

FIBROADENOMA IN A SUPERNUMERARY MAMMARY GLAND OF THE VULVA

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Congenital supernumerary breasts are not uncommon. A review of the literature shows that this anomaly has been known to occur in almost every part of the body, and is usually located between the groin and the axilla along the embryological "milk-line". But the location of supernumerary breast in some part of the vulva is extremely rare and very few cases have been reported so far.

Of the 430 cases recorded by Deaver and McFarland (1918), 90% were located in the axilla, chest or abdomen, the remaining 10% being scattered throughout other parts of the body. Only in one instance was the vulva the seat of this malformation. Series of cases collected from the literature were reviewed by different authors from time to time. De Cholnoky (1939) has reviewed the literature on the subject up to 1939, later Weinshel and Demakopoulos (1943) have brought the review up-to-date up to 1943. More re-

cently Tow and Shanmugaratnam (1962), after a careful search of the literature found only 15 recorded cases and added one case of their own, making a total of 16 cases.

Table I shows the list of case reports that appeared in the literature up to date on supernumerary breast in the vulva.

Of these, 7 cases of vulval breasts were discovered during pregnancy (Blasio, 1905; Bell, 1926; Purves and Hadley, 1927; McFarland, 1931; Mengert, 1935; Looney *et al*, 1959; Tow and Shanmugaratnam, 1962); 2 cases presented with milky discharge (Hartung, 1872; Bell, 1926); 1 case with milk-cyst formation (Bergner, 1934); 1 with abscess formation (McGee, 1925-26), 2 undergoing adenocarcinomatous change (Greene, 1936; Hendrix and Behrman, 1956) and 1 with sarcomatous change (Crumpe, 1854). Only 3 cases of supernumerary breast with fibroadenoma occurring in the vulva have been reported up till now. The first case was reported by Friedel in Germany (1932) and subsequent cases by Fisher (1947) and Siegler and Gordon in U.S.A. (1951).

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TABLE I

List of case reports on supernumerary breast in the vulva

No.	Author	Vulval involvement	Associated changes
1.	Crumpe (1854)	.. Unilateral	With sarcomatous change.
2.	Hartung (1872)	Unilateral	With milky discharge.
3.	Blasio (1905)	Bilateral	Developed at puberty and enlarged during pregnancy.
4.	McGee (1925-26)	.. Unilateral	With abscess formation.
5.	Bell (1926)	.. Unilateral	Discovered during 3rd pregnancy with milky discharge.
6.	Purves and Hadley (1927)	Bilateral	Enlarged during pregnancy.
7.	McFarland (1931)	Unilateral	Discovered during 3rd pregnancy.
8.	Friedel (1932)	.. Unilateral	With fibroadenomatous change.
9.	Bergner (1934)	.. Unilateral	With milk cyst formation.
10.	Mengert (1935)	.. Bilateral	Discovered during 4th, pregnancy and enlarged in puerperium.
11.	Greene (1936)	.. Bilateral	With adenocarcinomatous change
12.	Fisher (1947)	.. Unilateral	With fibroadenomatous change.
13.	Siegler and Gordon (1951)	Unilateral	With fibroadenomatous change.
14.	Hendrix and Behrman (1956)	Unilateral	Adenocarcinomatous change.
15.	Loony et al (1959)	Bilateral	Discovered during 3rd, pregnancy.
16.	Tow and Shanmugaratnam (1962).	Unilateral	Discovered during 5th pregnancy.

In so far as we have been able to ascertain from the review of the English literature of supernumerary breasts only 16 cases of vulval involvement have been reported. The present report brings the total number of known cases to 17 and this is the 4th. case of fibroadenoma occurring in a supernumerary mammary gland of the vulva.

CASE REPORT

Mrs. B. B., Regd. No. 30160/67, a married Hindu Assamese woman, of 26 years of age, was admitted into the surgical ward, Assam Medical College Hospital, under the care of one of the authors (M.A.C.) with the complaint of a lump over the left side of the vulva, associated with ulceration of 2 months' duration. It started as a nodule, which rapidly increased in size with ulceration of the overlying skin associated with foul smelling discharge, which made her very worried as she was also having amenorrhoea of 36 weeks' duration. Otherwise she was well and her past history was non-contributory.

