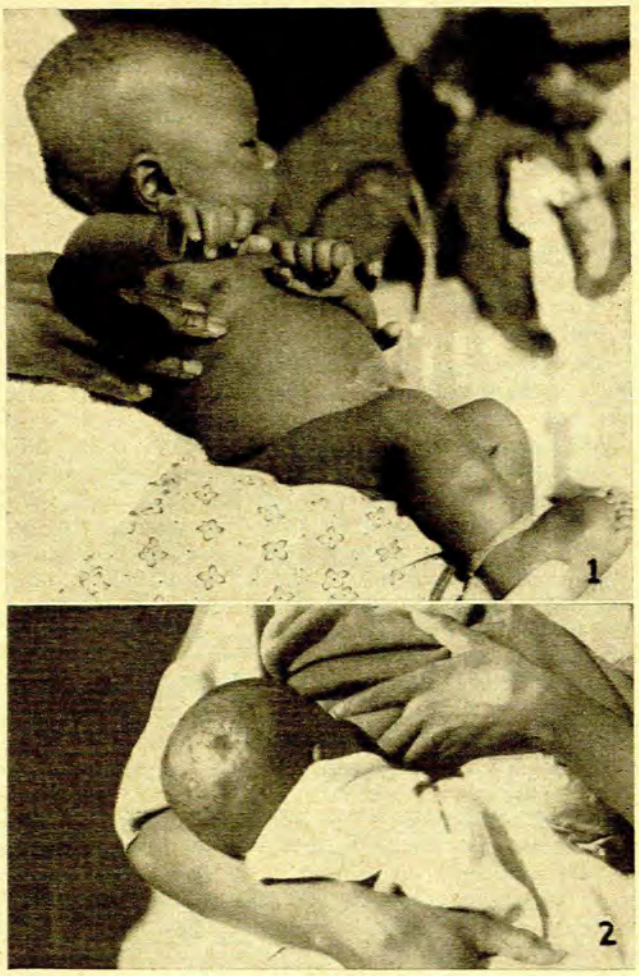

Fig. 1. Patient aged 1 month showing oedema of legs.
Fig. 2. Showing oedema of the scalp.

DIFFERENTIAL DIAGNOSIS

Physiological 'oedema noenatorum' is thought to be associated with physiological processes peculiar to the newborn. McCance[2] has been one of the foremost workers in this field, and he has shown how poorly the neonatal kidney performs in regard to urea and mineral clearances. The newborn human and newborn pig are very alike in this. Neither responds to extra salt in the diet with a corresponding rise in excretion. The extracellular fluid volume, which in any case is much larger in newborns relative to adults, expands with a rise in the concentration of sodium. This breakdown in homeostasis is well tolerated by the newborn, and in the piglet leads to a fall in the katabolism of protein.[3]

Failure to produce an adequately hypertonic urine is probably also concerned with decreased sensitivity of the tubules of the neonatal kidney to anti-diuretic hormone and decrease in production of this hormone.[4]

However, despite the apparently ineffective neonatal kidney in most newborns, the composition and volume of extracellular fluid *is* usually maintained at normal levels, and oedema does not result. This is because the food material absorbed is always being incorporated into the tissues in the process of growth and does not present itself for excretion by the kidney. The kidney of the newborn is therefore relieved of much of the work which it is called upon to do in adult life. Nevertheless, cow's milk presents to the baby's kidney a much greater osmolar load of electrolytes and nitrogen than breast milk,[5] which reminds one of the care necessary in choosing suitable feeds for the very young, and especially the premature.

Usually in 'physiological oedema' the baby is not ill and the oedema is transient, occurring at 2 or 3 days of age in the lower limbs, face and genitalia and clearing within a week.

The effects of cold in neonatal physiology has been studied in newborn pigs and also in puppies, which behave rather like newborn infants. They can tolerate a considerable fall of body temperature provided the return is made within a reasonable time.[3] However, long-continued exposure to cold gives rise to the syndrome of *neonatal cold injury*, so well described recently by Mann and Elliott.[6] The presenting features are increasing apathy, refusal of food, and oliguria, in an infant who does not look ill but is cold to the touch. He has pitting oedema, hypothermia, and striking erythema of the face, hands and feet. This illness is often complicated by purulent nasal discharge, and pulmonary haemorrhage has often been found after death. It is thought that Henoch's account of 'oedema of the newborn' in 1899 corresponds closely to this syndrome.

