

HEMI-HYPERTROPHY OF THE BODY WITH NÆVUS AND VARICOSE VEINS

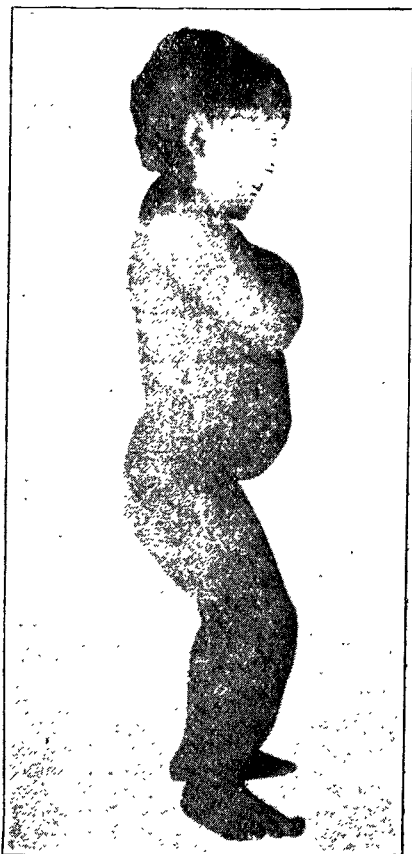
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A GOOD example of this remarkable and rare disease was recently admitted to the Manchester Residential School for Crippled Children.

The patient is a girl 7 years of age, who was admitted on account of genu valgum due to rickets. There is situated over the lower part of the trunk on the right side an extensive nævus. (Fig. 1.) The nævus is of the ordinary port-wine stain type, quite superficial, not raised above the surface, and showing a very sinuous outline with two or three out-lying splashes of similar nœvoid skin. It is entirely confined to the right side and does not cross the middle

FIG. 1.



Side view of patient.

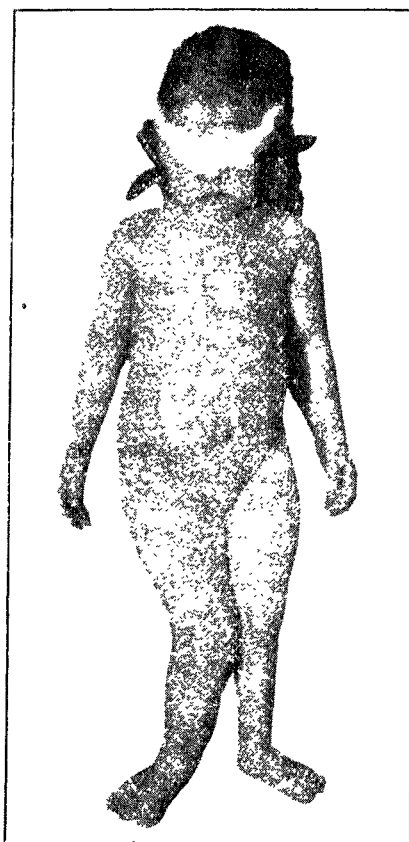
line at any point. The nævus is congenital and, excepting for an increase commensurate with the growth of the child, does not appear to have extended since birth.

The girl shows asymmetry of the body in a marked degree, due to overgrowth of the side on which the nævus is situated. There is slight but distinct asymmetry of the two halves of the skull and face, and the palpebral fissure is noticeably wider on the right side. The right upper extremity is distinctly larger than the left; the difference is especially marked in the hand. There is hardly any inequality in the two halves of the thorax, but the right side of the pelvis is larger than the left. Hypertrophy of the right side is, as the illustration shows, most marked in the right lower extremity, which from the iliac crest down to the toes is much larger than its fellow. (Fig. 2.) Measurement of the two limbs gives the following figures:—Total length: right, 20 in.; left, 19 in. Circumference of thigh: right, 16 in.; left, 14½ in. Circumference at level of knee: right, 10¾ in.; left, 10 in. Circumference of calf: right, 9½ in.; left, 7¼ in. The patella of the right side is distinctly larger than that on the left, and the disparity of the feet is well shown in Fig. 3.

Radiographic examination of the lower extremities shows that the asymmetry involves the bones equally with the soft parts, but there is no difference in structure or density of

the bones, and the changes due to rickets are equally marked in each limb, although it is interesting to note that to the eye the rachitic changes are more pronounced on the right side than on the left. The skin of the affected limb is normal, excepting for the varicosities to be mentioned later.

FIG. 2.



