A Rare Case of 12 Year Old Girl with Giant Fibroadenoma with Phylloides Tumour showing Features of Intraductal Papilloma

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Abstract
Breast tumours in prepubertal females are usually rare. Most commonly they present as adult type Fibroadenoma (FA). We report a rare case of a 12 year old girl who presented to our side with a painless progressive breast lump (Left side) since 5 months. The final histopathological diagnosis made was that of a giant fibroadenoma with phylloides tumour showing features of intraductal papilloma.

Introduction
Breast tumors most often seen in children and adolescents are fibroadenoma (FA), which comprises 44–70% of all breast lesions\(^1\),\(^2\). Although adult-type FA accounts for the large majority 93%\(^3\) of FA cases in childhood, a small proportion of fibroepithelial tumors clinically show rapid growth, often giving rise to large-sized mass lesions, and pathologically exhibit somewhat different histology, including cellular stroma and epithelial hyperplasia, which is often florid. These tumors are called giant fibroadenoma\(^3\).

Phylloides tumor (PT) is another fibroepithelial tumor, rarely seen (~1%) in children\(^1\),\(^2\),\(^4\). PT is distinguished from FA by higher degree of stromal cell growth and peculiar intracanalicular growth pattern, producing characteristic leaf-like structures with very broad cellular fibrous stroma.

PT is classified into three categories, namely, benign, borderline and malignant, depending on the degree of stromal hypercellularity, cytological atypia and mitoses, stromal overgrowth, and nature of tumor borders/margins\(^5\).

The case that we are reporting showed extremely unusual intraductal growth and prominent epithelial hyperplasia, showing unusual similarities with intraductal papilloma (IDP). Only a handful of such cases showing a link between FA, PT and IDP that in a 12 year old girl have been reported in literature.

Case Report
A 12-year-old female visited our hospital with a 5 month history of a painless progressive left sided breast lump. She had no particular past history and
family history for breast cancer and her general health condition was good. Patient attained menarche at 11 years. Physical examination confirmed a well-defined 13x13 cm mobile mass occupying the upper and lower outer quadrants of the left breast. It was firm to hard, irregular mass, not fixed to the underlying structures. No clinically palpable axillary lymphnodes were identified. There was no other significant systemic illness. All hematological and biochemical investigations were within normal limits. (Fig.1)

USG mammography: showed a well circumscribed 13*12*6cm hypoechoic heterogenous mass lesion containing many cystic spaces with variable posterior acoustic enhancement and showing mild to moderate internal vascularity. No obvious foci of calcification seen. Other breast and bilateral axilla within normal limits. BIRADS 4 grade. (Fig.2)

FNAC: mucinous blood mixed material aspirated. Smears prepared show irregular clusters, acini & tubules of mildly atypical epithelial cells with mild anisokaryosis and anisocytosis. Also seen large clumps of fibrocollagenised stromal cells. In the background lakes of mucin seen. Impression: Suggestive of atypical neoplastic lesion of breast.

D/D- Giant fibroadenoma with atypia

Based on the cytological and clinical findings a preliminary diagnosis of benign proliferative breast lesion closest to giant fibroadenoma was offered. However, a cytological possibility of benign phylloides tumor was not ruled out and thus the patient was taken up for surgery. Wide local excision of the tumor sparing the nipple areola complex was performed. The post operative period was uneventful and patient was discharged on 7th postoperative day. The patient has remained asymptomatic for past one month of followup. (Fig.3)

**Results**

Gross examination a lobulated mass 13.5*13.0*6.5 cm visualised. Leafy protrusion seen on external surface with one small lobe & other larger attached at middle cut surface mucoid sticky with lobated grayish to whitish. (Fig.4)

Microscopy: Multiple sections showing well circumscribed mass comprising of marked ductal and stromal hyperplasia. Ductal glands are dilated to large cystically dilated lined by tall columnar to pseudo stratified columnar mucus secreting cells and lumen filled with eosinophilic material. The glands are lined by hyperplastic myoepithelial cell layer. The stromal hyperplasia comprised of phylloides proliferation of fibroblastic cells with increased cellularity and mild atypia. Focal myxoid areas are also seen. (Fig.5-10)

Impression: Suggestive of benign neoplasia- Giant fibroadenoma with Cystosarcoma phylloides with features suggestive of Intraductal papilloma
FIG. 2: USG MAMMOGRAPHIC APPEARANCE OF THE TUMOUR

FIG. 3: INTRAOP PHOTOGRAPH OF THE RESECTED TUMOUR

FIG. 4: GROSS SPECIMEN OF THE TUMOUR
Discussion
Breast fibroadenomas are the most common solid lesions found in young women. They typically present as firm, mobile, painless, easily palpable breast nodules. Juvenile or Giant fibroadenoma is an uncommon pathology usually presenting in adolescents, characterized by massive and rapid enlargement of an encapsulated mass. Giant fibroadenomas which are approximately 4% of all fibroadenomas present as rapidly growing unilateral macromastia without definable borders or texture differences. However, full evaluation may reveal that the larger breast contains the abnormality with hypoplastic breast on the smaller side. Giant Fibroadenomas can be at times difficult to distinguish from phylloides tumor.
PT can occur in patients of all ages with a peak incidence in the fourth and fifth decade of life. The rarity of the malignant tumors of breast in adolescents does not exclude such possibility as about 2% of all primary malignant breast lesions occur under the age of 25 years in the females. However, it needs a careful diagnostic and clinical approach to rule out the possibility of malignancy. Cytological diagnosis of PT remains difficult with a significant overlap with fibroadenomas. The cytological smears of malignant PT is quite easy and well established but the differential diagnosis between fibroadenomas and benign or borderline phyllodes tumor is overlapping at times. The presence of large tumor size, low epithelial stromal ratio, epithelial atypical, columnar stromal cells with visible cytoplasm and stromal giant cells favors a diagnosis of PT over fibroadenomas.

In fibro epithelial tumors, including FA and PT, cyst formation within a tumor may be occasionally seen, but intraductal tumor growth is extremely rare, with only a small number of such cases having been described in the literature and its mechanism is unknown. It has been reported that PT arises from the stroma around the interlobular ducts rather than intralobular stroma, which may explain the histogenesis of intraductal papilloma.

Surgical resection of the tumour sparing the nipple areola complex was performed in this case. To conclude, giant fibroadenoma of the breast with phyllodes tumour showing unusual features of intraductal papilloma in a young female child is a rare occurrence which warrants reporting. However, the number of patients reported is too small, and so additional cases need to be analyzed before any conclusion is made.

References