Fibro-adenosis of Ectopic Breast Tissue in the Vulva and Perianal Region

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SUMMARY

A case of bilateral vulval and perianal fibro-adenosis in ectopic breast tissue is recorded. The literature is reviewed, the theories of origin are discussed, and the clinical features and management are described.


A review of the literature indicates that ectopic breast tissue is not uncommon. However, only 24 cases of ectopic breast tissue on the vulva have been recorded, and in only 1 case was bilateral fibro-adeno is of vulval ectopic breast tissue reported. No previous reported cases of fibroadenosis in breast tissue around the perianal region could be found.

This appears to be the first report of ectopic breast tissue in the vulva in the South African medical literature.

CASE REPORT

A Bantu patient, aged 25 years, gravida 2, para 2, complained of tender swelling of the vulva and pain with sexual intercourse. She had first noticed tender nodules on the vulva during the 16th week of her second pregnancy. The tumours gradually enlarged towards the end of pregnancy and became very tender, causing dyspareunia. She had a normal vaginal delivery at term and breast-fed her baby for 3 months. At the time of her admission to hospital the baby had been off the breast for 4 months and the patient had started menstruating normally again.

On examination it was noted that there was still active secretion from the breasts. The patient had small tender nodules on both the labia majora and around the anus (Fig. 1). Closer examination showed that there were altogether approximately 12 small tender nodules on the vulva and perineum, involving both labia majora and labia minora, and extending from the mons veneris next to the clitoris down to the anus. The nodules varied in size, from 0.5 cm to 1.5 cm in diameter. All were covered with normal skin except one small tumour on the right labium majus; this nodule was denuded of skin and had a reddish colour with an offensive discharge. Some of the nodules felt cystic on palpation, others were firm and all were tender and painful. Around the anus there were a further 11 small nodules resembling haemorrhoids.

Management

Laboratory investigations, which included urine analysis, haemoglobin estimation and blood count, fasting blood sugar, bilharzia complement-fixation test, serological tests for syphilis, cytologic examination of vaginal smears and X-ray examination of the sella turcica, were all normal or negative.

Two of the nodules on the vulva were removed for histological examination. The histological report was as follows:

'In the dermis numerous branching ducts, lined by a double-layered epithelium, were arranged in a lobular

Fig. 1. Gross appearance of vulval and perianal ectopic breast tissue.
pattern. Focal areas of adenosis were observed, some of the ducts were cystically dilated and a few were lined by "pink epithelium". Fairly marked peripheral fibrosis was present in the lobules and a focal lymphocytic infiltrate was seen.

'The histological picture in both vulva nodules is compatible with mammary tissue showing the features of fibro-adenosis.'

After the histology became known the patient was questioned further, and she admitted that the tumours became larger and more tender premenstrually. The patient was given a course of stilboestrol to attempt to depress the ectopic breast tissue, but there was no evident diminution in size, though the secretion from the normal breasts became less. Further treatment consisted of local excision of the vulval and perianal nodules. Most of the labia majora were removed, but the clitoris and labia minora could be spared. There was no difficulty in shelling the nodules out of the subcutaneous tissues. Before excision of the perianal nodules, the anus was first dilated and care was taken to prevent any anal stenosis. The remaining perianal nodules were removed on a separate occasion 10 days later; during this interval the patient was given Primolut-N 15 mg daily, to determine whether it would have any histological effect on the fibro-adenosis.

The histology report following the local excision of the tumours was virtually the same as the previous report and mentioned fibro-adenosis of ectopic breast tissue in the perianal tumours as well (Figs. 2 and 3). The Primolut-N had no histologic effect on the fibro-adenosis.

Recovery was uneventful and a satisfactory cosmetic and functional result was obtained.
DISCUSSION

Approximately 1 - 6% of all women show polymastia and in 90% of the cases the accessory breast tissue is found in the axilla or on the chest or abdomen. In the remaining 10% of cases the ectopic breast tissue is found in other parts of the body. Aberrant breast tissue on the vulva is extremely rare and in the 430 cases of polymastia reviewed by Deaver and McFarland in 1918, there was only 1 such case. In the 24 cases found in the literature there was no mention of fibro-adenosis of ectopic tissue situated peri-anally, although Paget’s disease is not uncommon in this region. The incidence of polymastia is low in Caucasians, whereas the condition seems to be relatively frequent among Japanese women.

Embryology

At approximately the 6th week of gestation, when the human embryo is 6 - 7 mm long, bilateral thickening called milk-lines or mammary ridges appear. These lines stretch along the ventrolateral aspects of the body from the axilla to the inguinal region and end in the medial aspects of the thighs. These ectodermal bands soon become part of and form a series of tiny buds or mammary Anlagen. The caudal two-thirds of the mammary ridges usually disappear by the 20 mm stage and only the pectoral parts of the Anlagen remain.

Fig. 4. Embryologic milk-lines (see text).

Supernumerary or ectopic breasts develop when the caudal portion of this milk-line persists. The distribution of aberrant mammary glands or nipples which are situated away from the conventional position of the mammary ridges indicate that not all cases can be explained by the ‘developmental rest concept’. To explain supernumerary breasts lateral to the milk-line, Castano mentions that Schultz suggested that there may be a dorsal emplace-

ment of the mammary ridges which at a later stage moves progressively towards the ventral surface as the result of a special growth process.

Theories of Origin

In attempting to explain the presence of aberrant breast tissue on the vulva it is necessary to theorize.