Local hardening of superficial tissues sometimes occurs over frankly oedematous parts, but this hardening, accompanied and preceded by oedema, differs from *sclerema*, where there is a diffuse hardening of subcutaneous tissue and the skin colour by contrast is yellowish-white. Underwood gave an accurate clinical description of this disease in 1811, but it has often subsequently been confused with the much milder *subcutaneous fat necrosis* of the newborn.

It is obvious that the patient described does not fit into any of the 'oedema neonatorum' syndromes. Tables I and II show causes of generalized oedema in the neonate, none of which are the probable reasons for the oedema in this patient. Although the distribution of oedema in this patient suggests a generalized form, in fact oedema has never been seen above the knee except in the region of the scalp, and there has never been sacral oedema or oedema of forehead and eyelids.

TABLE I. CAUSES OF GENERALIZED OEDEMA IN THE NEONATE

*'*Oedema of newborn*'	All have been known as oedema neonatorum
Neonatal cold injury	
Sclerema	
Subcutaneous fat necrosis	
Infant of the diabetic	
Erythroblastosis foetalis, hydrops foetalis syndrome—due to several causes	
†(*Oedema of tetany of newborn*)	
Oedema of post-acidotic state and other mineral depletion	
Congenital syphilis	
Renal oedema	
Cardiac oedema	
Hypoproteinaemia	
Angioneurotic oedema	
Hormonal oedema iatrogenic—e.g. cortisone.	
†Beri-beri	

TABLE II. CAUSES OF HYPOPROTEINAEMIA IN THE NEONATE

Prematurity
Congenital syphilis
Loss of protein *via*—
  (*a*) Kidneys in nephrosis
  (*b*) Haemorrhage
Protein dilution in excess fluid administration
Heart failure
Congenital hypogammaglobulinaemia
Hypercatabolic hypoproteinaemia—transient dysproteinaemia group
Hypoalbuminaemia with aminoaciduria.

TABLE III. CAUSE OF LOCAL OEDEMA IN THE NEONATE

**Birth trauma*
Hormonal change in female neonate's external genitals
Localized exposure to cold
Lymphoedema secondary to congenital bands
Congenital lymphoedema
Bonnevie-Ulrich-Turner syndrome
Infantile cortical hyperostosis
‡Venous thrombosis

* Italics indicate conditions confined to the neonatal period.
† Described in literature concerning neonates, but doubtful entities.
‡ Theoretically possible, but does not appear to have been described.

Table III shows causes of localized oedema in the neonate. Of these conditions the only likely diagnosis in this patient is *congenital lymphoedema*.

The original report of congenital lymphoedema was published in 1892 by Milroy in America,[7] but in 1891, Nonne,[8] in Germany, had also reported a condition of heredofamilial oedema of the lower limbs. Meige[9] reported a further series in France in 1899. Since then various authors have reported the condition. Rosenberg,[10] in 1940, divided the cases into 2 groups, those in whom the condition was congenital and those in whom it appeared in early adult life. The majority of Milroy's cases had shown oedema at birth, while Meige's cases had their onset at puberty.

In 1940 Herzberg[11] described a case of 'idiopathic lymphoedema' or 'Milroy's disease' of the upper extremity. It has also become apparent that not all cases of lymphatic oedema have a family history. Milroy, however, described his disease as characterized by (1) its congenital and hereditary nature, (2) the limitation of the oedema to one or both lower extremities, (3) the permanence of the oedema, and (4) the complete absence of constitutional symptoms.

McKendry, Lindsay and Gerstein[12] report that from 1938 to 1955 at the Hospital for Sick Children, University of Toronto, Canada, 8 patients with congenital lymphoedema were admitted. They report a case with chylous ascites and lymphoedema in which, at autopsy, the subcutaneous tissues of the involved limbs showed many dilated lymphatic channels. The pathological picture was one of multiple dilated lymphatic channels with failure of peripheral channels to communicate with the large lymphatics. This patient, therefore, may be an example of one of the rare causes of neonatal oedema—a congenital defect of the lymphatics.

SUMMARY

A case of neonatal oedema in which the oedema has persisted beyond the first month, is presented.

The differential diagnosis is discussed.

Congenital lymphoedema is suggested as the most probable diagnosis.

ADDENDUM

The patient has recently been seen in the out-patients department of Baragwanath Hospital, Johannesburg, aged 16 months. He is apparently healthy, with no constitutional symptoms. The oedema is still present, more brawny in nature, with less pitting, but still limited to the same sites. This is in keeping with the suggested diagnosis.

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