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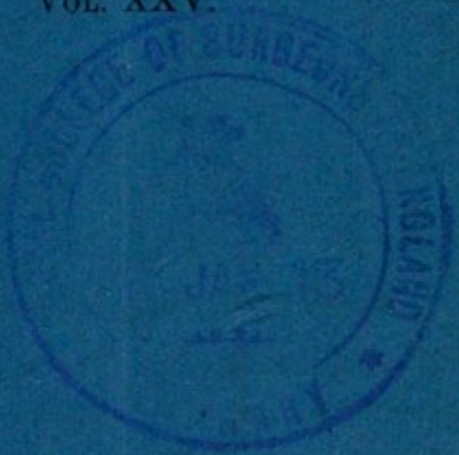

MAMMARY VARIATIONS *PER DEFECTUM.*

By W. Roger Williams, F.R.C.S., *Surgeon to the Western General Dispensary, late Surgical Registrar, Middlesex Hospital.*

Section I.

The development of the breast may fail at any stage of its evolution—from early embryonic life up to the climacteric period. When the morbid process sets in before the 2nd or 3rd month of intra-uterine life, there results complete suppression of the organ—*amazia.*

In animals having normally a large number of mammæ, some of them are often aborted in this way; but in human beings

Complete absence of both mammæ (Wylie).

and other bimastic animals, amazia is a very rare affection—much rarer, for instance, than polymazia.

Nearly all the cases hitherto recorded, in which the sex has been well marked, have been in females.

The deformity is frequently, but by no means invariably, associated with grave malformations *per defectum* of the adjacent chest or of the sexual organs. One or both breasts may be affected.

Complete absence of *both* mammæ is one of the very rarest congenital deformities.

Most cases have been met with in acephalous monsters, associated with deficient development of the thorax.

I know of only four instances unaccompanied by such conditions.

In the first case[1] (*see* fig.) the patient was a single woman, aged 21, who, when examined three months after giving birth to a healthy male child, was found to present no trace whatever of mammæ, areolæ, or nipples. Not a drop of milk had been secreted, so that she was unable to suckle. Menstruation set in at 15, and she had since been regular. But for the mammary deformity she was well made, and her health had been good. In answer to a letter of inquiry, Dr Wylie, her medical attendant, kindly informed me that there was no deficiency of the pectoral muscles or ribs, and that the external genitals, the teeth, hair, and other dermal appendages, were well developed.

The second case[2] occurred in the person of a woman, who one week previously had been prematurely delivered of a living child. She presented no trace of mammæ or nipples, but in the position of each of the latter was a pigmented patch of skin, the size of a sixpence.

The subject of the third[3] case was a boy 3½ years old, in whom complete absence of both mammæ from birth was associated with similar absence of hair, and an atrophic condition of the whole integument and its appendages, except that of the external genitals. The latter were well developed, except for phimosis; and presented a remarkably plump appearance, as compared with the shrivelled aspect of the rest of the body. The testes were well placed and normal. The boy's mother had suffered from *alopecia areata* from the age of 16.

Mr Hutchinson indulges in some interesting speculations as to the explanation of this remarkable case, on the basis of modern theories of heredity.

In the fourth case[4] the patient was a so-called hermaphrodite, aged 65, who had always passed for a female. On examination of the body after death, its general appearance was that of a male, and there was a tremendous beard. Both mammæ were completely absent. Menstruation had never occurred, nor was there any history of sexual desire. Further examination was

[1] Wylie, W., *Brit. Med. Jour.*, 1888, vol. ii. p. 235.

[2] Batchelor, H. T., *Brit. Med. Jour.*, 1888, vol. ii. p. 876.

[3] Hutchinson, J., *Med. Chir. Trans.*, vol. lxix. (1886), p. 473.

[4] Pilcher, *Lancet*, vol. i. (1838), p. 915.

limited to the genital organs. The clitoris was very large—quite as large as in many competent males. The corpora cavernosa were large; there was a bulb to the urethra, a rudimentary prostate, and rudimentary ejaculatores seminis. In addition there was a rudimentary uterus and vagina—the latter ending in the urethra. Nothing is said about ovaries or testes.

Complete absence of *one* breast is only a little less rare than that of both.

Several authors have stated (contrary to what happens in polymazia) that the right breast is the one more frequently affected. I am unable to support this statement; for, as will be seen below, in most of the cases collected by me the deformity was on the left side.

