An unusual case of a large fibroepithelial stromal polyp presenting as a nipple mass

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Abstract

Background: Fibroepithelial stromal polyps (FESP) are benign lesions that typically occur in the genital area and are known to represent a diagnostic challenge for pathologists. Not only do they have a spectrum of morphological changes that ranges from bland morphology to rather atypical appearances, but they also share morphological features with a number of benign and malignant lesions. This is a report of a rare presentation of a FESP of the breast.

Case presentation: We describe an unusual case of a large polypoid mass arising from the nipple and connected to it by a long pedicle in a female of 45. The lesion comprised spindle and stellate shaped cells with bizarre stromal giant cells. The morphological and immunohistochemical diagnostic features are provided together with a discussion of possible mimics.

Conclusion: FESPs may occur in the female breast. It is important to differentiate the lesion from other benign and malignant spindle cell lesions particularly metaplastic carcinoma.

Keywords: Breast, Nipple, Fibroepithelial polyp

Background

Fibroepithelial stromal polyps (FESP) are benign lesions recognised in the skin, oral cavity, urinary tract [1] and genital area [2]. They are recognised as posing diagnostic problems in the vulvo-vaginal region for the practising pathologist. These lesions, apart from being uncommon, have overlapping morphological features with other mesenchymal lesions specific to the vulvovaginal area. These lesions include angiomyofibroblastoma, aggressive angiofibroma, and cellular angiofibroma. Other lesions that can morphologically mimic FESP are leiomyomas, superficial angiomyxoma, perineureomas and neurofibromas. Although immunohistochemistry has traditionally been described to assist differentiating these lesions, they are of limited clinical value.

FESP are remarkable for their ability to exhibit a wide range of histological appearances. They present in young or middle aged women most commonly in the vagina but also in the vulva [3] and rarely the cervix [4]. Only the clinical presentation of one case of FESP has been recently described in the breast [5]. Here, we present a case of a fibroepithelial stromal polyp arising in the nipple and showing histological appearances of those described in the vulvovaginal region.

Case presentation

This is a 45 year-old lady with no relevant medical history. She presented with a 4x4cm painless nipple lump (Figure 1). The lump has been present for a long time and slowly increased in size. The patient was not pregnant and the increase in size was not related to a previous pregnancy. On clinical examination, the mass was well circumscribed, firm and covered by a stretched but otherwise unremarkable skin. It was connected to the nipple by a long pedicle (Figure 1A-D). The rest of the breast was normal. Breast imaging was otherwise unremarkable.

Clinically, the lesion was likely to be benign though rather unusual. The clinical differential diagnosis included a possible leiomyoma or dermatofibroma.

The patient was treated by local excision of the lesion under general anaesthetic as a day case procedure with no post-operative complications. No further local treatment was given.
The specimen was examined by both breast and soft tissue pathologists by H&E and immunohistochemistry. The latter included EMA, desmin, calponin, caldesmon, EMA, S100, CD34, factor XIIIa, oestrogen receptor and progesterone receptor.

**Macroscopically**

The lesion was a well-defined, polypoid mass that exhibited a pedicle. The mass was firm with a uniform, translucent, whorly, greyish cut section. Apart from the base of the pedicle, the lesion was covered by unremarkable skin. There was no evidence of haemorrhage or necrosis.

**Microscopically**

Sections revealed a rather hypocellular lesion that extended upwards to the overlying epidermis (Figure 2A-C). There was no demarcation between the lesion and the epidermis. The stalk comprised fibromuscular tissue of the nipple and was covered by unremarkable skin including sebaceous units. The lesion comprised a haphazard proliferation of rather bland spindle and stellate-shaped cells. The lesion showed variable cellularity with markedly hypocellular stroma alternating with more cellular areas. Occasional multinucleate stromal giant cells were seen. A sprinkle of mast cells was seen within the stroma. Scattered thin walled blood vessels were seen. There was
no evidence of necrosis and mitoses were not a feature. No plasmacytoid cells, thick-walled vessels or cells with epithelioid morphology were seen.

Immunohistochemistry showed the lesional cells to be focally immunoreactive for smooth muscle actin (5% of cells were stained), and CD34 but negative for desmin, calponin, caldesmon, EMA, S100, oestrogen receptor and progesterone receptor (Figure 2D-H). Scattered through the lesion were factor XIIIa positive dendritic-like cells (approximately 45% of cells were positive).

The patient remained well and follow-up showed no regrowth or recurrence.

