Pathologic breast conditions in childhood and adolescence: spectrum of US findings

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Learning objectives

Describe the different pathologic entities of the breast in pediatric and adolescent patients - and recognize their imaging findings.

Background

Discovery of breast masses in children and adolescents often causes tremendous parental and physician concern because of the high prevalence of breast cancer in the adult population.

Knowledge of the spectrum of pathologic conditions that affect the pediatric breast allows the radiologist to play an important role in providing an age-appropriate differential diagnosis.

The vast majority of conditions that cause breast masses or breast enlargement in children or adolescents are benign.

Imaging findings OR Procedure details

1. Embrology of breast development

- 6th week of fetal life: Beginning of breast development: the ectoderm on anterior body wall thickens into a ridge known as the "milk line", "milk ridge" or "hughes lines" (fig 1)

- Around 10th week: recession of the ridge above and below the area of pectoralis muscle leaving the mammary primordium, which is the origin of the lactiferous ducts.

- Between week 10 and 20: The initial ducts form and become interspersed through the developing mesenchyme, which becomes the fibrous and fatty portions of the breast.

- 34 weeks of gestation: The breast bud becomes palpable

- 8 months gestation: apparition of the nipple, initially as a depression and later as an elevation

2. Breast development

- Normal breast development is hormonally mediated:
- Adipose tissue and the lactiferous ducts grow in response to estrogen
- Progesterone stimulation results in lobular growth and alveolar budding

-Thelarche, or the onset of pubertal breast development, occurs between the ages 8 and 13, with an average of 11-11.5 years.

- The normal development of the breast occurs over a period of 2-4 years after thelarche and is classified by the Tanner system into five stages (fig 2)

- Menarche occurs approximately 2 years after the initiation of breast development.

- Lack of development by age 13 is considered delayed and warrants evaluation by an endocrinologist.

3. Evaluation of the pediatric breast

- Breast examination

- All pediatric and adolescent patients should have a breast examination at the time of their annual physical exam regardless of whether specific complaints are mentioned.

- For the adolescent patient, the patient is asked to lie supine with one arm over her head. The examiner should note Tanner staging of the breast.

- Self-breast examination may be taught to older adolescents as a mechanism to increase their understanding of the examination and put them at ease.

- Teenagers with history of malignancy or radiation to the chest, or a family history of BRCA1 or BRCA2 gene defects deserve early self-breast exam teaching as well as regular breast exams.

- Breast imaging study

- **Sonography**: the initial breast imaging study that performed in pediatric and adolescent patients (lack of ionizing radiation in a susceptible population and greater sensitivity in the relatively dense fibro-glandular tissue in young girls).

- **Mammography**: reserved for selected cases, role in evaluation of microcalcifications and suspicious of discrete breast masses in older adolescents;

- **MRI**: performed for breast masses that involve deeper structures or chest wall lesions.

- **Fine needle aspiration cytologic analysis and core needle biopsy**: rarely used (the extremely low prevalence of breast cancer in pediatric age group, much greater risk of intervention to the developing breast that to the mature breast.

- A conservative approach of clinical and sonographic follow up: more commonly adopted in children
4. Normal breast development: US characteristics

- Tanner stage I (prepubertal): in most cases US shows ill defined hyper echoic retroareolar tissue (fig 3a)

- Tanner stage II: it appears at US as a hyperechoic retroareolar nodule with a central star-shaped or linear hypoechoic area that represents mostly branched ducts

- Tanner stage III: hyperechoic glandular tissue is seen extending away from the retroareolar area, and a central spider-shaped hypoechoic region is noted (fig 3b)

- Tanner stage IV: (it is very transient and may not necessary appear) at US, in most cases, hyperechoic, mostly periareolar fibroglandular tissue is seen, showing a prominent hypoechoic nodule in the central region. Subcutaneous adipose tissue is identified in some cases (fig 3c-d)

- Tanner stage V: hyperechoic glandular tissue with increased subcutaneous adipose tissue anteriorly is found at US, without central nodule seen in tanner II, III and IV

5. Common development anomalies

5-1 Polythelia or supernumerary nipple: a common anomaly found usually along the milk line

5-2 Polymastia (more than two breasts): less frequently than polythelia

5-3 Accessory breast tissue: often found in the axilla

5-4 Amastia (congenital absence of the breast): rare, usually unilateral, may also result from trauma

5-5 Hypoplasia unilateral or bilateral of the breasts: more common than true amastia (small normal breast)

5-6 Breast asymmetry: usually of no consequence, substantial differences in breast size can result in a major cosmetic or self image problem and require sometimes a surgical option of augmentative and/or reduction mammoplasty.