It is alleged sometimes to occur independently of any other deformity. Birkett[1] cites a case (Marandel's) of this kind; but, on looking up his reference,[2] I found the record so meagre and unsatisfactory that I was unable to determine whether there was associated deficiency of the pectoral muscles or not.

In the following cases unilateral amazia was correlated with malformation of the adjacent chest wall:—

1. In a woman,[3] aged 30, who died of peritonitis eight days after her confinement, there was found in place of the R. breast a shallow depression; but no trace of the nipple, areola, or gland. Beneath the skin there was nothing but a thin layer of adipose tissue. The L. breast was well developed and full of milk. The anterior parts of the 3rd and 4th ribs were absent, together with the corresponding intercostal muscles, the sternal part of the pectoralis major muscle, the whole of the pectoralis minor, and portions of the serratus magnus. The gap was closed in by tough aponeurosis. The pleura and lung were normal.

2. Here the patient was a healthy-looking girl,[4] 5 years old, whose parents also were healthy and free from any deformity. Of the R. mamma, nipple, and areola there was no trace; the skin over this region was like that of the rest of the body. The pectoralis major and minor muscles were deficient, as well as the anterior part of the 4th rib and the adjacent

[1] *Diseases of the Breast*, 1850, p. 23.

[2] *Dict. des Sci. Méd.*, t. xxx. p. 378.

[3] Froriep, *Neue Notizen*, Bd. x., 1839, s. 9.

[4] Reid, *Froriep's Neue Notizen*, No. 500, Bd. xxiii., 1842, s. 254.

intercostal muscles. At this spot, during respiratory movements, hernial protrusion of the pleura took place, the overlying skin being in close contact with the latter. The six upper ribs, except the first, were markedly bent and arched forwards on both sides, causing considerable deformity of the pelvis. The L. mamma was normal.

3.[1] In this case complete absence of the L. breast was associated with absence of the L. upper limb, which was represented only by a small conical stump at the shoulder. There was also large deficiency of the thoracic wall on this side, through which the thoracic and abdominal viscera protruded, covered only by a membranous envelope.

4.[2] A girl, aged 10, with complete absence of the L. mamma, areola, and nipple. The sternal part of the pectoralis major also wanting. The other breast normal. No heredity. The mother attributed the disease to fright during pregnancy, from having seen a woman's chest after amputation of the breast.

5.[3] In a healthy married woman, aged 22, shortly after her first confinement, complete absence of the L. breast was noticed by the medical attendant. The nipple was represented by a small pimple. The pectoral muscles of the affected side were imperfectly developed. The woman's mother first noticed the deformity three weeks after the patient's birth. She attributed it to having been frightened when pregnant by a woman who called at her house and exposed her chest, showing marks from amputation of her breast for cancer.

6.[4] A single woman, aged 21, with complete L. amazia. The pectoralis major imperfect. The patient otherwise well formed. No heredity.

Similar cases have been recorded by Lousier[5] and Schlözer,[6] but I have been unable to get access to the original memoirs.

Referring to the former of these cases, St Hilaire[7] says: "Le

[1] Förster, A., *Die Missbld. des Mensch.*, 1861, s. 105, in atlas, Taf. xi. f. 16.

[2] King, *Med. Times and Gaz.*, 1858, vol. i. p. 527.

[3] Paull, *Lancet*, 1862, vol. i. p. 648.

[4] Widmer, *Corresp. Blatt. f. Schw. Aerzte*, 1888, s. 472.

[5] *Dissert. Anat. et Physiol. sur la sécretion du lait*, These de Paris, An. x. No. 53, p. 15.

[6] *Ueber die Angebornen Missbild. der gesam. weibl. Genitalien. Dissert. Erlangen*, 1842.

[7] *Histoire des Anomalies*, t. i. p. 710.

Docteur Lousier fait mention d'une dame qui privée d'une mamelle, transmit a sa fille le vice de conformation dont elle etait ellemême affectée." In accordance with this, almost all subsequent authors have referred to the case as an example of hereditary transmission; but, according to Puech,[1] Lousier never asserted this. All he said was, "J'ai connu une dame et une demoiselle chez les quelles la glande mammaire manquait complétement d'un côté."

In the two following cases congenital amazia was associated with total absence of the corresponding ovary. Both are reported by Scanzoni,[2] and I know of no others precisely similar, although, as I shall presently mention, there are on record many cases of micromazia associated with deficient ovarian development.

