Unusual presentation of more common disease/injury

Differential considerations of skin tumours with florid vascularisation: report of a solitary giant vascular eccrine spiradenoma

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Summary

The authors report the case of an 81-year-old male who presented with a 3-year-history of a bluish, nodular tumour located on the extensor side of his right forearm. Subjective symptoms included tenderness upon palpation and spontaneous haemorrhage. In order to exclude malignant neoplasms, for example, nodular melanoma, metastatic melanoma or angiosarcoma, the tumour was surgically removed and tissue submitted for microscopic examination. Histologically, the authors diagnosed this as giant vascular eccrine spiradenoma, a rare variant of eccrine spiradenoma, which can easily be mistaken for angiomatous lesions due to the haemorrhagic features and florid vascularisation. It is our aim to help clarify the diagnosis and differentiate giant vascular eccrine spiradenoma from other painful cutaneous tumours exhibiting a high degree of vascularisation, for example, angiosarcoma or venous thrombosis, as this case represents one of only seven found in published literature.

BACKGROUND

Giant vascular eccrine spiradenoma, a rare variant of eccrine spiradenoma, can easily be mistaken for angiomatous lesions due to the haemorrhagic features and florid vascularisation. In this case study, we aimed to shed light on a case of a clinically misdiagnosed giant vascular eccrine

spiradenoma in order to expand the potential clinical and histological criteria able to help clarify the diagnosis.

CASE PRESENTATION

An 81-year-old male presented with a blue nodule of approximately 3 cm in diameter localised upon the

Figure 1 A blue nodule of approximately 3 cm in diameter appearing sessile and firm in consistency.

Figure 2 Histopathologic features: (A) sharply circumscribed tumour localised in the upper subcutis and entire dermis, (B) papillary-cystic glandular structure containing partially hyalinised and partially solid eosinophilic mucoid material intermixed with inflammatory cells and erythrocytes; (C) remaining areas of the neoplasm were composed of smaller cystic formations of oval-shaped, hyperchromatic cell nuclei with basophilic or slightly eosinophilic cytoplasm; (D) focally weak expression of CEA.

extensor side of his right forearm (figure 1). The lesion appeared sessile and firm in consistency. The tumour was initially noticed 3 years ago and described as painful on palpation. The patient observed a progression of the size of the tumour over the last 2 years, as well as spontaneous haemorrhage. Ultrasonography showed a $14.7 \times 25.7 \times 28.6$ mm sharply circumscribed hyperechoic tumour with discrete vascularisation. A preliminary x-ray of the right forearm did not exclude muscle- or bone-infiltration. The patient's medical history was non-contributory and all results of laboratory investigations were within normal parameters. Surgical excision of the exophytic lesion on the right forearm was performed and the tissue was submitted for microscopic examination. To date (2 months postoperatively), the patient is alive and well with no recurrence or complication.

INVESTIGATIONS

Histopathological findings

Multiple sections of the tissue were obtained and revealed a sharply circumscribed tumour localised in the upper subcutis and entire dermis, causing a rounded protrusion of the epidermis (figure 2A). The tumour was predominantly composed of a papillary-cystic glandular structure containing partially hyalinised and partially solid eosinophilic mucoid material intermixed with inflammatory cells and erythrocytes (figure 2C). The remaining areas of the neoplasm were composed of smaller cystic formations of oval-shaped, hyperchromatic cell nuclei with basophilic or slightly eosinophilic cytoplasm (figure 2B). Between the tumour cells, multiple lymphocytes and plasma cells

were observed in the stroma, as well as wide and angiom-like vessels. Mitotic activity, however, was not increased. Proliferative activity varied between 0% and 15% as demonstrated by Mib1/ Ki67. The immunophenotype of the tumour exhibited characteristic features of eccrine differentiation along with strong expression of pan-cytokeratin MNF116, focal reactivity for S100, and weak expression of the carcinoembryonic antigen (CEA) (figure 2D). The melanocytic markers HMB45 and melan-A were negative. However, endothelial markers CD31 and 34, as well as epithelial membrane antigen (EMA), were not expressed.

DIFFERENTIAL DIAGNOSIS

Giant vascular eccrine spiradenoma may be easily mistaken for other lesions with a marked degree of vascularisation, such as:

- ▶ Angiosarcoma: vascular tumour with a typically high expression of endothelial markers such as CD31, CD34 and podoplanin (D2-40)
- ▶ Venous thrombosis: primarily clinical differential diagnosis – the histopathological findings are usually obvious
- ▶ Malignant melanoma: melanocytic tumour, typically expressing HMB45 and Melan-A; negative in our case; generally with a high reactivity for S100 (only focal reactivity for S100 in our case)
- ▶ Merkel cell carcinoma: neuroendocrine tumour, typically expressing chromogranin A and cytokeratin 20; negative in our case
- ▶ Neuroma: benign cutaneous tumour composed of a partially encapsulated mass of bland Schwann cells and innumerable tiny axons arranged in interlacing fascicles.

