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Case Report

The diagnostic and treatment challenges of a primary breast angiosarcoma: a rare case report

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ABSTRACT

Primary angiosarcoma of the breast (PAB) is an aggressive malignancy that represents less than 0.05% of breast malignancies. PABs can pose a diagnostic challenge as they present in the absence of risk factors and in younger patients where breast density weakens the diagnostic utility of imaging. Treatment decisions may also be challenging as there remains scant consensus on the most appropriate mix of surgical and adjuvant treatment modalities. We report the case of a 40-year-old female who presented with a 1-centimeter peri-areolar nodule. After initial suspicion for infection and failure of anti-microbial therapy, a shave biopsy was performed. Angiosarcoma was subsequently confirmed and further wide local excision demonstrated no residual disease, likewise imaging was also not concerning for metastases. Given reassuring clinical progress, consensus among the multidisciplinary team was for surveillance without adjuvant therapy. The case was fortunate to have early cutaneous manifestation, and thus lead to earlier presentation, intervention and a positive outcome. Nonetheless it demonstrates the potential challenges to diagnosis in young patients who present with an absence of risk factors, and the case-by-case deliberation regarding management. Clinicians should remain vigilant of atypical, indeterminate breast lesions and ensure progression to more sensitive imaging and prompt biopsy to prevent missed or delayed diagnoses. A diagnosis of primary breast angiosarcoma should have early expert involvement, aggressive intervention and thorough follow up to optimise patient outcome.

Keywords: Angiosarcoma, Primary breast angiosarcoma, Breast cancer, Breast surgery

INTRODUCTION

Angiosarcomas of the breast are often overlooked rare and aggressive endovascular lesions which occur most commonly as secondary lesions following radiation or chronic lymphoedema.[1,2] Rarely angiosarcoma of the breast occurs as a primary malignancy in the absence of these risk factors. Primary angiosarcomas of the breast (PAB) represent 0.04% of malignant breast lesions and around 20% of breast angiosarcomas.[3]

PABs present clinically as parenchymal masses which are often rapidly growing and occur most commonly in patients aged 30 to 50.[4] The increased density of breast tissue in this younger cohort makes mammography diagnostically difficult, with up to one third of cases presenting with a normal mammogram.[5] Hence, diagnosis is typically made histologically.[5] It is an aggressive malignancy with high metastatic potential, typically doing so via local invasion or hematogenous spread, rarely involving lymph nodes.[1,2] PABs carry a relatively poor prognosis with overall three-year survival rates for PABs being 23% compared to 60% for non-angiosarcoma breast sarcomas.[6]

Treatment is typically aggressive management with mastectomy due to a high rate of local recurrence with breast conserving procedures.[7] Given the rarity of PABs and limited data with conflicting outcomes the role of adjuvant chemotherapy and radiation remains unclear, and

its use is on a case-by-case basis.[8-10] The aggressive nature of these tumours coupled with their relatively early onset in the absence of clear risk factors raise both diagnostic and treatment challenges for patients.

CASE REPORT

A 40-year-old female was seen by the breast surgery department at a local hospital following general practitioner (GP) referral for an isolated 1 cm right peri-areola nodule biopsy confirmed to be an angiosarcoma. The lesion appearing a month prior, was initially misdiagnosed and treated as an infection.

Surgical review demonstrated a lack of causative factors including previous radiation, lymphoedema or prior surgery, confirming a primary breast angiosarcoma diagnosis. Furthermore, she had minimal other risk factors, experiencing menarche at age 14 years, 2 children and pregnancies, non-smoker and no history of hormone replacement therapy. She had no confirmed family history of breast cancer, and breast related surgical history only included a previous cyst aspiration, and regular ultrasound (US) follow up since then. The lesion having been shaved off for biopsy by GP was no longer evident in surgical clinic. Her breast exam was hence unremarkable for all but a 2 cm lipoma. Extensive diagnostic workup was performed after re-affirming histological diagnosis.

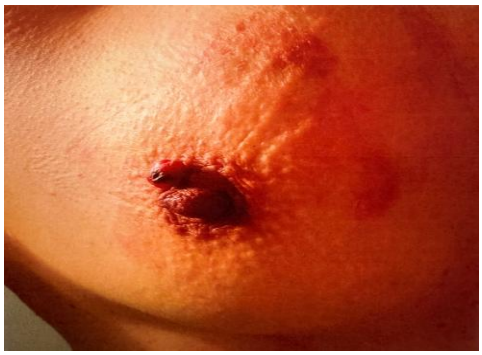

Figure 1: Photograph by GP on initial presentation demonstrating erythematous right periareola nodule with central black discolouration.

Histopathological examination of hematoxylin and eosin (H&E) stained sections demonstrated a nodular lesion comprising numerous small to medium-sized, blood-filled vascular channels exhibiting atypical features. More pronounced endothelial cell cytological atypia accompanied by conspicuous mitotic activity evident on higher magnification indicated a vascular malignancy (Figure 2). Subsequently, Immunohistochemistry confirmed endothelial/vascular origin of the lesion with strongly positive vascular markers CD31 and ERG. Ki-67 proliferation index was more than 50% positive, further confirming malignant nature of the vascular lesion. C-MYC expression was also upregulated within the atypical vascular component, although typically associated with secondary angiosarcomas, is also not uncommonly positive in primary tumours (Figures 3a-d).

