CONGENITAL ABSENCE OF HAIR AND MAMMARY GLANDS

WITH

ATROPHIC CONDITION OF THE SKIN AND ITS APPENDAGES

IN

A BOY WHOSE MOTHER HAD BEEN ALMOST WHOLLY BALD FROM ALOPECIA AREATA FROM THE AGE OF SIX.

BY

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The subject of this case, a boy æt. 3½, presented a very peculiar withered or old-mannish look, all his features being thin and pinched. His fingers were shrivelled, and dusky, and their nails, which also were remarkably thin, were curved backwards so as to present more or less of hollow in the middle. His head was large and the anterior fontanelle not quite closed; the scalp was exceedingly thin, and with the exception of a quantity of down, was quite bald. It looked semi-transparent and tight, and the veins coursing in it were everywhere conspicuous. The veins were probably larger than natural. A large trunk came down the forehead on each side of the eyebrow and communicated by a transverse branch at the
root of the nose. The inosculations across the middle line of the scalp were many. There was a peculiar blue tinge about the lips; it involved the skin and not the prolabium only. At first I thought that this was due to accidental staining; but after he had been half an hour in my room it much diminished, as did also the turgescence of the veins of his scalp. His lips were exceedingly thin. His teeth were all cut and were tolerably regular, but his incisors did not stand quite straight, most of them had some slight inclination into the mouth. On his shoulders he was so thin that his coracoids and the outlines of his acromion processes could be easily seen; the skin over them being not much thicker than brown paper. The tightness of skin was nowhere very conspicuous excepting on the scalp; thus, on the abdomen, arms, and thighs the integument was quite loose but everywhere very thin. His muscular development was slight in all parts excepting the thighs, which felt hard and had muscles quite out of proportion to the rest of his body (this remark does not apply to the buttocks). His genitals presented a very remarkable contrast to the rest of his body. The parts about the pubes and upper part of the scrotum were so full and plump that a suggestion occurred that he must have double hernia. This, however, was not borne out by examination, and I believe the simple fact was that the scrotum and adjacent parts of skin were in the state of those of a normally stout child, whilst everywhere else the skin, subcutaneous cellular tissue, and panniculus adiposus were almost absent. The true scrotum was small, naturally corrugated, and occupied only the lowest part of the genital pouch which I have described. I do not think that there was anything very unusual in this state in a child, but must admit that possibly there was some excess of subcutaneous development about the pubes and root of penis. His testes were well placed and of normal size. His penis, except that there was phimosis, was quite natural. His toes and their nails were in the same condition as his fingers. He did not walk quite perfectly,
always keeping his knees a little bent, but I could not make out any definite muscular defect. One other remarkable feature remains to be mentioned, he had no nipples and their sites were occupied by little patches of scar. These scars were exceedingly superficial and slightly marked, but I am sure that they were there. Nothing like a mammary gland could be traced.

The history which the mother gave me of the child was that he had had no ailments since his birth, was of cheerful disposition, and very intelligent. It had been necessary from cross presentation to turn during delivery, and for some days after birth he had been very blue, probably in a state of partial cyanosis. He was still liable to vary very much in blueness in connection with the temperature and states of excitement, but never now presented anything approaching a cyanotic condition.

I have now to relate the very extraordinary fact which is possibly explanatory of the singular condition of things just described. It will have been noticed that the chief defects present in the child were, an atrophic condition of the appendages of the skin and its accessory cellular tissue and fat, which became especially conspicuous in the absence of the scalp hair. With this we had a well-developed condition of the male sexual organs and an absence of the mammary glands and nipples. Now the mother of this child from the age of six to the present time had worn a wig on account of alopecia areata. At the age mentioned she began to lose her hair, which had previously been plentiful, in patches. She described the usual course of things, how the patches increased, and the whole scalp became smooth and bald, and how subsequently the eyebrows and eyelashes fell. After a considerable time her eyebrows and eyelashes grew again, and a few tufts of hair appeared on the scalp. But she had never regained her scalp hair sufficiently to dispense with her wig, and her eyebrows were still so poor that she was obliged to colour them. Excepting this alopecia she had no signs of deranged nutrition, being a florid, comely, well-developed
woman. The little boy was her first and only male child, but he had five sisters, all older than himself and all of whom had excellent development of hair.

Very curious speculations suggest themselves in connection with Darwin's theory of pangenesis. Under this hypothesis it may perhaps be possible that the germinal elements of the child's cutaneous system, and especially for his scalp, were derived from his mother, and were, in connection with her long baldness, very defective in vigour. With this would fit the entire absence of the mammary glands and their nipples; with this also would fit the normal development of the male genital organs and their skin, since he would be supposed to take these from his male parent. The fact that all his sisters had good development of scalp hair may be supposed to be explained by the suggestion that they inherited chiefly from their father.

It is to be added that the marriage was not one of consanguinity, and that no baldness or defects of development had been known in the family previously.

I may have perhaps a little over-stated the general absence of subcutaneous fat. Excepting on the head and hands, it was nowhere quite absent; and this remark especially applies to the abdomen and back. The deeply placed fat was less affected than the superficial. Thus, lumps of it could be detected at the root of the neck. The skin was everywhere destitute of natural elasticity and plump firmness, and where not dusky had an earthy pallor. The eyelashes were present but very weak. The eyebrows almost entirely absent.

I was indebted to Dr. Jago, of Mulgrave Place, Plymouth, for the opportunity of seeing this child and for some facts as to its history.

Remarks.—I prefer, for the present at least, to leave the above remarkable case without attempting to contrast it with other examples of congenital alopecia on record. From all these it differs, so far as I am aware, in the fact that the female sex organs (the mammæ) were absent, whilst the skin of the male sex organs was the only part
of the integument in a normal condition. These peculiarities become of the greatest possible interest when we remember that he appeared to inherit his defect from his mother. I am well aware that the explanation hinted at is a mere conjecture, and that there are a multitude of facts which might seem to militate against it. We cannot afford, however, in investigating the very difficult subject of hereditary transmission, to neglect any hint which the facts of pathology may offer. I need scarcely say anything as to the well-known law that defects, the result of disease or injury occurring in the parent and not congenital, are not transmitted to offspring. Everyone knows that circumcised fathers beget children in whom the prepuce shows no modification. To this law the case I have recorded seems to offer an exception, for there was not the slightest doubt that the mother's loss of hair was caused by the common form of alopecia areata, and did not begin till she was six years old. In fact, her hair grew again several times after its first falling, and again came off. Some will probably be inclined to consider that the mother's condition and that of her only son were associated as a mere coincidence and that the one was in no way dependent on the other. It is indeed precisely because this connection seems so probable, whilst it is in flat contradiction to received opinions, that I have thought the case worthy the attention of the Society.

(For report of the discussion on this paper, see 'Proceedings of the Royal Medical and Chirurgical Society,' New Series, vol. ii, p. 116.)