A rare cause of breast asymmetry is Poland's syndrome: the absence or hypoplastic breast is associated with ipsilateral chest wall congenital anomalies of absence or hypoplasia of the pectoral muscle, latissimus dorsi, ribs 2-4 and vertebral bodies. (fig 4-5)

5-7 Premature thelarche: breast development beginning before the age of 7½ years, unilateral or bilateral, ultrasound finds normal breast tissue. Most cases are idiopathic without any other signs of puberty; ultrasound is useful in the early differentiation between isolated premature thelarche and central precocious puberty by allowing measurements of the uterus and ovaries (fig6).
5-8 **Gynecomastia**: physiologic pubertal gynecomastia (excessive development of the breast in the male) occur in as many as 75% of healthy boys and may caused by drugs; Sonography permits differenciation gynecomastia and adipomastia (adipose tissue accumulation in the breast caused cases of general obesity) from other breast masses.

Other causes: some pathologic conditions: Klinefelter syndrome, anarchism, acquired testicular failure, androgen receptor defects)

5-9 **Juvenile or virginal hypertrophy or gigantomastia** (an extreme form of gynecomastia): excessive female breast enlargement that occur in a relatively short period of weeks to months, often begins shortly after menarche or during pregnancy, is thought to be due to an abnormal local reaction to normal hormone levels.

6-**Inflammatory lesions**: can occur in the neonate and also children and adolescents;

Breast abcesses manifest as tender, indurated or fluctuant erythematosus masses, they may result from obstruction of a mammary duct, infection of a retro-areolar cyst, irritation or abrasion of a nipple, or cellulitis of the surrounding chest wall area. Staphylococcus aureus is the major causative agent in documented series. These lesions apper at US as cystic or complex masses. US is not only diagnostic but also helps guide therapeutic needle aspiration. Doppler US may recognize abcess from mastitis. (fig7).

7. **Benign neoplastic masses**

7.1 **Fibroadenoma**: is a benign fibroepithelial tumor and is the most commun mass in girls younger than 20 years of age, accounting for well over half of tumors in surgical series.

At US, fibro-adenoma manifest as a well-defined hypoechoic homogenous, round, oval or macrolobulated mass, 1-20 cm in diameter. Theses masses may appear almost anechoic with low-level internal echoes. Slender, fluid-filled clefts may be seen within juvenile fibro-adenoma. In rare cases the structure is heterogeneous(necrosis, dystrophic calcifications); Posterior acoustic transmission is variable and is usually enhanced or intermediate, but posterior shadowing has been described (infarction); In avoid lesions, the growth pattern is horizontal or parallel. (fig8)

During Doppler evaluation, these lesions may appear avascular or may demonstrate some central vascularity.

The **juvenile or cellular subtype of fibroadenoma** is an uncommon histologic variant of fibroadenoma that frequently undergoes markedly rapid growth. A fibroadenoma over 5-10 cm in diameter is termed a **geant fibroadenoma**. Although most giant fibroadenomas are of the cellular subtype, not all cellular fibroadenomas are geant.
The histopathologic and imaging features of the cellular subtype of fibroadenoma and phyllodes tumor (a fibroepithelial neoplasm that may be malignant) overlap considerably, such that they are indistinguishable at imaging. The finding of peripheral cysts at sonography suggests phyllodes tumor, but definitive diagnosis requires tissue sampling.

Management and treatment: the pediatric patients with typical clinical and sonographic imaging adenofibroma would be managed conservatively with clinical and sonographic follow up. Fine needle aspiration or core needle biopsy may be used for patients for whom confirmation of the diagnoses is desired. Surgical excision is indicated for symptomatic or rapidly growing masses (juvenile subtype)

7-2 Other benign neoplastic lesions: include hemangiomas, papillomas, hamartomas, lipomas, lymphangiomas etc (fig 9,10).

8- Non neoplastic benign lesions

8-1 breast cysts: are solitary or multiple, unassociated with fibrocystic disease, 1-5 cm in diameter and most commonly located near the nipple and areola (Retroareolar "Montgomery"cysts due to obstruction of the glands of "Montgomery" at the edge of the areola). Cysts can manifest as a palpable mass or as of secondary infection. Uninfected cysts appear as anechoic masses in the breast tissue. Uninfected cysts may be multiple in number, round or lobular in shape, contain internal septations or isolated echoes and be avascular at Doppler. When infected, the cysts may contain echogenic debris, septations or fluid-fluid levels and increased vascularity is noted peripherally (fig 11).