Local examination revealed a large ovoid

pedunculated mass of 10 c.m. in diameter, situated on the vulva, just on the left side of the mons pubis. It was freely mobile, firm with a knobby feel and slightly tender. The whole covering surface was ulcerated and covered with necrotic slough with an offensive discharge. The mass was well localized and attached to the vulva with a peduncle. The skin was present and moved freely over the peduncle. The left labium majus was oedematous.

The uterus was enlarged to a size consistent with 36 weeks' pregnancy. Foetal heart sounds were heard.

Under general anaesthesia the whole pedunculated mass was excised and the wound was closed. The specimen was sent for histopathological opinion. The same evening she started having labour pains and delivered a normal full-term female baby. Convalescence was uneventful.

Pathological Report—Gross appearance:—The gross specimen measures 8.5 c.m. by 5.5 c.m. by 5 c.m. (Fig. 1). It is encapsulated and the whole surface is ulcerated and covered with necrotic slough. Other parts of the surface are greyish white and are gently lobulated, feel firm and elastic. Cut surface is uniformly greyish white and solid. Several firm yellowish white nodules

are present, which are distinctly raised from the surrounding areas.

Microscopic:- Sections from different areas stained with haematoxylin and eosin reveal distinct structure of well developed mammary gland in a state of activity, containing ducts, acini, fibro-areolar tissue and fatty tissue. (Fig. 2, 3). The ducts are moderately dilated and show marked proliferation with formation of acini. In general there is no secretory activity. The intralobular connective tissues are broken up and reduced to film-like partitions between the adjacent acini. The stroma is infiltrated with lymphocytes and plasma cells.

Sections from the nodular areas present a pattern of benign pericanalicular fibroadenoma. It is composed of proliferated ducts surrounded by a zone of well developed fibrous connective tissue. The ducts are not dilated or distorted. Special staining with Van Gieson and Mallory have confirmed these findings.

Such a picture suggests a supernumerary mammary gland in the vulva in a state of activity due to pregnancy and pericanalicular fibroadenoma.

The report aroused much interest and additional information was sought for concerning the accessory breast. The patient was re-examined on the 8th day following operation. She was a healthy adult with normally developed breasts, normally situated nipples and areolas. Montgomery's tubercles were prominent. Two small swellings were noticed in both the axillae with rudimentary nipples (Fig. 4). Milk-like fluid could be expressed from the rudimentary nipples. A thorough search over the whole body did not reveal any other supernumerary breast or nipple. She stated that during the postpartum period, the axillary swelling along with the normal breast became tense and painful for about 4 days. Her 3 previous pregnancies had resulted in normal deliveries at term. On further interrogation she gave a history of similar axillary swellings during her previous 3 pregnancies and lactations with milky discharge. These subsided afterwards and became impalpable. But no such swelling over the vulva was noticed till the last 2 months of her 4th pregnancy. There was

no familial history of supernumerary breast or any other congenital anomalies.

Discussion

The occurrence of supernumerary nipples (polythelia) and of supernumerary breast containing actual mammary gland tissue (polymastia) is well known; but involvement of the vulva is extremely rare. They may be unilateral or bilateral and may occur in both male and female individuals. They are said to be more common in coloured than in white individuals. There is a superstition that polymastia is usually associated with multiple births, but there is no substantial proof of this in the literature. Deaver and McFarland (1918) demonstrated a strong hereditary factor in the causation of this anomaly. De Cholnoky (1939) in a review of literature of supernumerary breast reported that one additional breast occurs in from 60 to 65% of the cases, two in 30 to 35%, three in 3.5 to 4% and four in 1.5 to 2%.

