

Epithelioid Myofibroblastoma of Mammary-type in Chest Wall – A Case Report –

Hyun Jung Kim
Hunkyung Lee¹ • Ok-Jun Lee²
Kyung Ja Cho² • Jae Y. Ro²

Department of Pathology, Inje University, Sanggye Paik Hospital, ¹Ewha Clinic, ²University of Ulsan College of Medicine, Asan Medical Center, Seoul, Korea

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Corresponding Author

Jae Y. Ro, M.D.
Department of Pathology, University of Ulsan College of Medicine, Asan Medical Center, 388-1 Pungnap-dong, Songpa-gu, Seoul 138-736, Korea
Tel: 02-3010-4550
Fax: 02-472-7898
E-mail: jaero@amc.seoul.kr

Mammary-type myofibroblastoma of the soft tissue is a benign mesenchymal tumor, and it is a recently established clinical entity. We report a case of myofibroblastoma of the chest wall with a prominent epithelioid feature, that occurred in a 12-year old female. Although the lesion occurred in the breast area, there was no breast parenchyma in or around the mass, which favored soft tissue of the chest wall origin. The tumor was immunohistochemically identical to the mammary-type myofibroblastoma with diffuse and strong positivity against CD34 and desmin. The myoepithelial differentiation of the tumor was further supported by the electron microscopic analysis. This case indicates that mammary-type myofibroblastoma can occur in a young girl. The mammary-type myofibroblastoma should be considered a differential diagnosis, among epithelioid soft tissue neoplasms in the chest wall when the proper immunohistochemical work-up is done.

Key Words : Mammary-type myofibroblastoma; CD34; Desmin; Epithelioid feature

Myofibroblastoma of the breast is a benign mesenchymal tumor that was first formally characterized in 1987 by Wargotz *et al.*¹ Mammary type myofibroblastoma is generally a well circumscribed tumor, composed of spindle cells in variable sized fascicles; it is embedded in a hyalinized stroma with varying amounts of intralesional and interlesional fatty components, and there are commonly prominent mast cells. The characteristic immunohistochemical profile displays a dual expression of desmin and CD34.^{2,3} Mammary-type myofibroblastoma of soft tissue was reported by McMenamin and Fletcher⁴ in 2001. They found the same immunohistochemical characteristics of mammary-type myofibroblastoma, and claimed that this tumor might be closely related to spindle cell lipoma. Unlike the previously reported cases, we experienced a case of a mammary-type myofibroblastoma with a prominent epithelioid feature occurring in the chest wall of a very young aged girl.

CASE REPORT

A 12 year-old female visited a local clinic with a left anterior

chest wall mass. It had been slowly growing for the past three years. Other past medical history and family history were unremarkable. On physical examination, a well-circumscribed, apple-sized mass was located in the subcutaneous tissue between the left breast and the anterior chest wall. Pain and tenderness were not present. The mass was completely excised and the patient has been alive and well for one year after the surgery.

The resected mass 5 cm-sized, well circumscribed, lobulated, round and firm mass. The cut surface was grayish-white and fibromyxoid without any areas of hemorrhage or necrosis (Fig. 1).

Light microscopic examination demonstrated a mass with sharp demarcation, nodular growth pattern and intervening collagenous stroma (Fig. 2). The tumor showed a biphasic pattern with epithelioid and spindle cells. The epithelioid feature was predominating, and was mainly seen in the center of the tumor. The tumor cells had oval to round nuclei and abundant eosinophilic cytoplasm. They had mild nuclear pleomorphism with occasional prominent nucleoli. Mitotic activity was absent (Fig. 3). These tumor cells were arranged in clusters or in an alveolar growth pattern. Some of the tumor cells had intracytoplasmic or intercellular lumina that contained eosinophilic material. In



Fig. 1. A well-circumscribed, 5 cm-sized grayish-white firm mass with fibromyxoid cut surface.

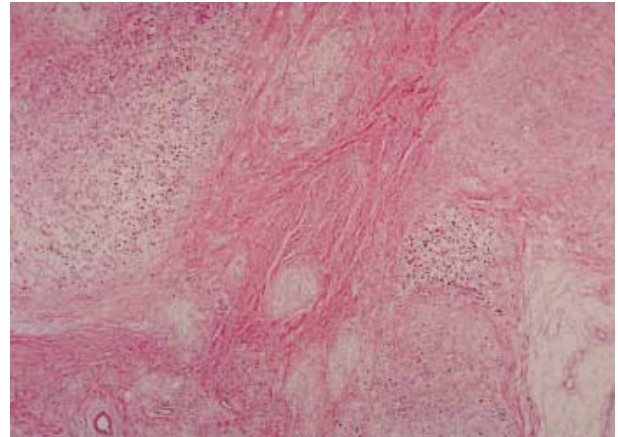


Fig. 2. Nodular growth pattern with intervening collagenous stroma ($\times 10$).

