

Extrapleural solitary fibrous tumour with haemangiopericytic pattern of the breast. An uncommon neoplasm

Tumor fibroso solitario extrapleural con patrón hemangiopericítico de la mama. Una neoplasia infrecuente

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SUMMARY

Extrapleural solitary fibrous tumour is a mesenchymal tumour, probably of fibroblastic type, occurring only rarely in the breast. A case of solitary fibrous tumour of the breast is presented and the possible origin of this neoplasm is discussed. A 43-year-old woman presented with a ten month history of a painless, enlarging mass in the left breast, which was seen on mammography to be a 10 cm, dense, nodular, well delimited lesion. A left mastectomy without axillary dissection was performed. Histologically, the tumour was composed of a proliferation of haphazardly distributed fusocellular elements and varying degrees of stromal collagenization. Medium-sized thin-walled blood vessels in a haemangiopericytic growth pattern were observed. The immunohistochemical study showed strong CD34 and CD99 positivity. This case represents a typical extrapleural solitary fibrous tumour with a haemangiopericytoma pattern. Our findings would indicate that the most probable origin of this lesion was the perilobular or interlobular stroma.

Keywords: Breast, solitary fibrous tumor, immunohistochemical analysis.

RESUMEN

El tumor fibroso solitario extrapleural es un tumor mesenquimal de probable origen fibroblástico. La mama es un sitio infrecuente para un tumor fibroso solitario extrapleural. Este trabajo documenta las características clínicas, histológicas e inmunohistoquímicas en un caso de tumor fibroso solitario de la mama, y discute el probable origen de esta neoplasia. Se trata de una mujer de 43 años que consultó por una masa no dolorosa en la glándula mamaria izquierda de diez meses de evolución. La mamografía izquierda mostró una lesión nodular de 10 cm densa, bien definida. Se realizó mastectomía sin vaciamiento axilar. Tras cirugía, la totalidad del tumor fue enviado para estudio histopatológico. El tumor estaba formado por una proliferación de células fusiformes distribuidas irregularmente en un estroma con áreas de variable colagenización y vasos sanguíneos de tamaño mediano y paredes delgadas en un patrón de crecimiento hemangiopericítico. El estudio inmunohistoquímico demostró fuerte positividad para CD34 y CD99. Nuestro caso representa un típico tumor fibroso solitario extrapleural con patrón hemangiopericítico. Los hallazgos descritos apoyan al estroma perilobular o interlobular como probable origen de estas lesiones.

Palabras clave: Mama, tumor fibroso solitario, inmunohistoquímica.

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INTRODUCTION

Extrapleural solitary fibrous tumour (ESFT) is a mesenchymal tumour of probably fibroblastic type which shows a prominent haemangiopericytoma-like branching vascular pattern (1-3). Most extrapleural SFTs were called haemangiopericytomas in the past. The breast is an uncommon location, with a few cases documented in previous reports (4-9). This report documents

the clinical, histological and immunohistochemical characteristics in a case of breast solitary fibrous tumour. We also discuss the probable origin of this neoplasm.

CASE REPORT

A 43-year-old woman presented with a painless growing mass in left mammary gland evolving for ten

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TABLE 1. Antibodies used for immunohistochemical studies

Antigen	Clone	Source	Dilution	Pretreatment	Tumour cells reactivity
CK AE1/AE3	AE1/AE3	Dako,Carpinteria, CA, USA	1:50	Steamer	Negative
CK 7	OV-TL 12/30	Dako,Carpinteria, CA, USA	1:50	Steamer	Negative
EMA	E29	Dako,Carpinteria, CA, USA	1:100	Steamer	Negative
Desmán	DE-R-11	Dako,Carpinteria, CA, USA	Prediluted	Steamer	Negative
Anti-Myo D1	5.8 A	Dako,Carpinteria, CA, USA	1:50	Steamer	Negative
S100	Polyclonal	Dako,Carpinteria, CA, USA	1:200	Steamer	Negative
CD34	QBEnd10	Dako,Carpinteria, CA, USA	1:50	Steamer	Positive
SMA	1A4	Dako,Carpinteria, CA, USA	1:100	None	Negative
CD99	12E7	Dako,Carpinteria, CA, USA	1:50	Steamer	Positive
BCL-2 oncoprotein	124	Dako,Carpinteria, CA, USA	1:100	Steamer	Positive focal
		Dako,Carpinteria, CA, USA	1:100	Steamer	Negative
CD117	Polyclonal	Dako,Carpinteria, CA, USA	1:300	Steamer	Negative
Vimentin	V9	Dako,Carpinteria, CA, USA	1:100	Steamer	Positive