Front view of patient

No increase in the secretion of sweat or sebaceous matter has been noticed; there is no abnormal growth of hair, and the nails are normal. The increase in the circumference of the limb appears to rest mainly with the subcutaneous tissue, which is increased in large pads and rolls. This overgrowth is especially marked in the calf, and the subcutaneous tissue is here compressible to an extent which suggests a con-

FIG. 3.



Showing disparity of feet

siderable nœvoid element in its structure. The muscular power of the right limb is good, but is in no wise better than its fellow.

There are scattered over the abdomen and the front of the thigh numerous small patches which consist of very delicate convoluted venules, and in a few instances present a bright red arterial appearance. The majority of these patches have undergone no change for some months, but others are evanescent and show a tendency to the formation of small subcutaneous hæmorrhages, which are subsequently absorbed with complete disappearance of the varicose patch.

The association of localised hypertrophy with nævus and varices, though rare, appears to form a definite clinical entity. The disease, when fully developed, presents three features, of which the nævus is congenital, the hypertrophy also congenital but more apparent as growth develops, whilst the varices appear later, and in some instances of the disease are small or entirely lacking. The interesting association of the three conditions, no less than the striking deformity that they produce, has given rise to a fairly considerable literature on the subject, and this literature is almost entirely of French origin. The first mention of the disease which I have been able to find is by J. Adams, who reported a case in THE LANCET of August 7th, 1858. This case, which is reported without comment as a curiosity encountered in the course of casualty work, was evidently a typical specimen of the disease. The first important reference is the research of Trelat and Monod on Asymmetry, which appeared in 1869. This was followed by papers by Leblanc (1897), Duplay (1898), and by a very complete article by Klippel and Trenaunay in 1900. Grimaud took the disease as the subject of a Paris thesis in 1901.

A study of this literature shows that the association of nævus, local hypertrophy, and varicose veins is a definite one which has been encountered in all parts of the body. It has, however, been met with most frequently in the lower extremities, and very curiously the reported cases refer, almost without exception, to the right side.

The main features of the disease are as follows. The nævus is usually superficial but very extensive, with an irregular, sinuous contour. However large the area of the nævus may be it is always sharply limited by the middle line, and several cases are on record in which the nævus was arrested at the scrotal raphe. The localised overgrowth of the part is usually most apparent in the bones, and has affected the bones of the skull, face, upper and lower extremities. The bones are increased in all dimensions, and the disparity of the two sides has amounted to difference of as much as four inches in the length of the lower limbs. The coexistence of dilated superficial veins is remarkable, since varicose veins are very rare in childhood. The varices vary much in number and extent in different cases, and are the least constant of the three chief features of the disease. They are usually superficial and small, seldom attaining the proportions seen in the varices of adults.

Whilst the hypertrophy, nævus, and varices constitute the three classical signs of the condition, there are often found other phenomena which are of interest as indicating the far-reaching nature of the congenital fault. The skin of the hypertrophied parts is in some cases thick, rough, and dry, the condition amounting at times to an ichthyosis. The hair may be abundant and coarse in texture. The nails of the affected limb have been described as deformed, showing furrows along their length. Excessive secretion of sweat and of sebaceous material has been a striking feature of some cases. The subcutaneous tissue and the muscle have usually shared in the overgrowth of the affected part.

The origin of the disease is unknown. Many theories, ranging from maternal impression to vaso-motor paralysis, have been offered in explanation, but a review of them merely confirms the opinion that the origin of this disease is one of the many mysteries in the field of ante-natal pathology.

Manchester.

A CASE OF MENINGITIS

IN WHICH THE ONLY ORGANISM THAT COULD BE CULTIVATED FROM THE CEREBRO-SPINAL FLUID DURING LIFE WAS A BACILLUS OF THE PROTEUS GROUP.

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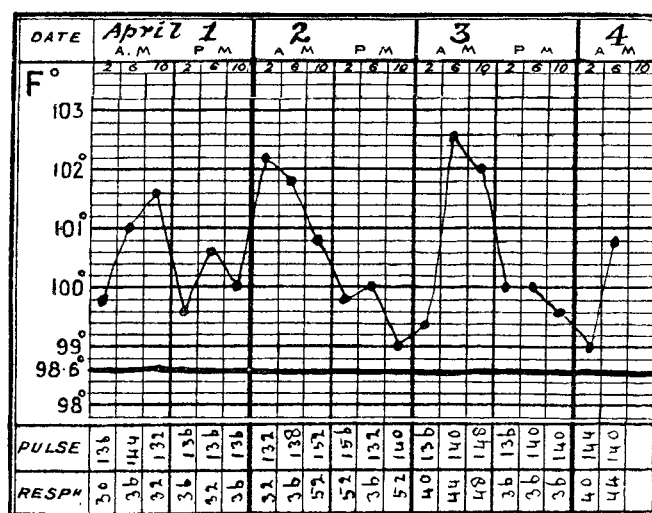
THE following is a brief abstract of the notes in this case, to which are appended the post-mortem and clinical pathological reports.

