Mammary Pseudoangiomatous Stromal Hyperplasia: A Mimic of Angiosarcoma

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Abstract

Pseudoangiomatous stromal hyperplasia (PASH) is a rare, benign hamartomatous lesion of mammary stroma with uncertain incidence as only case-reports have been reported in the literature. It is characterized by slit-like anastomosing channels lined by myofibroblastic cells. We report a case of PASH in a thirty year old lady who underwent lumpectomy for fibroadenoma.

Conclusion: Clinical and diagnostic significance of PASH is its potential confusion with low grade Angiosarcoma.

Introduction

Viuitch et al in 1986 first described PASH in mammary stroma. PASH resembles normal physiological changes in mammary stroma during luteal phase suggesting that this lesion represents a response to progesterone in oestrogen primed tissue. PASH can be seen as an incidental finding in fibroadenoma in female breast and with gynaecomastia in male breast. Extramammary sites include perianal area, perineum, labia majora and ectopic breast.

Case Report

A 30 year old lady had painless, movable, palpable mass in right breast since eighteen years. She had two children, younger is ten years old. She had no axillary lymph nodes and her systemic examination was unremarkable. She underwent lumpectomy for clinical diagnosis of fibroadenoma breast.

We received a well encapsulated mass measuring 10 x 8 x 5 cm, soft to firm in consistency with a smooth outer surface. Cut section was grey opaque with homogeneous appearance and a few slits (Fig. 1). No areas of necrosis or haemorrhage were seen.

Histological examination showed a well circumscribed mass composed of extensively collagenised intralobar and interlobar stroma with anastomosing slit-like spaces (Fig. 2) lined by myofibroblastic cells showing no Atypia (Fig. 3). Ducts were moreover attenuated while some of them were dilated and contained secretion. True capillaries filled with RBCs were seen. There was no evidence of malignancy in the sections studied. A diagnosis of benign mesenchymal neoplasm suggestive of PASH was rendered.

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Fig. 1: Gross photograph illustrating a well encapsulated mass measuring 10 x 8 x 5 cm with a smooth outer surface. Cut section was grey opaque with homogeneous appearance with a few slits.
Discussion

Regarding pathogenesis, Vogel et al.\textsuperscript{1,4} have described distinct alterations in mammary stroma during menstrual phase. The stroma is traditionally divided into hormonally responsive intralobular compartment and an hormonally unresponsive interlobar compartment. “Loose broken” stroma is seen in secretory phase. Lobulocentric pattern seen in PASH is similar to luteal phase, suggesting that it could be the result of expansion of intralobular stroma. Finding of distorted acini, absence of ducts, effacement of inter and intralobular distinction support the concept of lobular expansion. Also possible is the recruitment of less sensitive interlobar stroma in PASH.\textsuperscript{1,4} PASH is commonly seen in pre-menopausal women with a wide age range presenting as palpable mass commonly diagnosed as fibroadenoma.\textsuperscript{1} Our case presented at 30 years of age with palpable lump for 18 long years. Size of lump may vary from 2-15 cms with an average of 5 cm. Cut section is homogeneious or fibrous, tan grey to white with small cysts. Histologically, most striking finding is complex pattern of slit like spaces lined by plump myofibroblastic cells separated by crackled collagenous stroma. True capillaries with RBCs may be seen.\textsuperscript{1-5} Presenting case had similar morphological features. Histological forms of PASH include classic, tumourous and fascicular forms. Differential diagnosis varies with cellularity of lesion. Classic PASH closely resembles low-grade Angiosarcoma.\textsuperscript{1-6} Factors helpful in distinguishing PASH include narrow slits with its peculiar lobulocentric distribution which merges with interlobar stroma rather than infiltrate and destroying it as seen in Angiosarcoma which has more aggressive, infiltrative pattern. The lining cells of anastomosing spaces in PASH are lined by bland myofibroblastic cells which lack atypia, whereas in Angiosarcoma, lining cells show marked atypia as well as presence of mitotic figures. IHC markers in Angiosarcoma are positive for factor VIII and other vascular markers.

Tumourous form of PASH can be confused with entities associated with increased cellularity such as cystosarcoma phylloides. Fascicular growth, lack of stromal pleomorphism and mitoses despite increased cellularity, absence of stromal overgrowth and foci of classic PASH are distinguishing features.\textsuperscript{4}

Fascicular PASH should be differentiated
from Myofibroblastoma which has homogeneous but organized densely cellular pattern divided by collagenous bands. Wide local excision is treatment of choice, although PASH is known to recur.1 Diffuse PASH and recurrent cases may necessitate mastectomy.1,4 Our case is well till date, after one year of follow-up of lumpectomy.5

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References

CHRONIC SUBDURAL HAEMATOMA – TO DRAIN OR NOT TO DRAIN?
Thomas Santarius an colleagues report that a subdural drain reduced recurrences and mortality after burr-hole evacuation of chronic subdural haematomas (CSDH). This condition is common, dangerous, and treacherous, and mostly occurs in elderly individuals. CSDH is a capsulated haemorrhagic fluid collection that within a few weeks can grow large enough to cause progressive cognitive and neurological decline. Once diagnosed by CT or MRI, the treatment (virtually without an upper age limit) is rinsing of the subdural fluid space, usually through one or two burr holes.

Because of old age and fragility of most patients with the condition, however, an overall complication rate of 21% has been reported.

Recurrences are a major problem and require re-rinsing of the subdural fluid space, sometimes repeatedly. To prevent recurrences, some neurosurgeons place a subdural drain for a day or two; others do not for fear of puncturing the cortex and causing an intracerebral or subdural haematoma, or for fear of provoking a bacterial subdural empyema.

First, we rinse virtually all patients with a primary CSDH under local anaesthesia and sedation, making one burr hole only, compared with the two burr holes under general anaesthesia at Addenbrooke’s. Our rationale is that we feel that many patients with CSDH are too old and fragile for general anaesthesia.

In conclusion, drain or no drain has called for a randomized clinical trial for decades and here it is, impeccably done. The use of a drain drastically reduced recurrences, management, and mortality.