



CASE REPORT

ACCESSORY BREAST OUTSIDE MILK LINE

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Abstract

Polymastia or accessory breast tissue is an uncommon embryological defect and may present with or without nipple areolar complex. It is usually found along the milkline (most common being in axilla).Rarely ectopic breast tissue are found away from milkline and are called mammae erratae. Herein, we report a case of mammae erratae in the right loin in a woman and its management.

Keywords: Accessory breast, mammae erratae, milkline.

INTRODUCTION

The mammary glands are located on the anterior chest wall and are composed of mammary parenchyma with skin envelope containing the nipple-areola complex. During the early weeks (5th) of embryonic development, the mammary milk lines, which represent two ectodermal thickenings along the sides of the embryo, extend from the axillary region to the groin. In normal development, most of the embryologic mammary ridges resolve, except for two segments in the pectoral region, which later become the breast. A failure of any portion of the mammary ridge to involute may lead to ectopic breast tissue.^[1]

Ectopic breast tissue (EBT) is found in 0.4% - 6% of all women and 1-3% of males^[2], and occurs more commonly in Asian women^[3]. EBT is frequently located along the milk line, but can occasionally be detected in unusual locations, such as the face, posterior neck, shoulder, upper arm, flank, hip, posterior or lateral thigh, perineum, as well as midback^[2,3].

Herein, we report a case of an accessory breast tissue in the right loin in a woman and its management.



CASE REPORT

A 65yrs old female patient came to surgical op with complaints of swelling in right loin since birth. She observed an increase in size since 5yrs. The swelling had nipple areolar complex which was erectile. She had three children (P_3L_3), last child birth 30yrs back and all children were breast fed from normal breast. There was no lactation found from this swelling at the time of feeding. There was no history of pain associated with swelling. There were no associated urological symptoms. There was no history of breast cancer running in her family.

On examination, a single, non pigmented soft swelling of size 9*7 cm present over right loin with well formed erectile nipple areolar complex (Fig 1a, & 1b). Both breasts were normal each having normal nipple and areola. Left breast has an additional 1.5*1 cm non erectile pigmented nipple (Fig 2). There were no lymphnodes palpable in both axilla.



Figure 1 a.



Figure 1 b



Figure 2

The routine haematological and biochemical parameters are within normal limits. Ultrasound of swelling showed features suggesting breast tissue. Hence the swelling is clinically and radiologically diagnosed as ectopic breast tissue or polymastia not falling in milkline (a very rare presentation). The ectopic breast without nipple and areola are difficult to diagnose and the differential diagnosis of such polymastia are lipoma, hidradenitis, sebaceous cyst and that of additional nipple or polythelia is benign naevus.

The plan of management of this patient is excision. The excised swelling was found to extend upto deep fascia.



(Fig 3).

The histopathological examination showed breast fatty tissue with atrophy of lactiferous gland and ducts with no features of adenosis or malignancy (Fig 4)





DISCUSSION

Polymastia means the accessory breast glandular tissue with or without nipple & areola. Ectopic breast tissue may appear as anything from subcutaneous tissue similar in appearance to a small mole to that of a fully functioning breast. During the fifth week of gestation a thickening of ectoderm, known as the mammary ridge, appears on the ventral surface of the embryo. It correlates to a line from the axilla to the groin, called the milk line. The mammary ridge regresses the



(Fig 5)

following months, with the exception of paired tissue on the anterior chest, which forms normal, pectoral breasts. Failure of this regression may result in accessory breast tissue. Accessory breast tissue may occur at any point along the milk line most common being axilla^[4,5]. Very rarely, accessory breast tissue may occur outside of the milk line, a phenomenon known as mammae erratae. Cases have been reported on the buttocks, neck, face, arms, hips and back^[5].

Accessory breast has been classified by Kajava (1915) into 8 types as shown in table 1^[6].