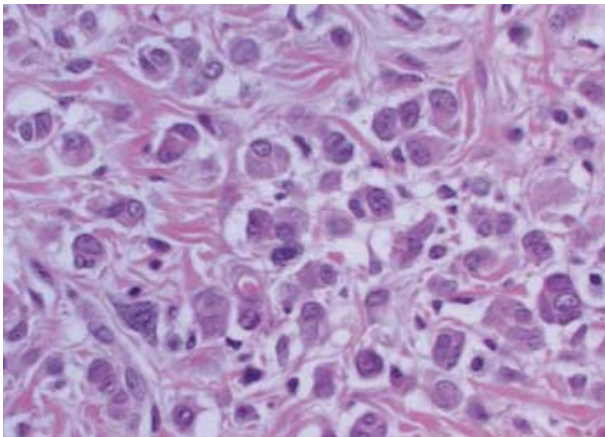


Fig. 3. Predominantly epithelioid features showing cellular clustering and alveolar pattern with oval to round nuclei and eosinophilic granular cytoplasm ($\times 200$).

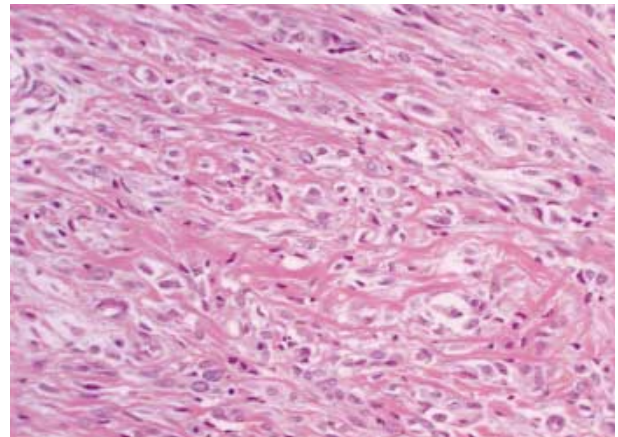


Fig. 4. Spindle cell areas consisting of plump elongated cells admixed with abundant collagenous stroma ($\times 100$).

some areas, tumor cells were compressed by collagenous stroma with an Indian filing arrangement, which was reminiscent of lobular carcinoma of the breast. Mast cells were frequently found throughout the tumor. The spindle cell component was minor and seen at the periphery of the tumor. The spindle tumor cells were admixed with abundant collagenous stroma, and the cells revealed elongated nuclei (Fig. 4). An adipose tissue component was not detected.

The tumor cells were diffusely and strongly positive against vimentin, CD34 (Fig. 5A), and desmin (Fig. 5B), and they were focally positive against smooth muscle actin and estrogen receptor; they were negative for cytokeratin, factor VIII related antigen, CD31, S-100 protein and progesterone receptor. The immunostaining results indicated a tumor with myofibroblastic differentiation.

The electron microscopic findings of the tumor cells were com-

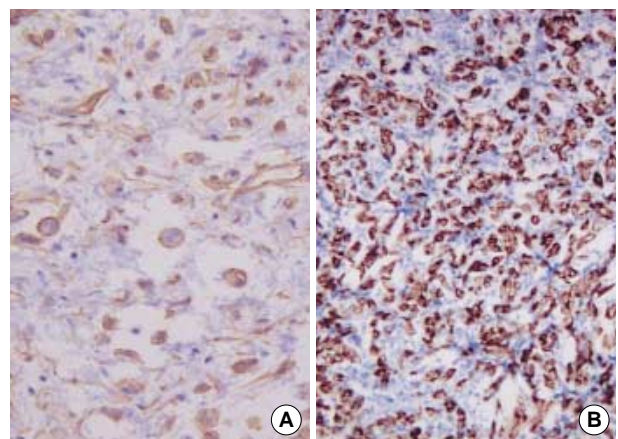


Fig. 5. Tumor cells with positive immunoreactivity for CD34 (A) and desmin (B).

patible with myofibroblasts having numerous dilated rough endoplasmic reticula, a large amount of microfilaments, and a focal

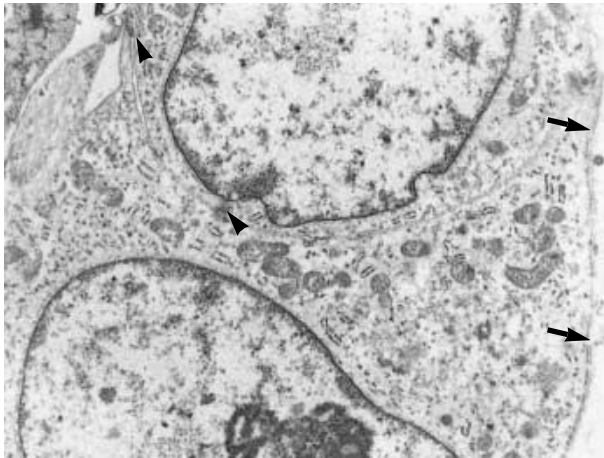


Fig. 6. Epithelioid tumor cells show dilated rough endoplasmic reticula, basal lamina, focal densities (arrows) and occasional well-developed cell junctions (arrow heads).

basal lamina with densities. In addition, the tumor cells revealed occasional well-developed cell junctions, that have not been described before in myofibroblastoma (Fig. 6).

