

## Angiosarcoma of the breast complicating pregnancy

Gurjeet Kaur MD, MMed Path.

Department of Pathology, University Sains Malaysia, Kelantan.

### Abstract

A 32-year-old pregnant lady presented with a rapidly enlarging right breast mass. A **fine** needle aspiration was suggestive of a malignant phylloides tumour. However histopathological examination after mastectomy revealed a moderately differentiated angiosarcoma. The histopathological and cytological features of this rare tumour together with the diagnostic pitfalls are discussed.

**Key words** : Angiosarcoma, breast, pregnancy.

### INTRODUCTION

Angiosarcoma of the breast is an uncommon but distinctive clinical and pathological entity. The earliest report of 'angiosarcoma' of the breast was in 1887.<sup>1</sup> This **tumour** accounts for approximately 1 out of 1700 to 2000 primary malignant tumours of the **breast**.<sup>2</sup> Up to 1995, 197 cases have been reported in the world literature<sup>3</sup> and several cases have been reported in pregnant **women**.<sup>4-8</sup> This is the first reported case in Malaysia in the past 15 years. In Hospital Kota **Bharu**, 73 cases of breast malignancies were diagnosed from 1994 until 1997. Of these, 70 were breast carcinomas. The other three consisted of malignant phylloides tumour, **non-Hodgkin's** lymphoma and this case of angiosarcoma.

### CASE REPORT

A 32-year-old lady **first** presented to the surgical outpatient department during the sixth month of her pregnancy. She complained of a rapidly enlarging right breast since the onset of her pregnancy, associated with some discomfort. On **examination**, the right breast was three times the size of the left breast. It was **diffusely** soft and not fixed to the underlying structures. The overlying skin was bluish-red and oedematous. The nipple was normal. The **axillary** lymph nodes were not palpable. A clinical diagnosis of phylloides tumour was made.

A fine needle aspiration of the right breast yielded blood. The smears were poorly cellular with fragments of spindle-shaped cells displaying nuclear **atypia** and occasional mitotic figures. Epithelial cells were absent. The aspirates were reported as consistent with phylloides tumour with probable malignant change (Fig. 1A and B).

A mastectomy was performed at 32 weeks of gestation. The tumour occupied the whole of the right breast, was attached to the pectoral fascia and in areas to the muscle. There were no post-operative complications.

The patient delivered spontaneously at 38 weeks of gestation. She was referred to the **oncology** unit in a nearby referral centre but has since been lost to follow-up.

### Pathological findings

The mastectomy specimen weighed 2.6 kg. The tumour occupied the whole breast and measured **19x17x10** cm. The tumour was diffusely haemorrhagic and spongy with **large** pools of blood. There were two small superficial ulcers over the skin. The nipple appeared normal. The nearest surgical margin was the posterior margin which was 1 mm away from the tumour. No lymph nodes were retrieved (Fig. 2).

Microscopical examination revealed the tumour to be composed of anastomosing irregular vascular channels, lined by plump endothelial cells with moderate cytological atypia and occasional mitotic figures. Focal intraluminal papillary projections lined by piled-up endothelial cells were present. The delicate supporting stroma contained similar 'endothelial cells'. Glandular elements and **fibroblastic** stroma were not seen. Haemorrhage and necrosis was confluent and extensive throughout the tumour. The tumour extended up to 1mm from the posterior surgical margin (Fig. 3).

### DISCUSSION

Older terms employed to describe primary **angiosarcoma** of the breast include angioblastoma,"