In general, more accessory mammae are noted on the left side of the body than on the right, and a great many more are observed below the normal breasts than above them. Those situated below the normal breasts are usually medial to the normal breasts, whereas those above the normal site are usually found lateral to the normal breast region. Those situated laterally are more apt to be of considerable size and to undergo normal lactation than those situated medially. In the great majority of cases the extra breast or nipple tends to appear as one would expect along the normal course of milk line (fig. 5a). Nevertheless, sporadic cases oc-

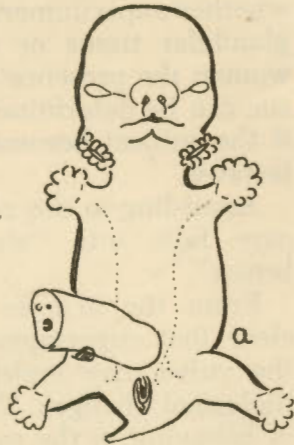


Fig. 5(a)

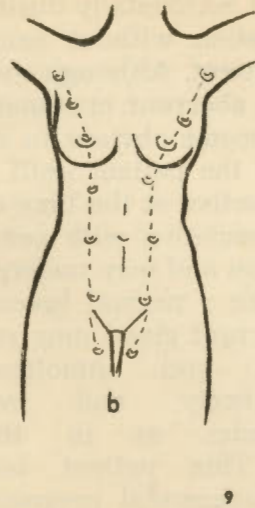


Fig. 5(b)

Fig. 5(a)—Schematic diagram showing the position of the "milk line" in a mammalian embryo.

Fig. 5(b)—Showing the commonest sites at which supernumerary nipples and mammary glands occur in woman.

cur in both sexes in which the breasts or nipples are present in areas not normally involved in mammary gland formation. Occurrence of supernumerary breasts has been reported on cheek, neck, ear, tragus and helix, upper part of left arm, thorax and abdomen, shoulder, back, lumbar region, public and inguinal regions, flank and hip, thigh and Scarpa's triangle, thigh and dorsolateral aspect, buttock, vulva, labia majus and minus. The composite diagrams (Fig. 6) indicates all the locations at which aberrant mammary glands or nipples have been reported, showing wide departures from the conventional positions along the milk line (Patten, 1955). Accessory breasts located in these atypical regions are rare and usually asymmetrical. In most cases, these abnormal organs are very rudimentary and suppressed in their development and are often represented by a minute

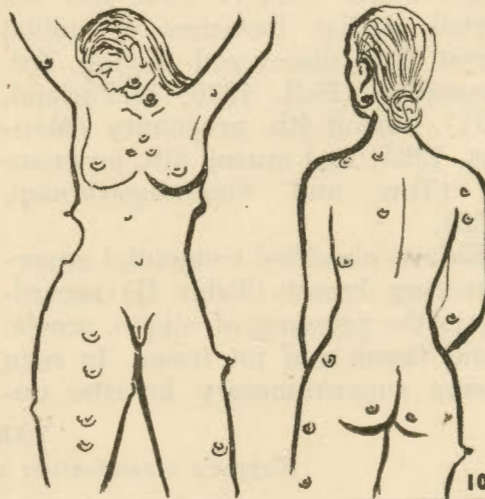


Fig. 6

Schematic diagram showing collected on single subject all the unusual locations in which supernumerary nipples and breasts have been reported (Patten, 1955).

nipple or pigmented areola. The supernumerary breasts are

usually diagnosed accidentally during physical examination without being noticed by the patient. Although congenital in origin, aberrant mammary gland may not become obvious to attract attention of the patient until it is enlarged or is active at the time of puberty or in association with pregnancy and lactation and may undergo cyclic changes like a normal breast. At times the aberrant gland may remain quiescent and unnoticed throughout puberty and repeated pregnancies as in the present case. This patient has gone through 3 successful pregnancies and all her babies were breastfed normally. It was not until the latter part of her 4th. pregnancy, that she discovered the mass over her vulva, particularly when it became rapidly large. Other authors have also reported similar instances of vulval breast first discovered during 3rd. pregnancy (Bell, 1926; McFarland, 1931), during 4th. pregnancy (Menger, 1935) and during 5th. pregnancy (Tow and Shanmugaratnam, 1962).