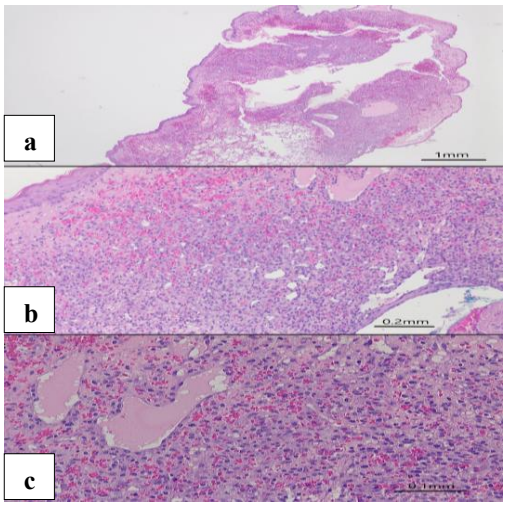

Figure 2 (a-c): Hematoxylin and eosin (H&E) sections of low, medium and high magnification respectively. The lesion forms a nodule comprising numerous small and medium-sized blood-filled vascular channels with atypical features. The high magnification image shows endothelial cytological atypia and significant mitotic activity. All findings expected of a vascular lesion such as an angiosarcoma.

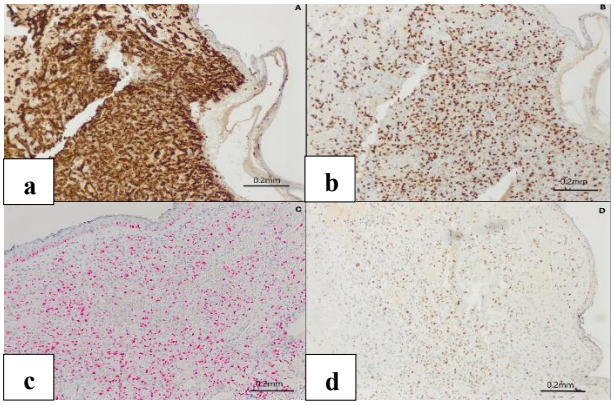

Figure 3: Immunohistochemistry staining with positive CD31 and ERG markers displayed (a and b) respectively highlight vascular lesion as expected with angiosarcoma, (c) shows Ki-67 which is markedly elevated and highlights high proliferation index, and (d) shows a positive C-MYC stain, which is a non-specific marker for secondary tumours.

Following histological confirmation, computer tomography (CT) - chest, abdomen and pelvis, bilateral mammogram and ultrasound of the breasts, magnetic resonance imaging (MRI) breasts as well as an fludeoxyglucose positron emission tomography (FDG PET) were performed. All investigations were unremarkable, with nil evidence of local, nodal or metastatic disease. Her PET scan showed a non-specific non FDG avid 6 mm subpleural nodule.

Three further punch biopsies around the initial focus were also all benign. The patient's care was subsequently transferred to a tertiary facility with a specialised sarcoma unit, where the decision was made to perform a nipple sparing wide local excision, removing surrounding areola and a cuff of skin. The histology of this was also benign, showing no residual angiosarcoma. Finally, after case discussion at a multidisciplinary meeting, the consensus was reached that this was a low risk angiosarcoma, detected early, and would therefore not require further therapy. The decision was reached to maintain close surveillance of the patient 4-monthly for 2 years, including CT chest to monitor the subpleural nodule.

DISCUSSION

PABs present both diagnostic and treatment challenges for clinicians. They may be easily missed with common screening methods such as ultrasound and mammogram.[11] The neoplasm has a high metastatic potential and three-year survival rate as low as 23%, making missed diagnosis and delayed treatment potentially fatal.[6] We present a fortunate and atypical case where a suspicious peri-areolar nodule and appropriate early action by the primary physician and surgeons triggered rapid diagnosis and intervention, culminating in a positive outcome. The presentation with an early visible cutaneous lesion is atypical, most cases of PAB present late with a palpable mass and may lead to delayed diagnosis.[1] Further contributing to potential delayed diagnosis is the relatively reduced utility of ultrasound and mammography in a younger cohort with increased breast density.[5]

With no clear consensus on management, each patient is typically assessed on a case-by-case basis with expert opinion tending to lean towards an aggressive stance with surgical excision or mastectomy depending on the extent of the tumour.[7] Breast conservative surgery versus mastectomy, as well as the role of radiotherapy and chemotherapy remain unclear.[6,12] Among the limited literature surrounding the topic there is also significant emphasis on high recurrence rate, with up to 23% recurrence with wide local excision.[2]

PAB is a rare diagnosis and justifiably has limited surrounding literature and no definitive consensus regarding its aetiology, pathogenesis or treatment. However, there appears to be agreement among the research sphere regarding its challenging diagnosis, high metastatic potential, recurrence rate as well as mortality.[1,2,6,7,10-12]

Hence, despite most surgeons and primary physicians being unlikely to encounter PAB, it should be among the list of differential diagnoses on initial workup of any breast mass, not overlooking patients who may not have any of the typical breast cancer risk factors. Any mass that may yield inconclusive or unclear findings on mammogram or US should proceed immediately to biopsy and MRI, especially if rapidly growing. Histologically confirmed PAB should have early expert involvement and aggressive management and thorough follow up.

CONCLUSION

PAB is a rare diagnosis that may often be overlooked, and any diagnostic delay at potentially significant detriment to the patient, given its high propensity for metastasis and poor prognosis. Therefore, despite being low on the list of differentials, it should be considered nonetheless when evaluating any rapidly growing breast mass, especially in young patients with no significant breast cancer risk factors. Atypical masses that yield inconclusive results on common screening methods such as mammogram and US should progress to obtaining MRI imaging, and early histological diagnosis. Subsequent aggressive surgical management and thorough outpatient follow up is also pivotal in achieving a favorable patient outcome.

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