The patient, a male child, aged 9 months, was admitted to the East London Hospital for Children on March 31st, suffering from stiff neck of two days' duration. The mother noticed it when she bathed the child. He could not keep his food down—vomited it at once. He emitted a peculiar squeal during most of the night. The child had not been well since he began teething three months before. The

bowels were constipated. The mother noticed that he used "to work his eyes a lot at night." She thought this was due to fits. There was nothing of importance in his previous history. With regard to the family history, four children were alive out of seven. Two died from bronchitis and croup and one from epilepsy. The father and mother were healthy.

On admission the child was well nourished, pale, and very drowsy; the head was retracted. There was marked rigidity of the neck. Kernig's sign suspicious on both sides. Abdominal reflex increased on the left side, less marked on the right. Knee-jerks were present. Fontanelle was not bulged. Discs were normal. Lumbar puncture, *nil*; three attempts, no fluid. On April 3rd rigidity was more marked. Head was very retracted and abdomen sunken and soft.

FIG. 1.



Temperature Chart.

Abdominal reflexes slight. On the 4th he was very rigid. 10 c.c. of spinal fluid were drawn off on April 3rd. Smear showed bacilli, some of which were intracellular, but most extracellular. Fontanelle bulged this morning; much worse. The patient died.

Abstract of the Post-mortem Report.

Brain: The entire surfaces at the vertex and base were covered with a thick layer of greenish-yellow pus, and the ventricles were also dilated and contained pus. The cerebral tissue was sectioned in all directions without finding any evidence of a localised abscess. Spinal cord: This thick greenish-yellow pus stripped away with the membranes and formed a layer nearly a quarter of an inch thick. Lungs: Emphysematous. Liver: Cloudy swelling and fatty change. Intestines: Both the large and small intestine showed patches of enteritis with injection and swelling of the mucous membrane.¹ Middle ears: The middle ears on both sides contained pus. The right more than the left. The cerebro-spinal fluid: This was withdrawn under aseptic precautions by Dr. A. G. Hamilton in a sterile tube and carried to the laboratory, where an examination was made immediately. The fluid was obtained some time before death on April 3rd; it was very turbid, and contained large numbers of bacilli. Most of the organisms were extracellular, but a fair proportion were seen to be inside the cells in stained film preparations. A differential count gave 93 per cent. of polymorphonuclear cells. Bacteriology: Pure cultures of the organism were obtained on agar. The bacilli varied in length and short forms were present. Gram-negative. Agar plate: Greyish white colonies, discrete, with a surrounding foggy haze which gradually spread over the plate. Neutral red agar plate: No growth. Litmus mannite: Acid; no gas. Litmus maltose and litmus lactose: Diffuse turbidity. No acid or gas. McConkey: No change. Litmus milk: Slight acidity. No clot in seven days. Gelatin slope: Rapid liquefaction of medium (in 18 hours). Broth: Diffuse turbidity. In maltose and lactose growth was fair, but film preparations from the McConkey tube showed that the organism had died out. The growth on gelatin was examined by Gram's method, and Gram-negative bacilli were found present in pure culture. Since a slight greenish tinge was observed in the exudate post mortem the possible presence of *B. pyocyaneus* had to be considered, although neither the tint of the colonies on agar nor the odour of the broth cultures was in the least suggestive. A peptone water culture incubated for a week gave a totally negative result.

Instances of meningitis in which bacilli, other than *B. tuberculosis*, figure in the cerebro-spinal fluid during life are distinctly uncommon. The recognised infective agents are as follows.

1. *The typho-colon group*.—The presence of a Gram-negative bacillus rapidly liquefying gelatin at once excludes this family with its many bacteriological complexities, including *B. paratyphosus* A and B with the other members of the *Salmonella* group. The acid and gas-forming powers of *B. proteus* in the litmus sugar media are very variable,

¹ These lesions may be of some importance in view of the bacteriological findings.