Discussion

In this report we present a case of 45 year old female presenting with a large polypoid nipple mass. The site and macroscopic appearances were rather unusual. The case was diagnosed as a benign fibroepithelial stromal polyp reminiscent of the lesions described in the female genital area. Only one recently published report described a similar clinical presentation in a 35 year old female [5]. However from the brief histological description provided, the lesion does not appear to contain stellate or giant stromal cells and therefore may not be identical to the current case.
Histologically, the lesions are characteristically polypoid and usually contain a conspicuous fibrovascular core. The stroma is the most distinctive aspect of the lesion and can exhibit a wide range of appearances. The stroma can be hypocellular, being composed of bland spindle shaped cells with indistinct cytoplasm set within a loose or finely collagenous matrix. Stellate and multinucleate stromal cells are characteristic, being more noticeable at the epithelial-stromal interface [6]. At the other end of the spectrum, the stroma of some FESP exhibit marked cellularity, nuclear pleomorphism and increased mitotic activity including atypical mitoses. These lesions were called “cellular pseudoangiomatous fibroepithelial stromal polyps” [6]. These lesions are morphologically worrisome and may be mistaken for a malignant neoplasm. Cellular FESP tend to exhibit a greater degree of cellularity in the centre of the lesion, becoming less cellular as the lesion extends up to the stromal epithelial interface [6]. The stellate giant cells in FESP of the oral cavity are shown to be derived from fibroblastic lineage [7].

Other lesions that occur in the breast have also been reported in the genital area (e.g. ectopic breast tissue, fibroadenomas, mammary carcinomas) [8,9]. The mammary line extends from the breast superiorly to the genital area and therefore, it is conceivable that certain lesions can be shared between both areas.

The morphological features of the reported lesion are fully in keeping with a benign fibroepithelial stromal polyp. This polypoid lesion abuts the overlying epidermis without a distinct Grenz zone. It had the stellate cells and occasional bizarre multinucleate cells characteristic of FESPs. A conspicuous fibrovascular core was identified together with a sprinkle of mast cells. There were no features to suggest malignancy and no mitoses were seen in the lesion. The MIB-1 proliferation marker is of limited diagnostic value in those lesions and there is no established cut off value for positivity to help separate from other mimics.

FESPs show variable morphological appearances and, as in the vulvovaginal area, need to be distinguished from other mesenchymal lesions. In the breast, a wide range of benign and malignant spindle cell lesions occur in the breast; for review see [10]. In this context, it is important to differentiate spindle cell metaplastic carcinoma—which can appear deceptively bland, from other mesenchymal lesions [11]. This current case shows no expression of epithelial markers. Atypical neurofibroma and pleomorphic fibroma are possible differentials. The former is S100 positive (negative in the current case). Pleomorphic fibroma has atypical single fibroblasts which are CD34 positive and giant cells are not a feature. In the current case, atypia was limited to giant cells and CD34 was only focally positive. Given the postulated origin, the FESPs should also be differentiated from the entity: pseudosarcoma botryoides (fibroepithelial polyps with atypical stromal cells) that was previously described in the vulva and vagina. Although histologically benign, two out of the 13 case series described recurred after incomplete excision [3]. Sarcoma botryoides (embryonal rhabdomyosarcoma), on the other hand, is a frankly malignant tumour that arises under the mucosal surfaces of body orifices such as vagina, bladder and cervix [12]. They often occur at a younger age (childhood, adolescence). The characteristic cambium layer and the pleomorphic spindle cells with rhabdomyoblasts were not seen in the presented case.

The significance of the focal staining with factor Xllla and CD34 is uncertain. Factor Xllla stains dermal dendrocytes and is expressed in a variety of mesenchymal skin tumours e.g. dermatofibromas and fibrous papule of the face. CD34 also stains a wide variety of mesenchymal tumours but the focal nature of expression in this case would reduce any diagnostic significance.

Conclusions

FESPs can occur in the female breast. The presentation is of a long standing slowly growing pedunculated polyp, arising from the nipple. The histological features are those of a variably cellular lesion with spindle, stellate and stromal giant cells. The cells are focally positive for smooth muscle actin but negative for caldesmon, calponin and epithelial markers. In the breast, it is important to differentiate the lesion from metaplastic (spindle cell) carcinoma which can present as a relatively bland spindle cell proliferation.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Abbreviations

FESP: Fibroepithelial stromal polyp; EMA: Epithelial membrane antigen; H&E: Hamatoxylin and Eosin.

Competing interests

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No
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No
Do you hold any other financial competing interests? If so, please specify.
No
Non-financial competing interests.
No

Are there any non-financial competing interests (political, personal, religious, ideological, academic, intellectual, commercial or any other) to declare in relation to this manuscript? If so, please specify.
No

Authors’ contributions
AMS: carried out the macroscopic handling, histological examination and immunohistochemical analysis of the lesion, reviewed the literature for histological differentials/mimics, took the microscopic photos and drafted the manuscript. PT: provided the clinical data, consented patient, provided the macroscopic photographs of the lesion and helped to draft the manuscript. WM: provided the soft tissue expertise into the diagnosis and differential diagnosis of the lesion, advised on the immunohistochemical panel and interpretation and contributed to the writing up. All authors read and approved the final manuscript.

Acknowledgements
The authors wish to thank the department of Medical Illustration at St James’s University Hospital, for the photographic assistance.

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Received: 31 January 2013 Accepted: 29 August 2013 Published: 30 August 2013

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doi:10.1186/1756-0500-6-345
Cite this article as: Shaaban et al.: An unusual case of a large fibroepithelial stromal polyp presenting as a nipple mass. BMC Research Notes 2013 6:345.