CK: Cytokeratin; EMA: Epithelial Membrane Antigen; SMA: Smooth Muscle Actin; Steamer: Epitope retrieval, Black & Decker steamer in Dako target retrieval solution High pH (30 min).

months. Left breast mammography showed a dense, nodular, well delimited lesion, 10 cm in diameter. Tru-cut biopsy was performed and the diagnosis was consis-

tent with mesenchymal neoplasm, even though we could not discard fibroadenoma, phyllodes tumour, metaplastic carcinoma or pure stromal tumour. A left mastectomy without axillary dissection was performed. After surgical treatment was carried out, the whole tumour was submitted for histopathological study.

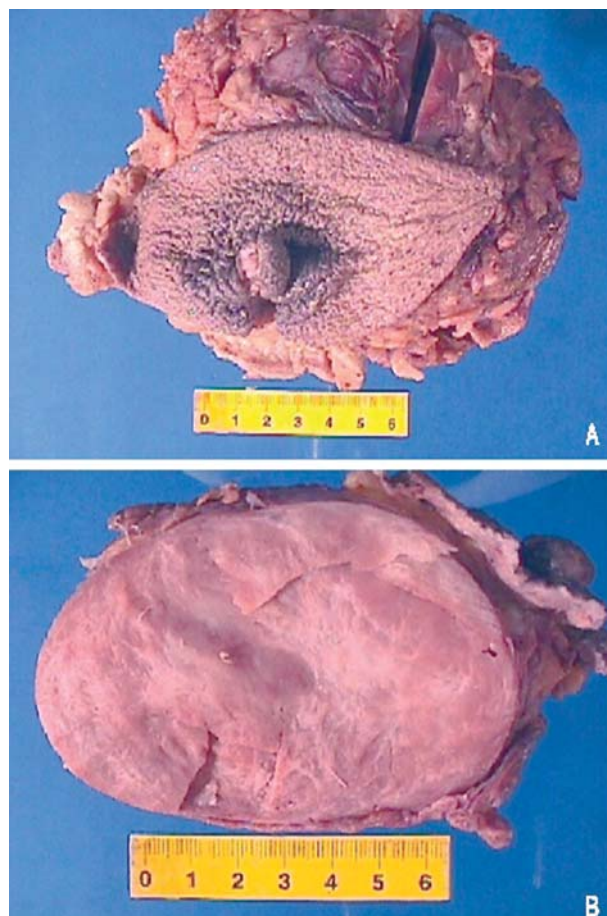


Fig. 1: Macroscopic aspect of breast ESFT. A. Mastectomy specimen showing areola-nipple complex without alterations. Breast skin is also normal. B. Well defined tumour, encapsulated, no hemorrhage or necrosis present.

MATERIALS AND METHODS

The excised specimen was fixed in 10% buffered formalin and processed for routine histopathological study. Immunohistochemistry was performed on paraffin embedded tissue. The thin-sliced materials were immunostained by an enzyme-conjugated polymer backbone (EnVision™ Systems, DAKO, Carpinteria, CA, USA). For specific immunohistochemical details see table 1.

RESULTS

Gross Pathology

Grossly the tumour appeared as an encapsulated, tan-pink mass measuring 10 × 6 × 4 cm. At cut sections the tumour was elastic, well delimited, with gray-white areas. Neither haemorrhage nor necrosis were present. The areola-nipple complex did not show alterations (fig. 1).