According to Darwin’s theory, it is suggested that the presence of aberrant breast tissue on the vulva is a further atavistic expression of our ancestral history or a reversion to a state pertaining to lower animals. Some animals (e.g. the sow and the bitch) have a series of nipples spread almost the entire length of the milk-line. In fruit bats and flying lemurs mammary glands occur in the axillary regions; in cetaceans (whales, dolphins and porpoises) breasts normally occur on the vulva.

Others attribute vulval breast tissue to a misplacement of embryonic mammary Anlagen. The presence of breast tissue on the vulva can possibly be explained in this manner but the occurrence of such tissue in the labia and especially perianally remains a problem.

Mengert and Burger and Marcuse believed that a fibro-adenoma is due to mammary differentiation in the apocrine sweat glands found on the vulva and sometimes also perianally. Foushee and Pruitt, on the other hand, suggested that it originates from ectopic breast tissue. Differentiating such tissue from vestiges of mesonephric ducts may present a difficult histological problem.

There is a strong familial trait in patients with supernumerary breasts. Our patient had no such family history and both her babies were without visible supernumerary breasts.

Pathology

Fibro-adenoma was the commonest pathological change in the 24 patients with vulval breast tissue reported to date. In all cases the distribution was unilateral except for Hassim’s case where the patient had bilateral fibro-adenoma on the vulva.

The histological diagnoses of the 24 cases were as follows: fibro-adenosis (9 cases); lactating breast tissue (6 cases); adenocarcinoma (4 cases); cystic disease (4 cases) and sarcoma (1 case).

Clinical Features

The lesions usually remain unnoticed and asymptomatic until a strong hormonal stimulus such as pregnancy supervenes. Most of the reported cases presented for the first time after the third or fourth pregnancy and the patients sought medical attention at about the 14th - 20th week of gestation. The usual complaint is a painless tumour or tumours on the vulva, sometimes associated with a milky or offensive discharge and with premenstrual tenderness. Dyspareunia and actual pain is usually a manifestation of inflammation of the ectopic breast tissue. In our patient, the perianal tumour caused slight pain with defaecation during the premenstrual phase.
Differential Diagnosis

Ectopic breast tissue on the vulva must be differentiated from the following conditions also causing tumours on the vulva or perianal region: Bartholin cyst or abscess; sebaceous cysts; epidermoid cysts; fibroma—a relatively common condition; lipoma; papilloma, including condyloma acuminata; haemangioma; mesonephric duct cysts; bilharzia granuloma; haemangioma; neurofibroma; leiomyoma; ganglioneuroma; lymphangioma; endometrioma; granular cell myoblastoma; naevis syringocystadenomatoj: papillferus and haemorrhoids.

Treatment

Once the diagnosis of ectopic breast tissue on the vulva or perianal region has been made, total removal of this tissue is necessary. Complications such as cyst formation, abscess formation or malignant change can occur in ectopic breast tissue. If all the accessory breast tissue is not removed, the condition may recur. Complete removal ensures asymptomatic relief and affords a good long-term prognosis.

I should like to thank Dr W. Kenny, Superintendent of H. F. Verwoerd Hospital, for permitting the publication of this case report.

REFERENCES

Book Reviews : Boekbesprekings

BIRTH CONTROL

It is inevitable that any book on birth control in America should, to some extent, also be a biography of that remarkable woman, Margaret Sanger. David M. Kennedy places the birth control movement in the larger context of the social revolution that was taking place in the USA at the time. He describes Mrs Sanger's dynamic leadership, combined with a disarming graciousness and charm, and her obsession with the idea that she was fighting against visible enemies in the Church, the State, and, to a lesser extent, the officials of the Church, the State, and, to a lesser extent, the officials of the medical profession who balked at her non-medical leadership in a medical area. Inevitably canners were drawn across her path, President Theodore Roosevelt advancing in 1905, the theory of 'race suicide' and the Catholic Church of America, the complicity of Russian agents in the birth control movement. But in the end she won her case and by 1940 birth control movement in the larger context of the social revolution that was taking place in the USA at the time.

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PREGNANCY PHYSIOLOGY

In the 7 years which have elapsed since publication of the first edition of this book much new information has been gathered about the physiology of normal pregnancy. This new information, necessitating a great deal of revision of the old text, has been succinctly incorporated into this volume which is directed at research workers and undergraduate students. The contents are deliberately restricted to the physiology of the mother. Fertilization and parturition are excluded and so is foetal physiology except in its relation to maternal physiology.

There is a good bibliography, while 4 useful appendices deal with the place of Prochownick in the history of weight control, the sex ratio, multiple births and mean birth weight by geographic location.

E.P.W.

PREGNANCY NEPHROPATHY

Dr Sophian's views on the uterorenal reflex in the aetiology of pregnancy toxaemia, 'a disease of theories', and cortical necrosis are well known. In these two volumes he extensively details the physiology and pathology of the kidney in normal and abnormal pregnancy. He produces much old and new evidence to support his hypothesis.

These expensively produced volumes will be of particular interest to the serious student of this fascinating subject.

J.D.

PSYCHO-OBSTETRICS

This is the fifth in a series on modern perspectives in psychiatry. The book is divided into general and clinical parts. Of special interest are chapters on the Couvade syndrome; childbirth as a family experience; and termination of pregnancy. A cumulative index to previous volumes is provided.

This fascinating book emphasizes the profound influence of human reproduction on every facet of life. It is thoroughly recommended to obstetricians, paediatricians, psychiatrists and not least, family doctors.

H.A.v.C.d.G.