The first patient was a beggar woman, who died, aged 64, of tubercle. There was complete absence of the L. mamma, nipple, and areola, and at the necropsy no trace could be found of the L. ovary. She had been subject to amenorrhœa since the age of 27, but she had previously menstruated regularly.

The second patient was a girl, aged 18, who died of typhoid fever. In this case R. amazia was associated with complete absence of the R. ovary. She had menstruated regularly.

Absence of the breast may occasionally be caused by inflammation and injuries in the newly born. Puech[3] relates the case of a girl, aged 17, of whose L. breast there was hardly a trace, although the R. was well developed. This resulted from acute inflammation of the part at birth, followed by suppuration and the formation of a large abscess, which had to be incised.

Section 2.

When the defect is less complete than in the above cases, we get a very small imperfectly-developed gland, like the normal male breast, or smaller—*micromazia*. This condition, though rare, is of more frequent occurrence than any of the foregoing. The rudimentary organs are useless for lactation. Both mammæ may be affected, or only one.

[1] *Les mamelles et leurs anomalies*, p. 63.

[2] Kiwisch, *Klin. Vorträge über spec. Path. u. Therap. d. Krank. des weib. Gesch.*, Bd. iii. (1855), s. 47.

[3] *Op. cit.*, p. 90.

The deformity occurs independently, or associated with malformations of the adjacent chest or of the sexual organs, as in cases of amazia.

The following instances of micromazia of independent origin are related by Puech :[1]—

In a single woman, aged 24, there was complete absence of the projection of the bosom on both sides, while the nipples and areolæ were small and stunted. Menstruation set in at 15, but the catamenia were not regular until two years later. Her mother and sisters had well-developed breasts.

In another case it was noticed that both the breasts of a woman about to be confined were rudimentary; and they remained so after delivery, only a few drops of colostrum being secreted. Her mother had a similar deformity.

In the next case only one breast was affected. The patient was a young woman who married early, and was the mother of three children. At puberty the asymmetrical condition of the mammæ was first noticed; for while the R. attained its full size, the L. remained undeveloped. After each pregnancy the R. gave plenty of milk, but the L. none.

As examples of micromazia, associated with malformation of the adjacent chest wall, I can cite the following:—

In a single woman, aged 21, seen by Engeström,[2] all that existed of the L. breast was a small stunted papilla, and under it "un petit amas de graisse, mais si insignificant, qu'il ne forme même pas une eminence." The sternal part of the left *pectoralis major* muscle was completely absent. She was otherwise well developed.

In another case by the same author, the patient was an emaciated, phthisical woman, aged 27, who had recently been delivered of her second child. The left breast was well formed and full of milk; but the right was very small, although its nipple and areola were normal, and only a few drops of milky fluid could be expressed from it. This secretion ceased shortly afterwards. Most of the sternal part of the corresponding *pectoralis major* muscle was absent. No history of any malformation in others of her family.

[1] *Op. cit.*, p. 89. [2] *Ann. de Gyn.*, t. xxxi. (1889), p. 84.

In a similar case, seen by Ebstein,[1] the R. breast was not larger than a hemp-seed. The sternal part of the *pectoralis major* and the whole of the *pectoralis minor* muscles were wanting.

Grüber[2] saw a young lady, aged 18, in whom nearly all of the costo-sternal part of the R. *pectoralis major* muscle was wanting; and whose R. breast was represented only by a malformed nipple, surrounded by a large areola, beneath which a thin glandular *plâque* could be felt. Menstruation set in at 15, but the R. mamma never developed, although the L. attained a large size. She was otherwise well formed; but thin and phthisical.

The following cases illustrate the connection between micromazia and defective development of the generative organs. In many of the individuals thus affected the secondary sexual characters are imperfectly evolved, and there is often manifest an approach to the male type of organisation.

In a woman, aged 26, seen by De Sinetz,[3] both breasts were like those of a girl before puberty; they had no areolæ, and their nipples were hardly perceptible. The uterus and vagina were of an equally stunted, infantile type.

In a case recorded by Greenhow,[4] the patient was an unmarried servant girl, aged 22, who was very flat in the mammary regions, and on careful manipulation no trace of either gland could be felt, although she had a small stunted nipple and areola in the usual position on each side. She was of spare, girlish aspect, and had never menstruated. The pelvis and hips were small, as also was the mons veneris, on which there were but a very few hairs. The vagina was small and narrow, with a well-marked hymen. The os and cervix uteri were absent; but on rectal examination a small hard lump was detected in the position of the body of the uterus. The ovaries could not be felt. In addition, she had bifid sternum, associated with congenital malformation of the heart, the exact nature of which could not be determined. She suffered much from palpitation of the heart, cough, and dyspnœa.