Kajava classified congenital supernumerary breast (Table II) according to the presence of nipple, areola, gland tissue and fat tissue. In men having supernumerary breasts, un-

less microscopic examination is made, it will be impossible to determine whether supernumerary breasts have glandular tissue or not; whereas in women the presence of glandular tissue can be determined with certainty if the subject becomes pregnant and lactates.

According to this classification, this case falls into "aberrant type of breast".

From the articles reviewed it is clear that supernumerary breasts in the vulva may undergo various pathological changes. They are capable of behaving in the same way as normally situated breasts and respond to hormonal influences at puberty, during menstrual cycle, pregnancy and lactation. A milky discharge was reported by Hartung (1872) and Bell (1926). Complications common to normal breasts have also been encountered, such as abscess (McGee, 1925-26) and milk cyst formation (Bergner, 1934). Neoplastic changes, both benign and malignant, may supervene. Fibroadenoma has been reported by Friedal (1932), Fisher (1947), Siegler and Gordon (1951) and adenocarcinoma by Greene (1936) and Hendrix and Behrman (1956). Aberrant mammary tissue in the axilla without nipple formatio.

TABLE II
Kajava's classification of supernumerary breasts

Type of breast.	Nipple	Areola	Gland tissue.	Fat tissue.
Complete	Present	Present	Present	Present
Supernumerary	Present	None	Present	Present
Supernumerary	None	Present	Present	Present
Aberrant	None	None	Present	Present
Pseudomamma	Present	Present	None	Present
Polythelia	Present	None	None	None
Polythelia areolis	None	Present	None	None
Polythelia pilosa—Only a patch of hair is present.				

is more prone to malignant change than in a supernumerary breast in which the frequency of tumour occurrence is seemingly the same as with normal breasts.

Fibroadenoma is a frequent tumour in a normal breast and is common in young women between 20 to 30 years of age. Their origin is in some way associated with oestrogenic stimulation of breast tissue. Pregnancy and lactation due to intense hormonal stimulation cause them to grow rapidly. The interesting finding in this case is that the breast tissue in the vulva has undergone pericanalicular fibroadenomatous change. While the normal mammary gland has reacted to the usual hormonal changes during pregnancy, no such changes are present in the areas affected by fibroadenoma. This may be explained by the fact that the neoplastic areas have become hormone independent. The fibroadenomatous change and active proliferation as a result of pregnancy have contributed to the rapid enlargement of the ectopic breast tissue with ulceration of the overlying skin.

Clinically, these ectopic breasts may resemble and are frequently diagnosed as lipomas. They should also be differentiated from chronic lymphadenitis due to syphilis, tuberculosis, Hodgkin's disease, lymphosarcoma, metastatic carcinoma and hypertrophied sudariparous and sebaceous glands.

The presence of breast tissue in some part of the vulva is explainable on the embryological basis. In a mammalian embryo the breast arises from a pair of band-like ectodermal thickening "primitive milk line",

which extends between the upper and lower limb buds on either side of the midline along the ventro-lateral body wall. These two "primitive milk lines" are detected in a 10 m.m. embryo and run from the axilla to the groin (Fig. 5a). In the course of development, towards the 2nd month of intrauterine life, these lines undergo fragmentation and the epithelium heaps up at regular intervals with the appearance of a series of little ectodermal eminences called "anlagen" or rudiment of future mammary gland. The number of "anlagen" is always in excess of requirements. The "anlagen" appear in the embryos of all mammals to be developed or suppressed and the location and their number being determined by phylogenetics of the mammal in question. In man the "anlagen", on each side in the pectoral region at the level of the 4th. rib, are retained to form future pectoral breasts and the remaining redundant "anlagen" are suppressed and disappear. In the human species the different heights at which the breasts are developed in different racial groups is indicative of the utilization of slightly different levels of the milk line in their development. Normally this variation occurs within rather narrow limits of the pectoral region. Incomplete or failure of suppression of redundant "anlagen" leads to the appearance of supernumerary breasts, which may be located anywhere along the milk lines on one or both sides (Fig. 5(b)).