Histopathology

Microscopically, the tumour showed a patternless architecture characterized by a combination of hypocellular and hypercellular areas with branching, medium-sized thin-walled blood vessels in a haemangiopericytic growth pattern. The round to spindle-shaped tumour

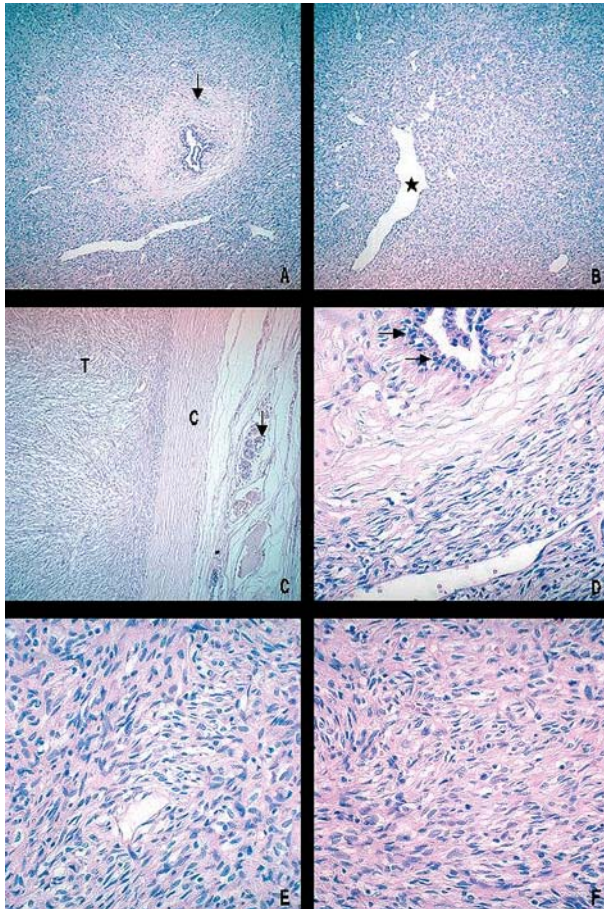


Fig. 2: Microscopic finding. A. Typical histopathologic aspect of ESFT showing hypocellular (peri-ductular, [arrow]) and hypercellular areas (H-E, 100X). B. Characteristic branching haemangiopericytoma-like vessels (asterisk) (H-E, 100X). C. Well demarcated, encapsulated (C) tumour (T), normal breast tissue is seen (arrows) (H-E, 100X). D. Detail of peri-ductular tissue, showing hypocellular area and normal secretory epithelium (arrows) (H-E, 400X). E,F. Characteristic spindle cell tumour cells with indistinct borders and disperse chromatin within vesicular nuclei. No cytologic atypia or necrosis was seen (H-E, 400X).

cells had little cytoplasm with indistinct borders. Nuclei were predominantly vesicular with dispersed chromatin. Cellularity greatly varied in different areas with predominance of hypercellular areas. Mitotic index was 2-3 per 10 HPF. At the periphery of the tumour a clear capsule was present. The epithelial elements were entrapped by the mesenchymal neoplastic proliferation (fig. 2).

Immunohistochemistry

tumour cells showed expression for vimentin, CD34, CD99 (fig. 3). Focal positivity to bcl-2 was present. Epithelial, neural, muscular markers and c-kit (fig. 3) were completely negative.