8-2 Fibrocystic changes and mammary dysplasia is a group of benign cystic, fibrosis and proliferative lesions with some potential for development of breast carcinoma that are most exclusively confined to adult breast, although they may be seen in late adolescence. US is nonspecific and may show solid or cystic masses, dilated ducts and echogenic foci representing fibrous tissue that may cause posterior sound attenuation. (fig 12)

8-3 Other benign lesions: include galactocele, post chirurgical fibrosis, fat necrosis, hematoma and extramedullary hematopoiesis

9- Juvenile papillomatosis: is a localized, proliferative disorder of young women and older adolescents. The mean patient age at diagnosis is 19 years. Patients presents with a firm, well defined, mobile mass in the periphery of the breast and without nipple discharge.

At gross examination, the resected mass appears well circumscribed and contains multiple small cysts (< 2cm) within a dense fibrous stroma, an appearance that have given rise to the term "swiss cheese disease". Yellowish calcifications are common. Tumors vary in size (1-8 cm). At histologic analysis, multiple macrocysts and dilated ducts are apparent within a fibrous stroma that blends into adjacent normal breast tissue.
The imaging appearance of juvenile papillomatosis is characteristic although not specific. At sonography, juvenile papillomatosis appears as an ill-defined mass with multiple small cysts, especially at the periphery, findings that reflect the gross pathologic features. Microcalcifications may be seen at sonography. Although results of mammography are usually negative, occasionally mammograms may reveal microcalcifications or asymmetric density. At MR imaging, juvenile papillomatosis has been described as a lobulated mass with small internal cysts (T2-weighted sequences and demonstrates marked enhancement with a benign enhancement profile.

Although juvenile papillomatosis is a benign condition, it is considered a marker for familial breast cancer.

10-Primary Malignant Tumors:

There is no clinical or radiologic difference between benign and malignant tumors, and histologic differentiation may be also difficult.

10-1 Phyllodes tumor or cystosarcoma phyllodes: is a rare fibroepithelial neoplasm that accounts for only 1% of breast lesions in children and adolescents, but it is the most common primary mammary malignancy in this age group. Phyllodes tumor shares many clinical, pathologic, and imaging features with juvenile fibroadenoma.

Phyllodes tumors demonstrate a wide spectrum of biologic behavior, and some have the potential for invasive growth, recurrence, or metastasis in rare cases. Most phyllodes tumors in adolescents are histologically benign.

Clinically, most patients present with a painless, rapidly growing, movable, rubbery mass, findings that simulate those of juvenile fibroadenoma. Most phyllodes tumors in children are larger than 6 cm at presentation.

The sonographic appearance of phyllodes tumor is similar to that of fibroadenoma. A well-circumscribed, round, or macrolobulated hypoechoic mass is identified, often with posterior acoustic enhancement. The internal echotexture is frequently heterogeneous; Anechoic cysts or clefts are very suggestive of this diagnosis but are not pathognomonic as they can also be seen in juvenile fibroadenoma. The imaging findings of benign and malignant tumors overlap significantly, and tissue sampling of suspect lesions is necessary for definitive diagnosis.

Most often, the prognosis for phyllodes tumor is favorable after complete surgical excision alone, but some of these tumors have the potential to recur or even metastasize.

Local disease and recurrence are treated with complete excision. Patients with malignant tumors should be closely monitored.

10-2 Carcinoma: Adenocarcinoma of the breast is extremely rare in children, accounting for less than 1% of breast masses in this age group, and may be less aggressive than in
adults. It usually manifests in the 1\textsuperscript{st} decade with a painless mass and the histopathologic results are similar to those of adult adenocarcinoma. The US appearance is variable and nonspecific; it manifests most commonly as a hypoechoic mass with inhomogeneous internal echoes, irregular margins, and variable acoustic shadowing.