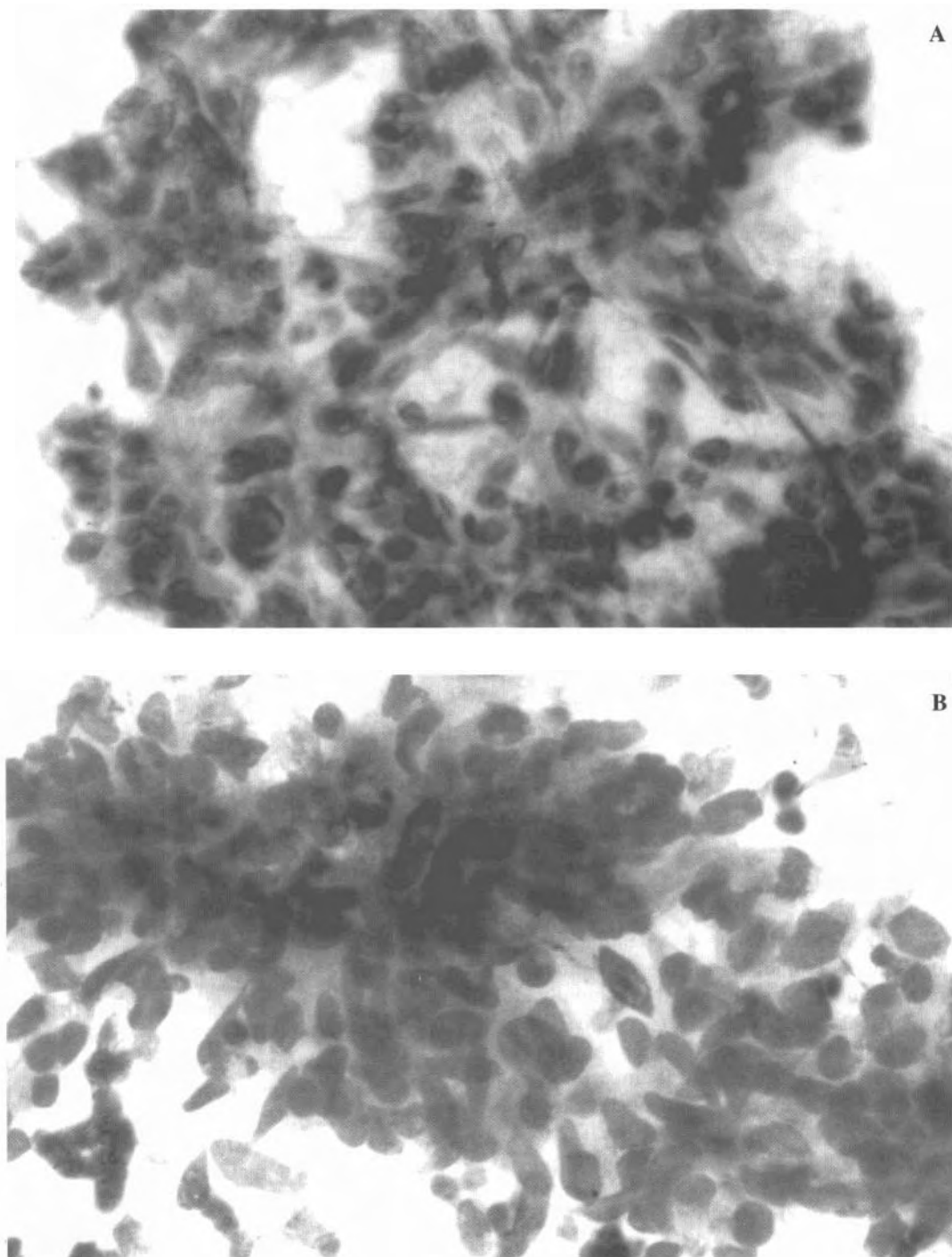


FIG. 1A&B: Atypical spindle cells on fine needle aspiration; A: PAP stain x 100. B: MGG stain x 100.

malignant haemangioendothelioma<sup>5,6</sup> and hemangiosarcoma.<sup>9</sup> It occurs exclusively in women<sup>2</sup> although a male patient with histologically proven primary angiosarcoma of the breast has been reported.<sup>10</sup> The incidence peaks in the third

to fourth decades of life. Thus far there have been only five published reports of this lesion occurring in pregnancy.<sup>4-8</sup> Although angiosarcoma is a rapidly growing tumour, there is a dearth of data as to whether the rate of growth is sub-

stantially different between pregnancy and non-pregnant patients. Immunostaining for oestrogen receptors in this case was negative but this does not exclude the possibility that the tumour is hormone dependent.