The capsule is composed of perineural cells that exhibit EMA positivity; negative in our case. Furthermore, most tumours of this aetiology measure ≤ 0.5 cm in contrast to 3 cm of diameter in our case.

TREATMENT

Treatment consists of a total surgical excision of the tumour.

OUTCOME AND FOLLOW-UP

Surgical excision of the exophytic lesion on the right forearm was performed. To date (2 months postoperatively), the patient is alive and well with no recurrence or complication.

DISCUSSION

Benign tumours of cutaneous appendages are divided into the following categories: eccrine, apocrine, follicular, and sebaceous differentiated neoplasm, as well as complex adnexal neoplasm. Spiradenoma are rare benign tumours of the sweat glands that tend to arise on the head; the neck; the trunk; and, less commonly, the arms followed by the legs. Lesions usually measure approximately 1 cm in diameter and remain stable in size. They are primarily described as sharply circumscribed gray, pink, purple, red, or blue nodules with a smooth surface. The lesions can be painful. Spiradenomas usually appear as solitary tumours, however, cases of linear/ zosteriform/ blaschkoid multiple spiradenomas have been previously described.^{1 2} The rate of malignant transformation is very low. Spiradenomas also may occur in Brooke-Spiegler syndrome, which manifests with cylindromas, spiradenomas, and trichoepitheliomas.^{3–5} Most spiradenomas have been reported to arise in those aged 15–35.

Giant vascular eccrine spiradenoma was first described by Cotton *et al* in 1986 as a rare variant of eccrine spiradenoma.⁶ They reported two cases of an unusually large eccrine spiradenoma with a considerable degree of vascularisation in which both measured over 2 cm. To the best of our knowledge, we are reporting the 7th case of giant vascular eccrine spiradenoma.^{6–11} In contrast to the data published on spiradenoma, in each of these cases the patients were far older than 50 years.^{6–11} Recently, Ko *et al* performed immunohistochemical staining to elucidate the histogenesis of this tumour.⁹ They found that giant vascular eccrine spiradenoma is typically composed of three types of cells: epithelial cells, small basal cells, and myoepithelial cells, and therefore concluded that the indicated origination was the eccrine gland with primary differentiation toward the secretory portion of the secretory coil.⁹ The unusual vascular component of the tumour is conceivably part of an ageing process with degenerative changes to the stroma ('life of lesion'). It could be a matter of so-called ancient lesions, originally described in melanocytic nevi.^{12 13} The vascular pattern in giant spiradenoma could possibly be the result of a reorganisation of the tumour stroma during the course of growing and ageing of the lesion. Due to the considerable degree of vascularisation and spontaneous haemorrhage it is often difficult, both for clinicians and pathologists, to differentiate these tumours from other angiomatous tumours. In the six cases described previously, the tumour was misdiagnosed as

Table 1 Clinical and histological criteria for giant vascular eccrine spiradenomas

Clinical criteria	Histological criteria
▶ Sharply circumscribed gray, pink, purple, red, or blue nodule with smooth surface	▶ Cystic-haemorrhagic glandular structures
▶ Solitary tumour	▶ Three types of cells: pale epithelial cells, small basal cells, and myoepithelial cells
▶ Diameter > 2 cm	▶ Wide, angiom-like vessels in the stroma
▶ Spontaneous haemorrhage	
▶ Can be painful on palpation	
▶ Arises in patients aged > 50 years	

angiosarcoma, malignant melanoma, neuroma, sebaceous cyst, angiolioma, or venous thrombosis.^{6–11} In our case, we discussed as primary clinical diagnosis 'merkel cell carcinoma'.

With the patient presented here we aim to report an additional case of a clinically misdiagnosed giant vascular eccrine spiradenoma, and we intend to characterise the clinical and histological criteria in order to help clarify the diagnosis (table 1).

Learning points

- ▶ Giant vascular eccrine spiradenoma is a rare variant of eccrine spiradenoma, and can easily be mistaken for angiomatous lesions due to the haemorrhagic features and florid vascularisation.
- ▶ Clinical criteria: sharply circumscribed gray, pink, purple, red, or blue nodule with smooth surface; solitary tumour; diameter > 2 cm; spontaneous haemorrhage; can be painful on palpation; arises in patients aged > 50 years.
- ▶ Histological criteria: cystic-haemorrhagic glandular structures; three types of cells: pale epithelial cells, small basal cells, and myoepithelial cells; wide, angiom-like vessels in the stroma.
- ▶ Therapy consists in a total surgical excision of the tumour.

Competing interests None.

Patient consent Obtained.

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