TABLE 1 Classification of accessory breast (KAJAVA)

| Group | Type | Terminology | Description |
|---|------|---------------------------------------|---|
| Glandular tissue with all or part of the nipple-areolar complex | I | Polymastia | Complete breast: nipple, areola, glandular tissue |
| | II | Supernumerary breast (without areola) | Nipple and glandular tissue |
| | III | Supernumerary breast (without nipple) | Areola and glandular tissue |
| Glandular alone | IV | Ectopic breast tissue | Glandular tissue only |
| Part of nipple-areolar complex alone | V | Pseudomamma | Nipple and areola only; fat replaces glandular tissue |
| | VI | Polythelia | Nipple only |
| | VII | Polythelia areolaris | Areola only |
| | VIII | Polythelia pilosis | Patch of hair only |

Although Ectopic breast tissue is present at birth, it remains dormant and is often noticed only during puberty, pregnancy or lactation due to hormonal stimulation. As compared to pectoral breast tissue, ectopic breast tissue shows same hormonal effects and is prone to develop breast



diseases like abscesses, mastitis, milk fistula, cyclical mastalgia, fibroadenomas, fibrocystic disease, phyllodes tumor, Paget's disease & all types of breast cancer^[7].

Accessory breast cancer appears to have a poorer prognosis than cancer in pectoral breasts because the cancer metastasises to lymph nodes more frequently and rapidly than that in regular breast tissue^[8] or due to delayed diagnosis, attributable to diagnostic difficulties (especially relating to aberrant breast tissue), plus a lack of awareness in the medical community. This is supported by the finding that prognosis in accessory breast cancer does correlate to disease stage (which is often advanced when diagnosed). The management of malignancies in accessory breast tissue are treated in a similar manner to pectoral breast cancers.

Polythelia or additional nipple is linked with urinary track abnormalities like supernumerary kidneys, failure of urine formation, carcinoma of kidney^[4]. Nonrenal anomalies include cardiovascular, vertebral disease, pyloric stenosis, testicular cancer and even familial alcoholism.^[9]

Commonly, polymastia and polythelia occur sporadically, but familial cases are reported. Toumbis-Ioannou and Cohen describe a woman with left sided polythelia and an ectopic right kidney. Her older sister had left sided polythelia, and her brother had a complete supernumerary breast on his left side. Klinkerfuss reported on polymastia in four generations of a family^[4].

The treatment of choice for symptomatic accessory breast tissue is surgical excision. Cosmesis is the main indication in the majority of cases. Removal of the tissue will relieve the physical discomfort and also confirms the diagnosis. Liposuction is used as an alternative if feasible.

In 2011, the American Society of Plastic Surgeons published an algorithm for treatment of axillary accessory breast tissue (Table 2). It advocates a combination of surgical excision and liposuction, according to the features of the tissue^[10]. The commonly reported complications after removal of axillary breast tissue are incomplete removal of the accessory breast, poor scar, intercostobrachial nerve injury and lymph edema of arm^[1].

The highest number of supernumerary breast, reported by Neugebauer in 1886 was eight in addition to the two normal breasts. A report in 1675 described the presence of five nipples on the left breast and two on the right. There are numerous reports of infants nursed from supernumerary breast. An often cited case from 1827 refers to Therese ventre of Marseilles, France. Her mother had a supernumerary breast beneath her normally positioned right breast. Ventre had an accessory breast on side of left thigh. This breast enlarged during puberty and when she became pregnant, it produced milk. It was offered to her infant who took it willingly. She apparently nursed five children during her life from three of her breasts^[4].

**Table 2. Classification of Excess Axillary Breast Tissue and Management Algorithm**

| Type | Features on Examination | Management |
|------|---|--|
| I | Small palpable mass, barely visible on examination; little or no skin excess; feels distinct from normal breast, and/or has a firm central core | Direct excision with no skin removal |
| II | Small visible mass; little or no skin excess; feels or appears contiguous with normal breast, and has similar consistency | Suction lipectomy alone |
| III | Visible mass; excess skin present; contiguous with the normal breast, and of similar consistency | Suction lipectomy with skin excision |
| IV | Large mass; excess skin present; distinct from normal breast tissue, and/or has a firm central core | Direct excision of tissue and skin, with or without suction lipectomy to improve final contour |



CONCLUSIONS

Accessory breast tissue, an uncommon embryological defect, may present as inconspicuous polythelia or fully-formed polymastia which may be a diagnostic dilemma in the absence of areola & nipple. It is also vulnerable to the same diseases both benign and malignant as normal breast tissue. The ectopic tissue can be surgically excised if symptomatic or if it represents a cosmetic problem. Proper timing of surgical intervention is necessary to optimize the functional, psychological, and aesthetic outcomes.

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