

Recurrent Atypical Eccrine Spiradenoma of the Forehead

SERGIO DELFINO^{1,2}, VITO TOTO¹, BENIAMINO BRUNETTI¹, ALEXANDER BIANCHI³,
ALFONSO BALDI³ and PAOLO PERSICHETTI¹

¹Department of Plastic and Reconstructive Surgery, University "Campus Bio-Medico", Rome;

²Department of Plastic and Reconstructive Surgery, University "La Sapienza", Rome;

³Department of Biochemistry and Biophysics "F. Cedrangolo",
Section of Pathology, Second University of Naples, Italy

Abstract. Eccrine spiradenoma is an uncommon but well recognized benign adnexal tumour of the eccrine sweat glands. It can appear at any age, but the highest rate of incidence is observed among young adults, without any sex predilection. The commonest clinical presentation is a solitary blue-red dermal or subcutaneous cystic nodule, ranging from 0.3 to 5 cm in diameter. Malignant transformation is a rare phenomenon, presenting as a rapidly enlarging nodule within a long-standing lesion. We report a case of eccrine spiradenoma in a thirty-six-year-old man who presented with a recurrent soft-tissue neoplasm of the forehead, with cytological atypia. The differential diagnosis of this tumour, as well as its possible malignant transformation, is discussed.

Eccrine spiradenoma, also known as spiradenoma, is a benign tumour of sweat glands, first described by Kersting and Helwing in 1956 (1). It usually presents as a single skin tumour, with typical histological features.

It is a relatively uncommon neoplasm that appears mainly in young adults, equally in both sexes and is rarely familial. Its most common anatomical sites are on the back of the trunk and proximal limbs. The lesion is typically solitary and painful and consists of a firm, rounded, bluish, dermal nodule 3-50 mm in diameter. The nodule develops most usually on the head, neck and dorsal aspect of the trunk and it is usually recurrent. A frequent symptom of this neoplasm is spontaneous pain or tenderness on palpation. Malignant transformation is rare, but possible and described in literature

Correspondence to: Dr. Sergio Delfino, Department of Plastic and Reconstructive Surgery, Faculty of Medicine, University Campus Bio-Medico of Rome, Via E.Longoni,83 - 00155 Rome, Italy. Tel: +390622541567, Fax: +390622541456, e-mail: s.delfino@unicampus.it

Key Words: Eccrine spiradenoma, histopathological subtype, anatomical distribution.

(2). It is sometimes misdiagnosed and confused with other cystic neoplasms, which causes inadequate surgical excision with consequent high rate of recurrence.

Differential diagnosis includes other sweat gland tumours, sebaceous cysts and benign subcutaneous connective neoplasms such as fibroma, fibrolipoma and dermatofibroma (3). Radical surgical excision is mandatory to avoid recurrence.

Case Report

A 36-year-old man presented to our Department with a recidivant palpable lesion on his forehead. Family history and past medical history were not significant.

Physical examination revealed a tumefaction of 2x1 cm, tender, painful, movable on deep layers and adherent to superficial derma, with clinically distinct margins. The lesion was previously excised twice in another hospital, but the patient did not provide any documented histological diagnosis. The tumour was covered by clinically normal skin with the presence of a well-formed scar of 1.5 cm of length. At first the lesion was clinically suspected to be a cystic neoplasm, but in consideration of its recurrence, an ultrasonographic examination was programmed. Radiological evaluation revealed a nodular, anechoic soft-tissue mass with regular borders and posterior acoustic increased echo, and suggested its excision and histological characterization. The tumour was excised *en bloc*. Histological evaluation revealed several neoplastic nodules located in the dermis without connection with the epidermis; the lobules were sharply demarcated and displayed a fibrous capsule (Figure 1A). On higher magnification, the epithelial cells within the tumor lobules were found to be arranged in intertwining cords; moreover, some cytological atypia were present (Figure 1B).