Pears[5] has related the case of a dwarfed woman, aged 29,

[1] *Deutsch. Arch. f. Klin. Med.*, vi. s. 283.
[2] *Arch. f. Path. Anat.*, cvi., 1886. s. 501.
[3] *Traité de Gyn.*, 1884, p. 947.
[4] *Med. Chir. Trans.*, vol. xlvii., 1864, p. 195.
[5] *Philosophical Trans. R. S.*, Lond. 1805, p. 225.

who was only 4 feet 6 inches high. Her breasts and nipples were like those of a male. She had never menstruated. There was no hair on the pubes, nor any other of the signs of puberty. "She always expressed an aversion to young men who were too familiar with her." She had been subject to violent fits of coughing and convulsions for several years, and in one of these she died. At the necropsy the uterus was of the infantile type. "The ovaria were so indistinct as rather to show the rudiments which ought to have formed them, than any part of their natural structure." Analogous instances have been recorded by Baynham,[1] Caillot,[2] Rénauldin,[3] and others.

According to Puech,[4] the infantile condition of uterus, which generally goes with absent or rudimentary ovaries, is nearly always correlated with defective mammary development.

When, however, the uterus is really absent, the mammæ are, as a rule, unusually well developed; and in these cases the ovaries are generally normal.[5] This is just the converse of what happens in males, in whom absence of the testes is usually associated with exaggerated mammary development.

Section 3.

Congenital absence of the nipple—*athelia*—is much commoner than any of the foregoing anomalies.

Inasmuch as this structure is formed by upheaval of the area of skin perforated by the ducts of the nascent gland, it follows that in true athelia none of the nipple structures—skin, connective tissue, vessels, nerves, ducts, &c.—are really wanting, as Duval erroneously supposed; but we have to do simply with failure of the normal mammillary outgrowth. This condition is usually unaccompanied by any other malformation, and it generally affects both breasts. I lately saw a healthy young lady, aged 18, with marked defect of this kind, associated with eczema of the malformed parts. Her breasts were large,

[1] *London Medical Gazette*, vol. iii. 1829, p. 72.

[2] *Mém. de la Soc. Méd. d'Emulation*, Paris, t. ii. (1798), p. 270 *et seq.*

[3] *Séances de l'Acad. Roy. de Méd.*, Fev. 28, 1826.

[4] For two well recorded cases confirmatory of this view, *vide* Warren, *Surgical Observations*, Boston, 1867, p. 305.

[5] *Les ovaries et leurs anomalies*, Paris, 1873; see also *Op. cit.*, p. 91.

and otherwise well developed; but their nipples were completely absent, and in the place of each was a small transverse groove, surrounded by a diminutive, stunted areola. The affection dated from birth. There was no family history of any similar deformity. Menstruation was normal.

Analogous cases have been seen by Cruveilhier,[1] Davis,[2] and many others. Persons affected usually have plenty of milk during lactation, but they are nevertheless unable to suckle their children.

Like other forms of defective mammary development, athelia is sometimes found associated with defective development of the genital organs, as in the following remarkable case, related by Chambers.[6] The patient, aged 24, had the general appearance and external sexual organs of a female, including the well-developed female bust. She had always passed as a female, having been engaged as a housemaid. Both nipples were completely absent, and the place of each was occupied by a small rose-coloured spot, representing the areola. There was no hair in the pubic region. The mons veneris was ill developed. The vagina was small, and ended in a *cul-de-sac*, and there was no trace of uterus or ovaries. She had never menstruated, nor had she ever experienced menstrual molimina. In each inguinal region she had an irreducible congenital hernial tumour, each of which contained a firm circumscribed body, thought to be an ovary. These bodies were excised, but on microscopical examination after removal they proved to be testes.

In such cases as the foregoing we evidently have to do with defective nipple evolution almost *ab initio*. In consequence, we often see persistence of the depression which, in the normal course of development, marks the site where the nipple will subsequently arise. When the morbid process supervenes at a somewhat later period, then we get some of the various minor degrees of congenital mammillary imperfection which are of such frequent occurrence: thus the nipple may be small, stunted, flattened, depressed, or otherwise malformed.