10-3 Other primary malignant breast tumors: include lymphoma, rhabdomyosarcoma and angiosacoma (fig13)

11- Secondary malignant tumors:

Malignant breast lesion are more likely to be secondary to metastatic or disseminated tumor in children and are described in lymphoma, leukemia, rhabdomyosarcoma and neuroblastoma. Their appearance at US is nonspecific. In leukemia, they may manifest as solid, well-defined, and relatively hypoechoic mass that is sometimes bilateral. In metastatic neuroblastoma, US may reveal multiple hypoechoic breast lesions (fig 14)

Images for this section:
Fig. 1: Fig1: projection of the « milk lines"
Fig. 2: Tanner staging of breast development (Stage I to V)

Fig. 3: Tanner stages of normal pubertal breast development (a) at the tanner stage 1, sonogram a midly heterogeneous retroareol subcutaneous tissue anterior to the pectoralis muscle, (b) sonogram of Tanner stage 3 breast development in a 14 year-old girl demonstrated an echogenic glandular tissue with a retroareolar stellate, (c-d) sonogram of tanner stage 4 shows more echogenic fibroglandular tissue, note (d) the increased subcutaneous fat anterior to the glandular tissue
Fig. 4: Poland's syndrome in a 18months old girl, photography of the chest (a) shows anomalies of the right nipple areolar complex and the breast associated with ipsilateral chest congenital anomalies. Chest XR (b-c) shows hypoplasia of the right pectoral muscle and ribs2-4 resulting of deformity of the sternum. Ultrasound (d-e) was confirmed the asymmetry of the breast and chest wall related to hypoplasia of the right breast and the major chief of pectoral muscle.
**Fig. 5:** Another case of « Poland syndrome » in a young woman who had consult about asymmetry of breasts, chest XR demonstrates the hypoplasia of the right pectoral muscle, pectus excavatum and the asymmetry of the dense breast projection; sonography shows absence of the right pectoral muscle.

**Fig. 6:** Unilateral premature thelarche in a 6-years-old girl: sonography shows increased thickness of the left breast bud without any masses.
Fig. 7: Breast abscess in a 15-years old young girl who consult for mastalgia with local inflammatory signs: ultrasound demonstrates a septated fluid collection filled with echogenic material with a thick regular wall suggestive of a retroareolar abscess. Fine needle cytology with bacteriological investigation concluded staphylococcal abscess.
Fig. 8: Juvenile fibroadenoma in a 17-year-old adolescent referred for a breast mass, sonogram reveals a homogeneously hypoechoic, well-marginated mass with hyperechoic septations, posterior acoustic enhancement. Color Doppler image demonstrates internal vascularity.

Fig. 9: Cystic lymphangioma 15-year-old patient with subclavian tumefaction extended to the axilla, US exam and lung CT-scanner showing a cystic lesion involving the anterior thoracic wall and the left axilla.
Fig. 10: Hamartoma 18-years-old patient referred for mastodynia with palpable uncircumscribed nodule in the left lower quadrants union at the clinical examination. US exam shows a hypoechoic heterogeneous mass with liquid foci and pseudocasule, suggestive of an hamartoma. Lateral mammograms reveal an heterogeneous uncircumscribed mass with limits hidden by the normal gland of the same density located at the left lower quadrants union.
**Fig. 11:** Retroareolar cyst in a 14 years-old girl: ultrasound shows a small well-defined retroareolar anechoic small mass.

![Image](image1)

**Fig. 12:** Fibrocystic changes: in a 19-year-old adolescent who presented with unilateral nipple discharge, US scan demonstrates a large heterogeneous, ill-defined, retroareolar mass with duct ectasia which were well identified in fat saturation T2 weighted MR image.

![Image](image2)

**Fig. 13:** Angiosarcoma in a 16-years-old adolescent who presented a large mass and a reddish discoloration of the right breast skin. Ultrasound scan shows a voluminous hypervascularized heterogeneous well-defined mass in the right lower quadrants of 97×76 mm. Mammography reveals a mass occupying almost all the right breast area without any microcalcification in projection; MRI demonstrates the well-defined hypervascular heterogeneous tissular aspect of the mass, MRI showed
numerous little vessels running through the mass. Percutaneous US-guided microbiopsy suggested the diagnostic of breast angiosarcoma, confirmed histologically.

**Fig. 14:** Fig 14- breast Metatasis of ganglioneuroma in a 3 years- old girl who referred for a breast mass, CT scans demonstrate multiple tissular masses (right breast [*], retroperitoneum[*], subpleural space [*])
Conclusion

Pathologic breast conditions are rare in childhood and adolescence. The spectrum of breast disease in this age group is different from that in adults; most lesions are benign and represent either normal breast tissue, cysts, or fibroadenomas.

US can help evaluate breast masses in children. It is the ideal imaging modality to study the pediatric breast and can be useful in all cases in identifying and characterizing the abnormality and guiding further investigation. Knowledge of the US appearance of normal breast development and specific lesions is essential for successful use of breast US.

Personal Information

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