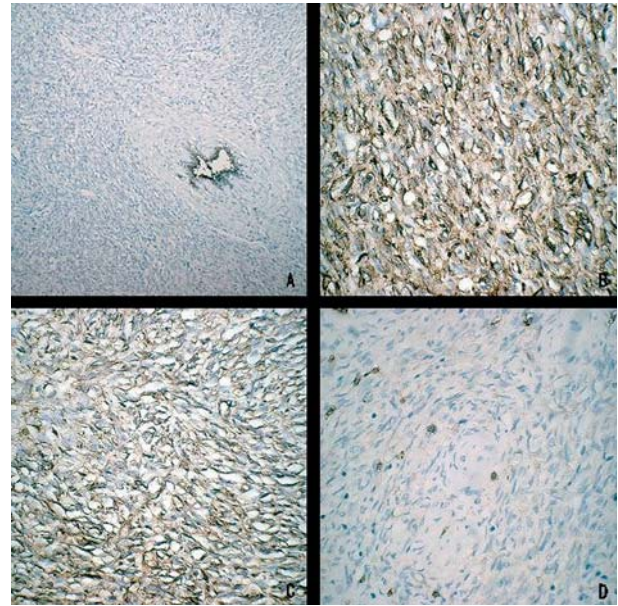


Fig. 3: Immunohistochemical profile. A. Cytokeratin 7 stain showing a strong reactivity in the normal ductal cells. No positivity is seen in neoplastic cells (DAB, 100X). B,C. CD34 and CD99 expression in the spindle-shaped tumour cells (DAB, 400X). D. Mast cells are immunoreactive to CD117. Note the negativity in the tumour cells (DAB, 400X).

DISCUSSION

Extrapleural solitary fibrous tumour is an infrequent breast neoplasm of which at least twenty cases, previously called haemangiopericytoma, were described in English literature (4-9). Clinically, patients present painless tumours, and mammograms show well circumscribed dense areas (8). Our case represents a typical ESFT example with classical branching haemangiopericytoma-like vessel pattern. We prefer the term solitary fibrous tumour rather than haemangiopericytoma since the identification of an haemangiopericytoma as a separate entity may become obsolete because its histological features are shared by a variety of soft tissue tumours (10).

In small biopsies diagnosis represents a challenge and differential diagnoses should include cellular fibroadenoma, phyllodes tumour and metaplastic carcinoma. Cellular fibroadenoma shows proliferative stromal areas pushing out into a prominent frond-like pattern. Fibroblast density is increased, but nuclei are not enlarged or anaplastic. Some typical mitoses can be seen (11) Phyllodes tumours are macroscopically characterized by cysts. Microscopically, phyllodes tumours show frond-like proliferation of epithelial and stromal elements, and marked cellular fibroblastic components with variable mitosis and anaplasia (11) degrees. Finally, metaplastic carcinomas may be indistinguishable from mesenchymal tumours, and multiple histological sections and antibodies to keratins are usually useful in resolving this problem. The ESFT is a pure mesenchymal lesion with no

frond-like epithelial proliferation of epithelial, and as we demonstrated, normal breast epithelial component is entrapped by the stromal proliferation. The use of immunohistochemical studies is of limited value, because fibroadenomas and phyllodes tumours show similar expression for CD34, bcl-2 and CD99 (12).

The origin of breast ESFT is unknown. In normal breast, CD34 staining is seen in the perilobular and to a lesser extent in the interlobular stroma (13-16). In several tissues, CD34 positive fibroblast-like cells have been described in the mesenchyme, especially surrounding structures such as vessels and nerves in several tissues (17-20, 15-17). A number of CD34 positive lesions, such as dermatofibrosarcoma protuberans in the skin (21), intestinal fibroid polyps (22), occur at sites where there are normal CD34-positive fibroblast-like cells. Thus it has been suggested that such lesions may arise from these CD34 positive fibroblast-like cells. Interestingly, previous reports of breast ESFT, as in our case, have showed strong CD34 expression. Probably this finding is consistent with the spindle cell component of these lesions arising from the perilobular or interlobular stroma in a similar manner to that proposed for CD34 positive lesions at other locations.

In conclusion, we present a typical breast ESFT case. These tumours have a benign clinical course. Wide local surgery is often enough for complete tumour excision. Probably, the histogenetic origin of breast ESFT is the peri-ductular or peri-lobular CD34-fibroblast positive cell.

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