This anomaly can also be explained in terms of Darwin's theory of "descent of man", which considers it as an atavistic or reversionary mani-

festation, in which, for some obscure reasons, a remote ancestral characteristic unexpectedly makes its appearance. Since the breasts of Cetacea (whales, dolphins, porpoises, lemurs etc.) are normally located in the vulva, the presence of vulval breasts in man appears to support this atavistic theory.

According to Siegler and Gordon (1962), the anatomical situation of the lesion and its histological structure makes a differentiation from a lesion of the vestiges of Wolffian or Mesonephric ducts almost impossible. But the character of the lining epithelium and the presence of small clusters of acini resembling those of a breast tissue, makes it the most acceptable histological diagnosis.

These however, do not adequately explain the occurrence of accessory breasts in unusual locations, such as neck, face, ears, arms, legs, buttocks etc. In these awkward situations, the anomaly may be attributed to misplacement of redundant "anlagen" (Deaver and McFarland). Champneys believes that supernumerary mammary glands are enlarged sweat glands or mammary differentiation of apocrine sweat glands (and not true breasts), a theory which was supported by Tandlaus's observation of a patient in whom there was complete failure of development of the breast and nipple, together with total absence of sweat glands all over the body and partial absence of sebaceous glands.

The harmless existence of the supernumerary breasts usually requires no medical or surgical treatment. They serve no useful function in man, but rather are a potent source of

disease, e.g. pain, abnormal discharge, infection, neoplasm and malignant change. Therefore, when discovered, they should be promptly removed. Besides there are cosmetic and psychological reasons for a surgical intervention.

Summary

A rare case of supernumerary mammary gland in the vulva with fibroadenomatous change, discovered during the fourth pregnancy, has been presented, which has been diagnosed by histological examination after resection.

The literature of supernumerary breast is reviewed and its incidence, classification, location and embryology are discussed.

Acknowledgement

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References

1. Bell, J. Warren: *Am. J. Obst. and Gynec.*, 11: 507, 1926.
2. Bergner, E. A.: *Acta Obst. Gynec. Scand.*, 14: 205, 1934.
3. Blasio de T.: *Arch. Psichiat. Torino.*, 26: 171, 1905.
4. Champneys, F. H.: *Med. Chir. Tr. London*, 69: 419, 1886.
5. Crumpe, F.: *Dublin Quart. J. Med. Sci.*, 17: 466, 1854.
6. Deaver, J. B. and McFarland, J.: *The Breast. Its Anomalies. Its Diseases and Their Treatment*, London, 1918, William Heineman.
7. De Cholnoky, T.: *Arch. Surg.*, 39: 926, 1939.

8. Fisher, J. H.: Am. J. Obst. and Gynec., 53: 335, 1947.
9. Friedel, R.: Virchows Arch. Path. Anat., 286: 62, 1932.
10. Greene, H. J.: Am. J. Obst. and Gynec., 31: 660, 1936.
11. Hartung: Inaugural Dissertation, Erlangen Germany, 1872, (Quoted by Greene, 1936).
12. Hendrix, R. C. and Behrman, S. J.: Obst. and Gynec., 8: 238, 1956.
13. Kajava, Y.: Duodecim, 31: 143, 1915.
14. Looney, C. M., Reichman, S. C. and Noel, O. F.: Am. Surg., 25: 219, 1959.
15. McFarland, J.: Arch. Path., 11: 236, 1931.
16. McGee, W. A.: Virginia Med. Mth., 52: 179, 1925-26.
17. Mengert, W. F.: Am. J. Obst. and Gynec., 29: 891, 1935.
18. Patten, B. M.: Human Embryology, ed. 2, London, 1955, p. 240, J. & A. Churchill Ltd.
19. Purves, R. and Hadley, J. A.: Brit. J. Surg., 15: 279, 1927.
20. Siegler, A. M. and Gordon, R.: Am. J. Obst. and Gynec., 62: 1367, 1951.
21. Tandlaus: Quoted by Weinshel and Demakopoulos, 1943.
22. Tow, S. H. and Shanmugaratnam, K.: Brit. Med. J., 2: 1234, 1962.
23. Weinshel, L. R. and Demakopoulos, N.: Am. J. Surg., 60: 76, 1943.

Figs. on Art Paper VI