The histological examination demonstrated incomplete tumour excision and a new surgical intervention was programmed. During the second procedure, a larger resection was performed involving the scar and 3 mm of clinically normal surrounding tissue until the frontalis muscle. The

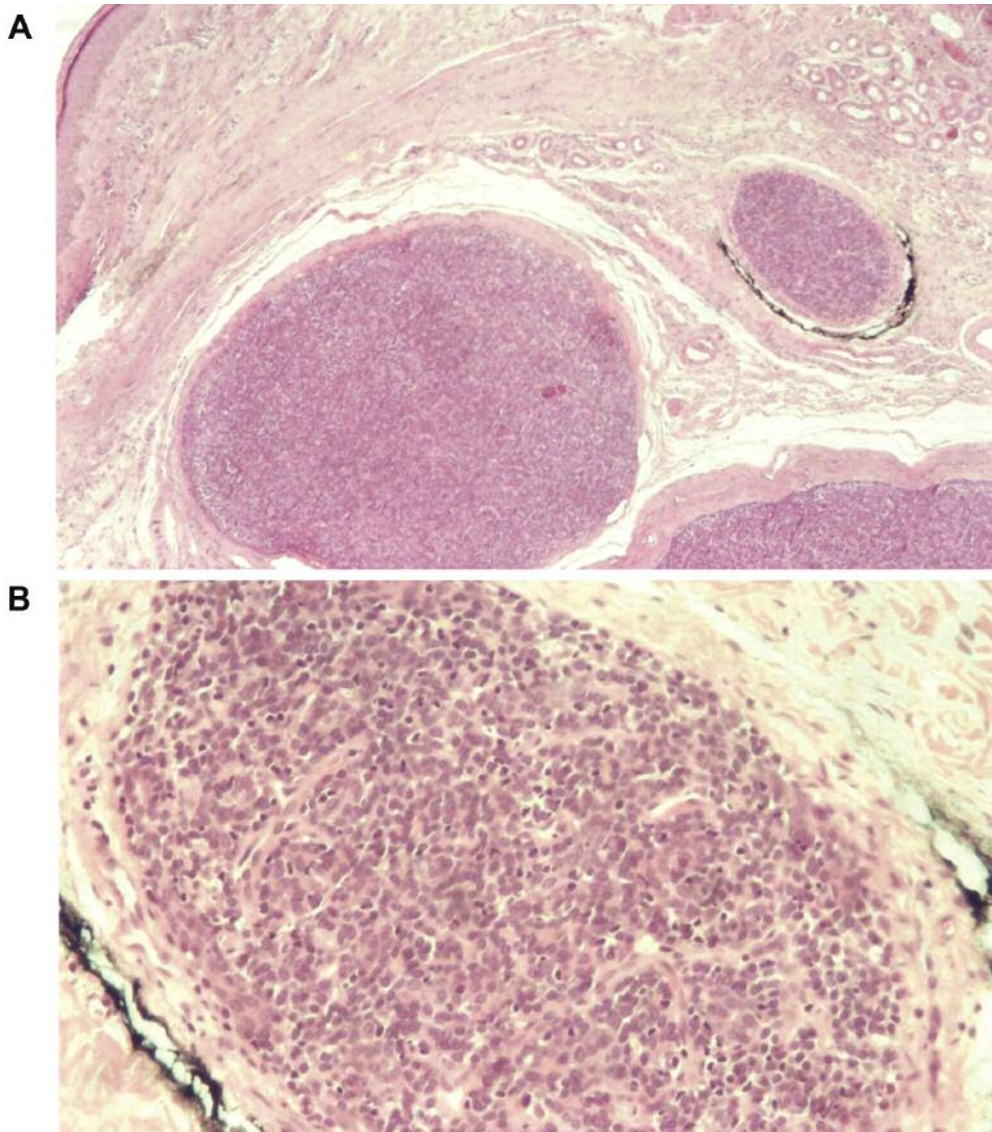


Figure 1. A. Microscopical aspect of the neoplasm showing the cystic appearance (Haematoxylin/eosin, original magnification $\times 10$). B. Higher magnification of the neoplasm (Haematoxylin/eosin, original magnification $\times 20$).

subsequent histological evaluation confirmed the diagnosis of eccrine spiradenoma with some atypical cells and frequent mitoses, and showed tumour-free margins.

