III.

Partial Macrosomia.

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(Report on Microscopic appearances by T. LAW WEBB, M.D. (Birm.),
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THE following case seems worthy of publication on account of its rarity and the obscurity which still surrounds the pathogenesis of the disease.

C.E., a schoolboy, aged seven years, was admitted to the Salop Infirmary, Shrewsbury, on the 21st of December, 1905, suffering from a congenital tumour of the right forearm and hand, which had recently become greatly inflamed as the result of an injury.

The mother stated that when the child was born the affected arm and hand were in much the same condition as regards relative size and general appearance as they were on admission, but that at birth there was no inflammation present in the limb.

About five days before admission, whilst playing, he knocked his arm against an apple tree. He did not complain at the time, but in the course of a day or two the forearm became very much inflamed and swollen, and he complained greatly of pain in it. He was seen by a medical man who ordered the application of poultices. As these did not relieve the pain or reduce the inflammation, he was brought to the Infirmary and was admitted under the care of Mr. L. J. Godson, through whose kindness I am enabled to publish this report of the case.

The patient was a pale, rather poorly-nourished, delicate-looking boy, with an anxious, fretful expression. The cranium was considerably larger than normal, indeed somewhat hydrocephalic in appearance. The root of the nose was broad. The two upper central incisors were loose and deficient in enamel, but they did not show pegging or notching; they were probably, however, temporary teeth not yet shed. The temperature on admission was 101°F.

The accompanying photograph shows fairly well the general appearance and relative size of the affected limb. Excepting the thumb and the tips of the fingers, which were normal, the right

hand and forearm were uniformly and generally swollen. The dorsum of the hand was soft and fluctuating, and might aptly be likened to a miniature water-bed. The forearm was inflamed and brawny; an indefinite sense of fluctuation was obtained in places, especially over the posterior aspect of the limb. Considerable pain was complained of, and some parts of the forearm were tender to the touch. There was a hard, circumscribed swelling about the size of a marble, just anterior to the insertion of the deltoid. In the axilla a few enlarged glands were felt.

The mother stated that the boy was a full-term child; the labour was rather tedious (as one would expect from the condition of the limb) but not instrumental. The usual "maternal impression" was absent in this case, but there was a history on the part of the mother of having had several frights during the pregnancy, and also of having fallen on a bank about the second month and bruised herself severely. There was no history of any attack of lymphangitis or erysipelas during the pregnancy. The parents had never been abroad.

At birth, as already stated, the forearm and hand were in much the same condition as on admission, except that there was no inflammation present then. As the boy grew older he was able to use the limb but only to a limited extent, raising it up and picking up light articles; but he never had much power in it, and could not write with it or use it to feed himself. The swelling never diminished in size at any time.

He was the ninth child of a family of eleven. All the other children were healthy, and no family history of a similar condition could be obtained. There was no history of miscarriages or any evidence of specific disease in the parents.

As there were signs of suppuration, on 22nd December, under chloroform, I made two small incisions into the inflamed tissues, but found no pus, only blood and serous fluid escaping. Boracic acid fomentations were applied. The inflammation subsided rapidly and the incisions quickly healed up. Four days after admission the temperature was normal.

The condition was diagnosed as a congenital tumour, most probably of the lymphangiomatous type. The limb was examined by the Röntgen rays, but the bones were found to be normal in appearance.

The child was put on full diet and was given Kepler's extract of malt and cod liver oil, and also port wine. In spite of this his general condition did not improve and he was fretful. The temperature occasionally rose to 101°F. A consultation was held, and it was decided that as the limb was practically useless, and the condition

was materially affecting the health of the child, the best treatment was amputation. Accordingly, on 9th January, 1906, the patient being under chloroform, Mr. Godson amputated the limb at the junction of the middle and lower thirds of the upper arm. The wound healed by first intention. His general health rapidly improved, and he was discharged in good health on 27th January. I saw him again on 18th April. He was looking strong and healthy. The small, hard nodule, at the insertion of the deltoid could still be felt, but otherwise the stump was good, with a firmly healed cicatrix.

Dr. T. Law Webb reported on the microscopic appearances as follows:—"Vertical sections show the skin itself to be fairly normal. Much of the connective tissue beneath, however, is of embryonic type with many nuclei. There is much fatty tissue in this region. The thick layer of tissue lying beneath the subcutaneous fat and above the deep fascia is peculiar, and this constitutes the chief bulk of the enlargement of the limb. It consists of adenoid tissue, having a fine network of branching fibres, densely packed with what appear to be lymph-cells. In places the fibres are coarse and easily seen; in other directions the rounded lymph-cells are so crowded together that the fibres are not visible. There are large tracts in which lymph-cells and connective-tissue fibres are placed in about equal proportions. Here and there wide, fatty areas, separate the masses of adenoid growth. The vessels are numerous, and their middle and outer coats are greatly thickened. In places the growth is so vascular that it might be taken for part of an angioma. Necrotic areas of considerable extent are present, where the tissue either stains faintly and diffusely or not at all. In some of these unstained regions the cell-elements have disappeared, leaving only an exceedingly fine, fibrillar network, having a very unusual appearance. The lymph-vessels and spaces are not enlarged. No portion of the growth shows any appearance of lymphangiona. Imperfectly developed muscle fibres are seen in the deeper parts of the sections, but no situation is visible in them."