Such conditions are fruitful sources of trouble during lacta-

[1] *Traité d'Anatomie Descript.*, ed. 1874, t. 2me, p. 525.

[2] *Medical Times*, vol. i., 1852, p. 250.

[3] *Trans. Obstet. Soc. Lond.*, vol. xxi. (1879), p. 256.

tion; they are, indeed, the chief causes of the acute inflammations and abscesses so common at that period.

According to Birkett,[1] out of 97 cases of acute mammary abscess, there was imperfect development of the nipple in 48, or in half the total number.

Congenital imperfection of the nipple is often found in association with neoplasms.

Of 137 consecutive cases of primary *cancer* of the female breast, I found congenital malformation of the nipple in 19 (13·8 per cent.). And of 42 cases of *adenoma*, there was similar malformation of the nipple in 10 (23·8 per cent.).

Absence of the nipple is not infrequently ascribable to traumatisms in the newly born, such as bites, burns, wounds, abscesses, ulcers, &c.

When the nipple is deficient, the areola is often stunted or absent; but it is a great rarity to find the areola absent when the nipple is well formed.

In a case of this kind described by Dr O'Flynn,[2] the patient was a healthy-looking woman, aged 30, the mother of seven children, who, when she came under observation, was pregnant for the eighth time. Her breasts were small and flaccid like those of a girl at puberty, and during her previous pregnancies they had never enlarged nor given any milk. The nipples were prominent, but neither of them had an areola. Her mother's breasts were rudimentary, and though she had eleven children, no milk was ever secreted.

Section 4.

At the climacteric period the breasts normally undergo atrophic changes, which usually affect the fibro-fatty as well as the glandular elements. The degree to which this involution takes place, and the age at which it sets in, are variable. Occasionally these changes begin at a very early period of life, and proceed to such a degree as to constitute veritable disease.

A well-marked instance of this affection lately came under my notice in the person of a young widow, aged 30, both of

[1] Art. "Diseases of the Breast," *Holmes' System of Surgery*, vol. iii. (1883), p. 435.

[2] *Dublin Medical Press*, vol. liv. (1865), p. 312.

whose breasts were small and flaccid, like those of a thin old woman. Before the death of her husband, two years previously, she had a well-formed bust; but since then the breasts have gradually wasted away. Her general health was good, but she was less plump than formerly. The catamenia were scanty, but regular. Shortly after the death of her husband she lost her only child.

Here grief, and suppression of the sexual function, seem to have been the determining causes.

Another somewhat similar case[1] is that of a married lady, aged twenty-four, who had noticed her breasts wasting away for four months. They were formerly well developed, and she knew of no cause for their atrophy. The general health was good, there was no organic disease, and menstruation was normal. On examination the mammæ were found very atrophied. She had been married for two years, and one year after marriage gave birth to a child, which died a few weeks afterwards of bronchitis.

In sterile women, and in those who have neglected to suckle their offspring, the mammæ often shrink to quite small proportions. Reynolds[2] has given an account of a woman, aged 21, who, having suckled her first child only for a few weeks, soon afterwards saw both her breasts completely disappear, so that not a vestige of them could be felt. Yet when she became pregnant again, they both enlarged to a fair size and gave milk; but, as she neglected to suckle, the glands soon wasted away as before—and so after each pregnancy.

There can be no doubt that the habit of weaning children does in the long run tend to defective mammary development. Disuse leads to atrophy and the result is inherited.

De Sinéty[3] says he has seen several examples of this in the women of families whose children have been weaned for several generations; and two of them, although very prolific, were unable to suckle, through failure of lacteal secretion.

Altmann[4] has recently published an interesting essay on this subject.

[1] *Lancet*, vol. i. (1884), p. 782.
[2] *Ibid.* vol. i. (1884) p. 331.
[3] *Traité de Gyn.*, p. 918.
[4] *Arch. f. Path. Anat.*, Bd. cxi. (1888), s. 318.

Atrophy of the ovaries, and morbid conditions interfering with their integrity, induce a somewhat similar condition. Thus in Potts'[1] well-known case the catamenia became suppressed, the mammæ wasted, and the body got thinner in a healthy and plump young woman, aged 23, each of whose ovaries presented as a hernial swelling at the inguinal rings, and were excised in consequence of their incapacitating the patient from work.

Other alleged causes of mammary atrophy are prolonged and excessive suckling, exhaustive illnesses, and the internal administration of iodide of potassium in large doses.

[1] *Surgical Works*, vol. iii. p. 329.