Cellular Angiofibroma of the Vulva – Case Report

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ABSTRACT

Cellular angiofibroma (CA) is an uncommon benign stromal tumour first reported in 1997. We report the first case in Singapore of a 37 year old woman who presented with a painless vulvar lump which was clinically diagnosed as a Bartholin's gland cyst. A simple excision was carried out under general anaesthesia, with a histopathological diagnosis consistent with CA. There was no documented recurrence post-resection. Differential diagnoses include vulvar-specific lesion like aggressive angiomyxoma (AAM) and angiofibroblastoma (AMF).

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CASE REPORT

A 37 year old, para 4, Chinese woman was referred to our outpatient clinic for further evaluation of a right labial mass that had been enlarging over the past three months. She denied any pain, fever or abnormal discharge. Her last Pap smear performed a year ago was normal. Physical examination revealed a 4 cm mass that was fluctuant non-tender and involving the posterior aspect of the right labia majora. There was no erythema of the overlying skin. Further gynaecological examination was normal and there was no lymphadenopathy. The provisional diagnosis was an uninfected Bartholin's cyst and patient planned for an elective marsupialisation of the cyst under general anaesthesia. The lesion was excised as a single tissue specimen and sent for histopathological analysis. Postoperatively, the patient was covered with oral broad-spectrum antibiotic amoxicillin/clavulanic acid for five days, but an infection of the surgical wound was detected during an outpatient review two weeks later. A pus culture and sensitivity testing of the infected wound grew *Streptococcus agalactiae* sensitive to penicillin, ampicillin and amoxicillin. The patient was restarted on amoxicillin/clavulanic acid and recovery thereafter was uneventful.

PATHOLOGICAL AND IMMUNOHISTOCHEMICAL FINDINGS

The resected specimen weighed 40g and measured 6.8 x 5 x 3.5 cm. Grossly, it was a lobulated but well-circumscribed ovoid mass with finger-like projections. There was an adherent rough fibroadipose tissue on one surface measuring 4 x 2 x 3.5 cm. The cut section showed a pale pink homogenous appearance. Microscopic examination revealed a heavily collagenized spindle cell lesion with plump fibroblast-like cells with no real atypia or mitotic activity as shown in Figure 1. Some cells showed plasmacytoid morphology. A collection of mononuclear inflammatory cells, as well as some mast cells, are present within some entrapped fat as shown in Figure 2. The margin of the lesion varies from well-defined to mildly infiltrative, with finger-like projections also noted. Focally, it extended into striated muscle. The resection margin appeared clear. CD34 stain reveals the presence of an intricate network of capillaries and small vessels. Occasional spindle cells were also CD34 positive. ER and PR were both moderately positive (25% of nuclei staining positively). In view of this above findings, a diagnosis of CA was made. The patient was followed up for 20 months without any evidence of recurrence.

DISCUSSION

CA is an uncommon mesenchymal neoplasm first described by Nucci et al in 1997[1] and so-named due to its two principal components: uniform stromal cells, and prominent blood vessels with mural thickening and hyalinization. In this descriptive study, all four initial cases and two addendums were reported only in middle-aged women and almost exclusively in the vulva, as in our case. Since then, it has been found that CA occurs in both men and women, mainly in the inguinoscrotal and vulvovaginal region[2-4], but females are affected more commonly and they tended to be younger[3, 4]. Other atypical sites of CA have been reported in the knee, eyelid, chest wall, retroperitoneum and oral mucosa[4-7].

CA which presents in the vulva is generally painless and small (approximately 3 cm)[1, 3, 4, 8, 9], although growth of up to 14 cm has been reported[3], and in our patient, the widest diameter measured 6.8 cm. Clinically, it is frequently misdiagnosed as cystic lesion, such as a Bartholin's gland cyst[1, 4].

Classically, the tumours are well-circumscribed, but unencapsulated. They are composed of bland spindle-shaped cells arranged in short, irregularly-intersecting fascicles in a fibrous stroma with bundles of wispy collagen, and numerous small to medium sized vessels with prominent hyalinization of the walls. Mast cells and a variable component of peripherally-located mature adipocytes are also noted[1, 12]. Mitotic activity was described as common in the original series by Nucci et al[1]. However, various deviations from the usual histological picture of CA have been described in the literature. These features include: absence of prominent vascular hyalinization, hypocellular hyalinized areas, dilated haemangiopericytoma-like vascular channels, stromal lymphoid aggregates, centrally-located adipose tissue and foci of nuclear pleomorphism that resemble symplastic change within a uterine leiomyoma[9]. A recent study has reported nine cases of CA with sarcomatous changes, but with no added aggressive biological activity[13]. However, there is limited follow-up data for these patients.

Immunohistochemically, CA is positive for vimentin, and usually negative for smooth muscle markers such as aSMA, desmin and h-caldesmon[1, 9]. This aids diagnosis as most other vulvovaginal stromal lesions are at least focally positive for smooth muscle markers, and especially with desmin. Unfortunately, the above-mentioned markers were not tested in our case. Instead, we note positive staining for CD34, in contrast to the negative results obtained in the original four cases reported by Nucci et al. It was initially thought that CD34 was a differentiating factor between CA and a top differential diagnosis, spindle cell lipoma, which is CD34 positive. However, subsequent two cases in the addendum refuted the original analysis, as were the cases in subsequent publications in the literature[3, 4]. Hence, differentiation between the two entities was to be based on histological findings. As is with our case, most CAs stained positively with ER and PR[9].