Discussion

Eccrine spiradenoma is a benign tumour of sweat gland origin, arising from the intradermal straight part of the duct of eccrine sweat glands. The lesion commonly appears as a solitary nodule but multiple lesions may also be present, becoming confluent or remaining discrete. The epidermis that covers the lesion may be normal in colour or pinkish and ulcerated (4).

Eccrine spiradenoma belongs to the group of the painful tumours of the skin, also known with the acronym BLEND AN EGG. This group includes blue rubber bleb naevus, leiomyoma, eccrine spiradenoma, neuroma, dermatofibroma, angioliopoma, neurilemmoma, endometrioma, glomangioma and granular cell tumour (5).

Histologically, the tumour is lobular, with two types of cells in the islands. Larger, paler cells are grouped around lumina and smaller, darker cells form the periphery (6). Cystic spaces or small, tubular structures may also be present. Large, thin-walled and dilated vascular channels often occur and condensed connective tissue surrounds the lobules of the neoplasm (7).



Figure 2. The scar 6 months' post-operative.

Eccrine spiradenoma can cytologically simulate an adenoid cystic carcinoma on fine-needle aspiration cytology (FNAC), because both tumours contain hyaline globules (8). The pain due to an eccrine spiradenoma is related to the presence of small unmyelinated axons in the context of the connective tissue around the tumour, or to the expansion of the cysts. In contrast, the pain arising from an adenoid cystic carcinoma is related to its infiltrative growth and tendency for perineural invasion.

Old tumours often present degenerative changes. When a malignant transformation occurs, necrosis and degeneration obscure the histological features and only focal areas of the neoplasm show the typical features of a spiradenoma. Malignant degeneration of spiradenoma usually appears in long-standing tumours and is clinically revealed by a rapidly enlarging tumour mass (9). In these cases, the diagnosis of spiroadenocarcinoma is made only if a residual benign component is identified. Clinical differentiation from other dermal tumours and cysts may be made if the tumour is a firm dark-blue colour nodule.

The lesion reported in this paper was initially benign, but the last histological evaluation revealed the presence of some atypical cells. With this report, we would underline the importance of a correct diagnosis of this tumour, followed by an early and complete excision (Figure 2), to avoid recurrence and possible malignant transformation in long-standing lesions.

Acknowledgements

This work has been supported by grants from the International Society for the Study of Comparative Oncology, Inc. (ISSCO) Silver Spring, MD, USA and FUTURA-Onlus to A. Baldi.

References

- 1 Kersting DW and Helwing EB: Eccrine spiradenoma. *Arch Dermatol* 73: 199-227, 1956.
- 2 Mambo NC: Eccrine spiradenoma: Clinical and pathologic study of 49 tumors. *J Cutan Pathol* 10: 312-320, 1983.
- 3 Castro C and Winkelmann RK: Spiradenoma: Histochemical and electron microscopic study. *Arch Dermatol* 109: 40-48, 1974.
- 4 Kaleeswaran AV, Janaki VR, Sentamilselvi G and Kiruba Mohan C: Eccrine spiradenoma. *Indian J Dermat Venereol Leprol* 68: 236-237, 2002.
- 5 Naversen Dn, Trask DM, Watson FH and Burkett JM: Painful tumours of the skin "BLEND AN EGG". *J Am Acad Dermatol* 28: 298-300, 1993.
- 6 Hashimoto K and Lever WF: Histogenesis of skin appendage tumors. *Arch Dermatol* 100: 356-369, 1969.
- 7 van den Oord JJ and de Woolf-Peeters C: Perivascular spaces in eccrine spiradenoma. *Am J Dermatopathol* 17: 266-270, 1995.
- 8 Kolda TF, Ardaman TD and Schwartz MR: Eccrine spiradenoma mimicking adenoid cystic carcinoma on fine-needle aspiration. A case report. *Acta Cytol* 41: 852-858, 1997.
- 9 Cooper PH, Frierson HF Jr and Morrison C: Malignant transformation of eccrine spiradenoma. *Arch Dermatol* 121: 1445-1448, 1985.

Received May 5, 2008

Revised August 6, 2008

Accepted August 27, 2008