A congenital tumour such as that described above may belong either (1) to the partial macrosomia of the embryonic or foetal period of antenatal life, or (2) to the congenital elephantiasis group, which arise in the foetal period. It is often a very difficult matter to decide to which group any given case belongs. I have placed the above case in the group of partial macrosomia for reasons to be stated later.

To begin with it may be advisable to state briefly the leading characteristics of each of these groups as they are described in Dr.

J. W. Ballantyne's standard work on "Antenatal Pathology" (to which and to the kind assistance of Dr. Ballantyne himself I am deeply indebted.)

The partial macrosomias belong to the group of localized monstrosities, which may arise either in the embryonic or foetal period of antenatal existence. They are characterized by a local overgrowth of the body, one part such as an arm, a leg, a toe, or one half of the face, considerably exceeding the other parts in size. As a rule, all the constituent structures of the affected part are involved in the overgrowth. Here we are specially concerned with the variety in which one extremity alone is affected. The whole of one limb or a part of one limb is hypertrophied at birth. The upper extremity is more often affected than the lower. It has been noted in several cases that while the whole area was larger than normal, only two or three of the fingers showed marked hypertrophy. In the case recorded above, the thumb and tips of the fingers were normal. In the upper limb the commonest form of localized enlargement is that in which only one or two of the fingers are involved (megalodactyly or macrodactyly). Occasionally the lower limb or the foot alone is affected.

The pathogenesis of the condition is obscure. When it arises in the foetal period it may sometimes be explained by lymphatic obstruction or lymphangiectasis, or by a diffuse fatty infiltration of the subcutaneous and muscular layers of the part. The latter appears to be the only feasible explanation of the condition as found in the above case. When all the structures are involved the condition most probably arises in the embryonic period, and two theories have been advanced to explain its pathogenesis under these circumstances. The first is the vascular theory of Trélat, Monod and others, which ascribes to slowing of the capillary circulation an increased nutritive activity of the tissues; the other is the nervous theory—a trophic action of the special nerves of the part.

Congenital elephantiasis, the allied condition, is characterized by a hypertrophic or hyperplastic state of the subcutaneous tissues present at birth. The name covers a variety of conditions, *e.g.*, (1) the soft cystic variety of the disease, (2) the hard and soft varieties which may be widely distributed or strictly localized, (3) multiple cutaneous neuro-fibromata and fibroma molluscum are also sometimes included in this group. The condition has to be diagnosed from congenital unilateral hypertrophy or partial giant growth (partial macrosomia) on the one hand and a heterogeneous group of neoplasms (including nerve naevi) on the other hand.

In the common type of congenital elephantiasis the hyperplasia is confined to the subcutaneous tissues with special involvement of the lymphatics. Occasionally, the vessels, nerves, and even the bones, and muscles, and fibrous tissue of the neighbouring parts may be involved. In the above recorded case, as will be noted from Dr. Webb's report, the changes were confined to the subcutaneous tissue, *but the lymphatics were not involved.*

Occasionally, a hereditary history is obtained. This was absent in the present case and beyond the injury and frights mentioned the pregnancy had nothing unusual about it.

The lower limbs are most frequently the seat of the disease. When this is the case there is usually some difficulty in walking; but otherwise the condition may not interfere materially with the life of the child. A tendency to diminution in size is sometimes noticed. The skin is usually normal. Grooves on it have been described which are probably due to amniotic bands. These were absent in the case described.

Amniotic bands encircling the limb have been advanced as an explanation of the cause of the condition, and this view may occasionally be correct. A true elephantiasis arabum is out of the question in most cases, as no traces of filaria are found. A streptococcal infection of the infant by the mother has been regarded as an explanation, but a history of maternal erysipelas or lymphangitis during the pregnancy is rare. A hereditary history also is rarely obtained. Others think that the condition may be due to a toxin circulating in the blood and causing long continued irritation of the subcutaneous tissues, but this is at least not proved.

The above case presents some of the characteristics of each of these groups, but on the whole, I think the evidence—especially that of the microscopic appearances—favours the diagnosis of partial macrosomia arising in the foetal period of antenatal life. The absence of any special involvement of the lymphatics is very much against the case being one of congenital elephantiasis, and there is nothing in the personal or family history pointing that way. On the other hand, the microscopic appearances, the distribution of the disease—the thumb and finger tips escaping,—the whole clinical picture, and the hypertrophy of the head which was noted, all point to the case being one of partial macrosomia (or congenital unilateral hypertrophy.)

Three chief methods of treatment have been recommended. First, elastic compression of the hypertrophied parts maintained for some

time. At the consultation this line of treatment was suggested, but at that time the inflammation and tenderness in the limb were too great to warrant its adoption. Second, ligature of the chief artery supplying the enlarged area has been tried. Third, amputation of the affected limb. This, however, is said sometimes to be followed by the excessive growth of neighbouring structures. In the above case there was no signs of this three months after amputation, and the child's health had greatly improved. As regards treatment each case must be judged on its own merits.

Tube.

Tube.

Ovary.

Fibro-
myomata
enucleated.

Dr. Lloyd Roberts's specimen.—Fibro-myomatous Uterus removed by supra-vaginal Hysterectomy after preliminary enucleation of several of the tumours.