Other vulvar neoplasms which may be confused with CA, by virtue of their location, include aggressive AAM and AMF. However, AAM as its name suggests, is locally infiltrative and tends to recur. It is often a large lesion, with an extensive myxoid stroma, and is paucicellular. AMF, on the other hand, shows characteristic alternating hypercellular and hypocellular areas. There are typically foci of cells with epithelioid or plasmacytoid morphology in the more cellular areas and tumour cells tend to be arranged around small-calibre vessels. Both AAM and AMF, in contrast to CA, are desmin and actin positive[7, 12].

To date, no cases with metastasis have been reported in the literature and it appears that CA pursues a benign clinical course. Simple local excision with negative margins is the treatment of choice with curative effects[1, 3, 4, 7]. Local recurrence after excision of the lesion with clear margins is uncommon[10, 11]. The patient we presented in this report had no documented recurrence post-excision of the tumour.

Figure 1. Heavily collagenized spindle cell lesion with plump fibroblast-like cells with no real atypia or mitotic activity

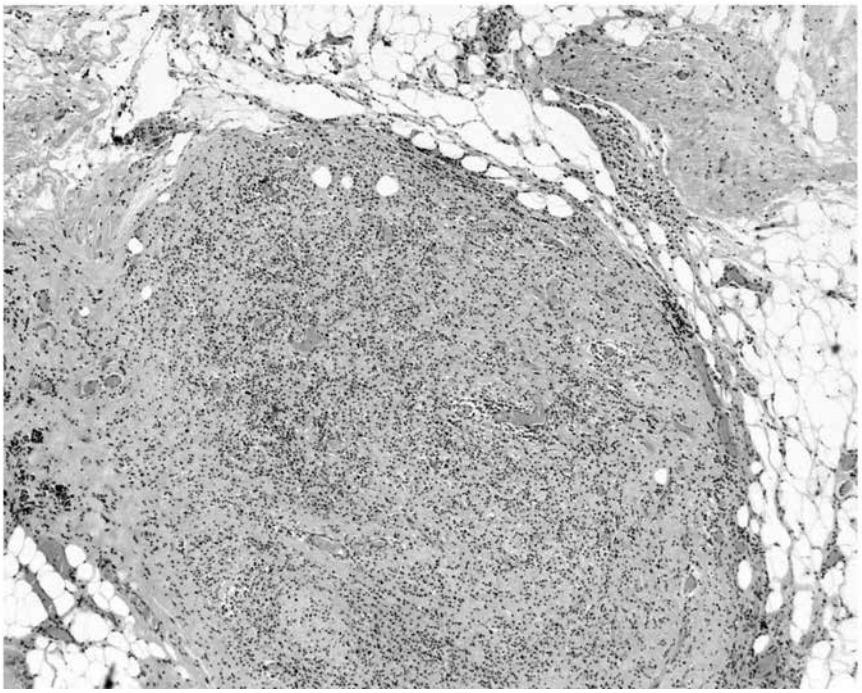
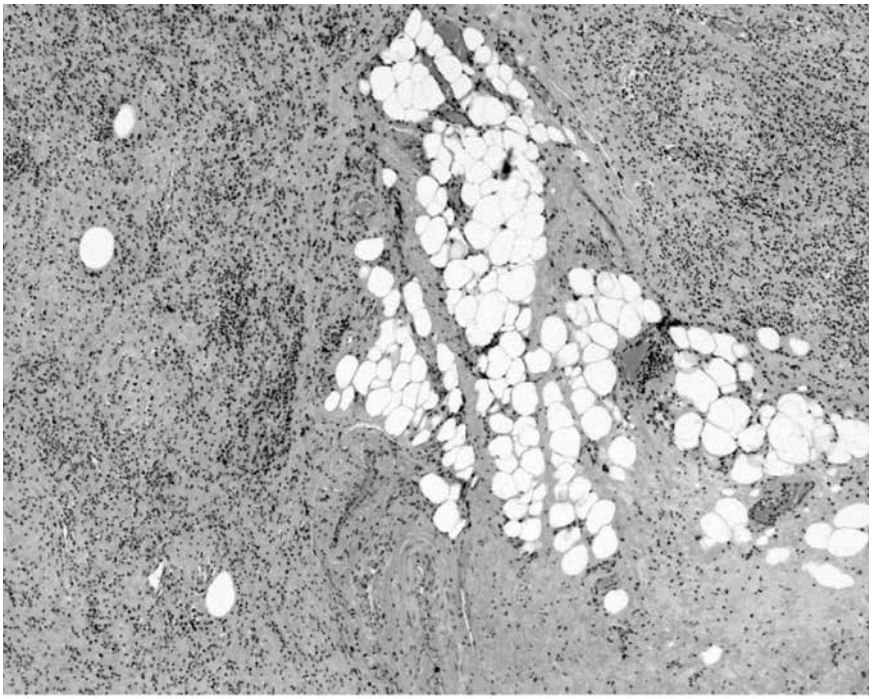


Figure 2. A collection of mononuclear inflammatory cells, as well as some mast cells are present with some entrapped fat



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