ACCESSORY BREAST TISSUE:
BASIC INFORMATION FOR
GYNAECOLOGISTS

By
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Introduction

"Accessory Breast Tissue " is a topic of interest to the surgeons, radiologists and definitely to the Gynaecologists as well. Accessory breast tissue is defined as the residual breast tissue that persists from the normal embryologic development. It is also known as ectopic breast tissue. An ectopic breast tissue is a broad term that refers to both supernumerary breasts as well as the aberrant breast tissue. Supernumerary breasts are found along the “milk line,” that runs bilaterally from the anterior axillary folds to the inguinal region and medial thigh. “Aberrant breast tissue” is defined as “an island of breast tissue usually located close to the normal breast” and they consist of accessory fragments of breast tissue outside the periphery of the gland . In contrast to supernumerary breasts, aberrant breast tissue lacks an organized secretory system.
Incidence
Accessory breast tissue is found in 0.22 to 6% of the population. It is found to be the highest among Japanese population and is the lowest among the white population. Most cases are sporadic. It may have an autosomal dominant inheritance pattern with incomplete penetrance.

Location
Accessory breast tissue is often located in the axilla. It can also be found in locations outside the milk line, including the face, posterior neck, chest, middle back, buttock, vulva, flank, hip, shoulder, upper extremities, and posterior and lateral thigh have also been reported.

Embryology

During the intrauterine fetal development, a pair of mammary ridges are derived from the ectoderm. These ridges are referred to as "milk line". They run bilaterally along the ventral surface from the anterior axillary folds to the medial aspect of inguinal folds. These ridges regress normally except at the site of the breasts. This explains the accessory breast tissue development along the milk line but not for the accessory breast tissue found outside the milk line. Hughes, in 1950 proposed that certain migrating nests of primordial breast cells can develop randomly and later Craigmyle suggested that an accessory breast tissue can develop from modified apocrine sweat glands. An accessory breast tissue contains all the elements of the normal breast tissue such as the nipple, areola and the breast parenchyma.

The association of accessory breast tissue with urogenital anomalies range between 11 to 27%. More commonly seen are the supernumerary kidneys, renal cell carcinoma and failure of renal formation.
Classification

The Kajava classification system for supernumerary breast tissue was published in 1915. Class 1 is termed “polymastia” and consists of a complete breast with a nipple, areola, and glandular tissue. Class 2 is a supernumerary breast without an areola, consisting of glandular tissue and a nipple. Class 3 consists of an areola and glandular tissue. Class 4 is glandular tissue only. Class 5 contains a nipple and areola only and is termed “pseudomamma.” Class 6 consists of just a nipple, also known as polythelia, and class 7 consists of just an areola which is known as “polythelia areolaris”. The last one, class 8 is termed “polythelial pilosis” and consists of just a patch of hair. Class 4 is the most common clinical manifestation of accessory breast tissue, which is fibroglandular tissue in the axilla.

KAJAVA’S CLASSIFICATION

<table>
<thead>
<tr>
<th>TYPE OF ANOMALY</th>
<th>DESCRIPTION</th>
<th>TERMINOLOGY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class 1 (Complete)</td>
<td>Nipple + areola + glandular tissue</td>
<td>Polymastia (mammae erratica)</td>
</tr>
<tr>
<td>Class 2</td>
<td>Nipple + glandular tissue</td>
<td></td>
</tr>
<tr>
<td>Class 3</td>
<td>Areola + glandular tissue</td>
<td></td>
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<tr>
<td>Class 4</td>
<td>Glandular tissue</td>
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<tr>
<td>Class 5</td>
<td>Nipple + areola + fat tissue</td>
<td>Pseudomamma</td>
</tr>
<tr>
<td>Class 6</td>
<td>Nipple</td>
<td>Polythelia</td>
</tr>
<tr>
<td>Class 7</td>
<td>Areola</td>
<td>Polythelia areolaris</td>
</tr>
<tr>
<td>Class 8</td>
<td>Patch of hair</td>
<td>Polythelia pilosa</td>
</tr>
</tbody>
</table>

Clinical presentation

Most of them remain asymptomatic and are unaware of the presence of accessory breast tissue. The accessory breast tissue responds to hormones - estrogen, progesterone, prolactin and human placental lactogen. As a result of these hormonal changes, they are recognized by the majority of these patients only during menarchy, pregnancy or lactation. Affected individuals may undergo premenstrual changes such as tenderness, swelling, and difficulty with shoulder range of motion and irritation.
They usually present with complaints of masses which can be associated with discomfort, pain, milk secretion, local erythema which becomes more prominent during pregnancy and lactation. Always check the contralateral side when you examine for accessory breast tissue as they usually have a bilateral presentation.

**Differential diagnosis**

Some of the other conditions which could be confused with an accessory breast tissue are lipoma, sebaceous cyst, lymphadenopathy, vascular malformations and carcinoma. We also need to keep in mind that the pathological conditions that are seen in a normal breast like mastitis, fibroadenoma, fibrocystic changes, lobular hyperplasia and malignancy can also occur in an accessory breast tissue.