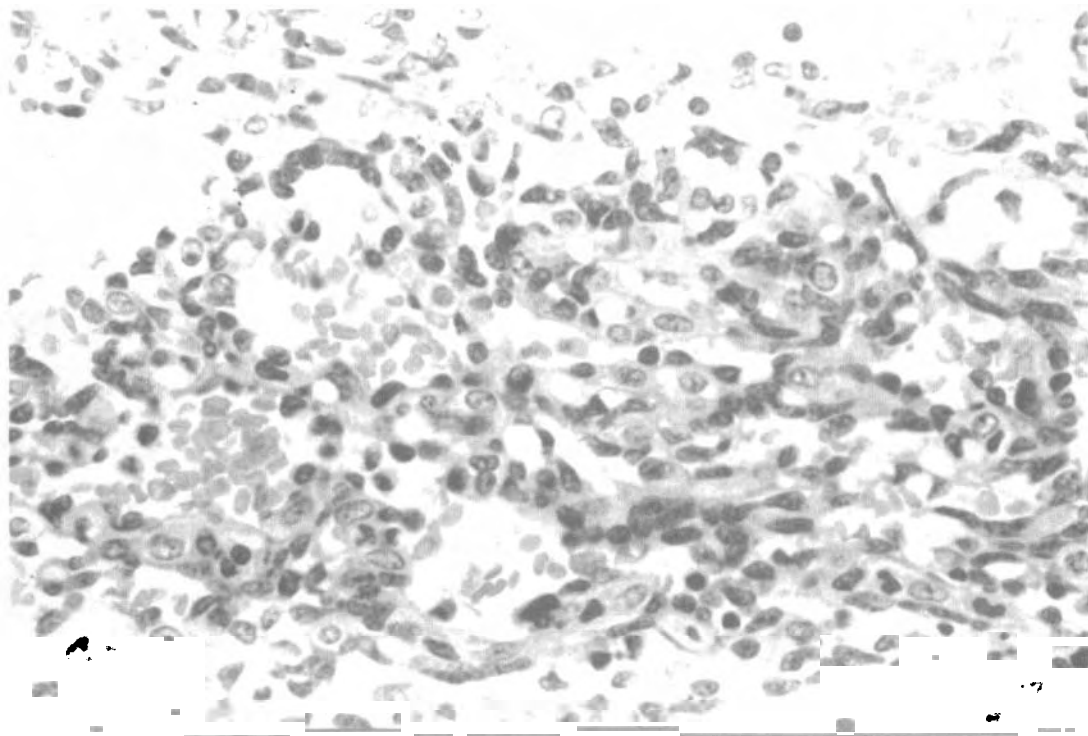
This patient demonstrated the classical presentation of a rapidly enlarging, painless breast mass,

associated with blue-red discolouration of the skin. Common signs of breast carcinoma such as skin retraction, nipple discharge, and axillary node enlargement were absent.<sup>2,10</sup>

The fine needle aspiration cytology findings of numerous red cells and few atypical spindle-shaped tumour cells are reportedly quite typical



**FIG. 2 :** Gross appearance of a diffuse, haemorrhagic breast mass.



**FIG. 3 :** Irregularly anastomosing vascular channels lined by atypical endothelial cells. H&E x 100.

of **angiosarcoma**.<sup>11</sup> However, there is considerable overlap with phylloides tumour which makes differentiating the two tumours with any degree of certainty very difficult. Some features that are common to both lesions include the relative **sparsity** of an epithelial component, as well as the presence of spindle-shaped cells which may show nuclear **atypia**, pleomorphism and mitotic figures.

The macroscopic findings of a haemorrhagic, spongy mass is characteristic of an angiosarcoma. The size of the tumour in our case was large compared to **5cm** which was the mean size reported in a study of 20 patients with **angiosarcoma**,<sup>10,12</sup> although they ranged from 1 cm to 14 cm in **diameter**.<sup>10</sup>

Histologically, angiosarcomas show a variable appearance, even within the same tumour. Basically it is composed of anastomosing irregular vascular channels lined by atypical endothelial cells. In some areas the tumours may be highly cellular while in others they consist only of **bland**-appearing infiltrative vascular elements that resemble benign capillary haemangiomas. The latter not infrequently results in histological **misdiagnosis** in biopsy **specimens**.<sup>10,13</sup> Donnell *et al*<sup>14</sup> stratified angiosarcomas into three categories grouped by histological appearance, correlating with tumours that are well differentiated (group I), moderately differentiated (group II) and poorly differentiated (group III). The present case corresponded to group **II** or the moderately differentiated category.

Angiosarcoma is the most aggressive and lethal of all breast neoplasms. The majority of patients die within two years of **diagnosis**.<sup>2</sup> Axillary nodal status has a minimal impact on prognosis. The best standard treatment of **angiosarcoma** is early and complete surgical excision of the mass with adequate negative margins. Axillary node dissection is not **indicated**.<sup>3,12,13</sup> The role of adjuvant therapy (chemotherapy **and/or** radiotherapy) is still not clear.

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