DISCUSSION

Myofibroblastoma of the breast is a benign mesenchymal tumor that was first formally characterized by Wargotz *et al.*¹ in 1987. This tumor has been reported in the literature under different names such as “benign spindle cell tumor”,⁵ “spindle cell lipoma”,⁶ “fibroma”,⁷ “solitary fibrous tumor”,⁸ “myofibroblastoma”,¹ “myogenic stromal tumor”⁹ and “atypical variant of leiomyoma”.⁹ Mammary myofibroblastoma is generally well circumscribed and is composed of spindle cells in fascicles. These fascicles are embedded in hyalinized stroma, that contains numerous mast cells. The tumor usually has a fatty component that shows intralesional and interlesional variability in amount, and a few cases have shown smooth muscle or cartilaginous differentiation.¹⁰ The mammary myofibroblastoma is characterized by dual expression of desmin and CD34. Some cases are also positive for smooth muscle actin. Mammary-type myofibroblastomas have been reported as being definitely benign with no recurrence or metastasis after surgical excision. The earlier reports showed an apparent predilection for old male patients, and especially in patients with gynecomastia.¹¹

Variant forms of myofibroblastoma have been reported,¹² including the collagenized/fibrous, epithelioid, cellular and infiltrative types. The epithelioid myofibroblastoma with sclerotic stroma, as was seen in the present case, has a linear growth arrange-

Table 1. Clinicopathologic features of mammary-type myofibroblastomas arising in extra-mammary locations⁴

Case No.	Sex/Age	Site	Size (cm)
1	M/52	Anterior abdominal wall	6 × 4.5
2	F/58	Posterior vaginal wall	2 × 1.5 × 1.5
3	M/52	Left inguinal region	7.3 × 6.0 × 2.5
4	M/67	Mid-back	2 × 1.5 × 1.5
5	M/62	Right groin	6
6	M/35	Left inguinal area	6 × 3 × 1
7	M/60	Left parasternal area	13
8	M/53	Right inguinal area	3.5 × 3.0 × 1.4
9	F/43	Left buttock	5.5 × 4.0 × 3.5
10	M/59	Suprapubic region	4
The current case	F/12	Anterior chest wall	5

ment of epithelioid cells that may resemble an invasive lobular carcinoma. However, the epithelioid cells were not immunoreactive for cytokeratin and they are strongly positive for vimentin, desmin and CD34; this indicates a mammary-type myofibroblastic differentiation.

A mammary-type myofibroblastoma of the soft tissue was recently described by McMenamin and Fletcher,⁴ and only a few more cases have been reported since then. McMenamin and Fletcher⁴ reported 10 cases of myofibroblastoma, which were morphologically similar to the spindle cell tumor of the breast, and they were designated as “mammary-type myofibroblastoma of the soft tissue”. The paper stated that an important clinical feature was the old male predilection [male:female=9:1 and old age range, 45-67 years (median, 53 years)]. However, our case revealed an unusual presentation that occurred in a young girl (Table 1). The site distribution in the above study disclosed a striking predilection along the embryonic milk-line with five cases in the inguinal/groin area, and one case each in posterior vaginal wall, buttock, anterior abdominal wall and mid-back. The tumor sizes ranged from 2 to 13 cm (median: 6 cm), and all the lesions were well circumscribed. Most of the cases had a component of adipose tissue, which occupied 10-60% of tumor volumes. Focal cellular atypia and multinucleation were also described, and one case showed a tumor with some resemblance to pleomorphic lipoma.¹³ Our case had no adipose tissue component, but the immunohistochemical profile was compatible with myofibroblastoma of the mammary type.

When soft tissue tumors show a prominent epithelioid component with cellular clustering and intercellular lumina, as was seen in the present case, and when they occur in the chest wall, the possible differential diagnoses include epithelioid myofibroblastoma, myoepithelioma, epithelioid hemangiioendothelioma, epithelioid smooth muscle tumor, alveolar rhabdomyosarcoma

and lobular carcinoma of the breast. The characteristic immunohistochemical findings, including positive immunoreactivity for CD34 and desmin with cytokeratin negativity, are diagnostic for mammary-type myofibroblastoma.

On the electron microscope study, the tumor cells of the current case disclosed abundant rough endoplasmic reticula, focal basal lamina and densities, which were the usual features of myofibroblastomas. However, the well-developed cell junctions were a very unusual feature for myofibroblastoma. Because the previous electron microscopic studies¹⁴ on myofibroblastoma were based on the classic spindle cell type of myofibroblasts, the significance of the well-developed cell junctions in an epithelioid variant, as seen in this case, awaits further study.

Although a pure or predominating epithelioid myofibroblastoma is an unusual occurrence in the soft tissue, the possibility of myofibroblastoma should be raised when we perform a proper immunohistochemical study.

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