**Investigations**

**Mammogram**

It shows fibroglandular elements interspersed with fat like that of a normal glandular parenchyma but is located separately from the breast parenchyma. The axillary tail of Spence is a continuous extension of the main breast tissue into the axilla where as the accessory breast tissue is found discontinuous from the main breast parenchyma.

Mammogram shows patch of pliable fibroglandular tissue with interspersed fat discontinuous with majority of breast tissue more inferiorly, which is consistent with accessory breast tissue, and benign appearing axillary lymph nodes.

( *American Journal of Roentgenology* )

Asymmetry (circle) is seen on right, upper hemisphere, posterior plane on mediolateral oblique view.
Ultrasound
It is the first choice of imaging modality for palpable mass outside the breast when the patient is below 30 years. The appearance of accessory breast tissue is the same as that of normal breast with fibroductal tissue and fat lobules present.

Arrow indicates dermis, half bracket indicates retromammary layer, and bracket indicates accessory breast tissue between dermis and retromammary layer.

( American Journal of Roentgenology )

MRI
It is found to be most useful for the diagnosis of ectopic breast tissue in prepubertal and pubertal age group, who present with an axillary mass. On MRI, the accessory breast tissue appears similar to the normal breast parenchyma but it is discontinuous from the main breast tissue. It could be small, moderate or large. It can be symmetrical or asymmetrical.

Normal appearance of accessory breast tissue (bracket). Small to moderate amount of accessory breast tissue is seen in left axilla with mild enhancement.

( American Journal of Roentgenology )

FNAC
In case of any suspicious mass in the axilla or any other region or if the patient is symptomatic FNAC is done. FNAC can help to differentiate a fibroadenoma, lipoma, fibrocystic disease, lymphadenopathy or an
accessory breast tissue. FNAC remains an important tool in preventing unnecessary surgery in such cases. Hence, FNAC can be considered as the first diagnostic tool for palpable lesions.

Pathological conditions

Benign
All diseases of breast can also occur in an accessory breast tissue. Fat necrosis, Hamartomas, fibroadenomas and lactation adenomas are some benign conditions that can also occur in an accessory breast tissue.

Malignant
As of now the increased risk of malignancy in accessory breast tissue when compared to that of the pectoral breast tissue is still a controversy. The most commonly reported malignancy in accessory breast tissue is infiltrating ductal carcinoma (74%). There are also some rare reports of Paget disease, cystosarcoma phylloides, Leiomyosarcoma and invasive secretory carcinoma.

Accessory breast tissue malignancy can also occur concurrently with pectoral breast cancer. First case was reported in 1979 of a woman who presented with primary breast cancer and ectopic breast carcinoma in vulva.

Management
Majority of the patients with accessory breast tissue are not aware of the condition and are usually asymptomatic. They are usually identified during the routine surveillance of breast in high risk patients. As they are mostly asymptomatic, no treatment is usually required but the possibility of disease in accessory breast tissue should be considered. A patient with accessory breast tissue in pregnancy and her fetus will have an excellent overall prognosis with supportive treatment.

In case of a symptomatic patient with accessory breast tissue in the axilla, surgical excision of the mass is done to relieve the physical discomfort caused due to the large masses of accessory breast tissue. A surgical excision, liposuction or both can be done for axillary accessory breast tissue.

In case of an accessory breast tissue cancer some authors recommend radical mastectomy of the ipsilateral breast while others report radical mastectomy does not improve the prognosis of an ectopic breast tissue cancer. Others recommend the surgical procedure of choice for ectopic breast carcinoma is wide resection of the tumor with surrounding tissue, skin and lymph nodes. Post operative management is the same as that of breast cancer. Estrogen receptor/progesterone receptor and Her2/neu testing has to be done in an accessory breast tissue tumor just as in a case of breast cancer. Radiotherapy, Chemotherapy and hormonal treatments are given as per the recommendations followed for breast cancer.
**Follow up**
Patients with accessory breast tissue should be followed annually to be certain that no masses develop in the accessory breast tissue. If masses develop, a needle biopsy should be performed to rule out malignancy.

**Conclusion**
Accessory breast tissue, though being a rare congenital condition can be diagnosed by a thorough physical examination along with ultrasound and/or MRI. The accessory breast tissue is similar to the pectoral breast tissue. Hence it is prone to the same physiological and pathological changes as that of a pectoral breast tissue, which is more often exacerbated during pregnancy or lactation. As majority of these patients are asymptomatic, an accidental diagnosis of the accessory breast tissue often requires no intervention unless they are symptomatic or for cosmetic purpose. The clinician should always be prepared for the wide possibilities that may arise with accessory breast tissue, which could range from benign breast diseases to malignant breast cancers. FNAC helps to clinch the diagnosis in clinically suspicious cases.

**References**


