

Myxoinflammatory Fibroblastic Sarcoma in the Chest Wall

Kyoung Shik Narm, M.D.¹, In Kyu Park, M.D., Ph.D.², Mi Kyung Bae, M.D.¹, Gi Jeong Kim, M.D.³

Myxoinflammatory fibroblastic sarcoma (MIFS) is a recently defined rare tumor. It is mainly found in the upper and lower extremities of adults. Due to its high local recurrence rate and low metastatic rate, it is classified as a low grade-malignancy. Accurate diagnosis and early, wide excision are important for prognosis. Herein, we report a case of MIFS in a 35-year-old male patient that presented in an unusual location, the left chest wall. To our knowledge, this is the first reported case of MIFS in Korea and the second case to be reported within the global scientific literature involving the chest wall.

Key words: 1. Chest wall
2. Sarcoma

CASE REPORT

A 35-year-old man visited our hospital presenting with a palpable mass on his left lateral chest wall, which was movable, rubbery, and non-tender. A gun biopsy assisted by ultrasonography was performed, but failed to confirm a specific diagnosis; therefore, we performed a local excision. The mass was 3×2×1 cm, non-adhesive, round, and pink; the cut sections evidenced well-demarcated, gray-white gelatinous masses. A microscopic examination revealed a tumor in the subcutaneous tissues without invasion to the adjacent structures (Fig. 1). The specimen exhibited a mixture of myxoid, hyaline, and inflammatory zones (Fig. 2). Scattered and enlarged tumor cells, each with a large eosinophilic nucleus resembling a virocyte, were observed, consistent with a diagnosis of myxoinflammatory fibroblastic sarcoma (MIFS) (Fig. 3A, B). An immunohistochemical study revealed that the tumor cells had a strong positivity for vimentin and focal positivity for

CD34 and CD68, confirming the diagnosis of MIFS.

A microscopic histological analysis of the specimen revealed that the mass was abutting to the resection margin in a series of multifocal areas. Thus, an additional wide excision was advised for an adequate tumor-free margin. On combined positron emission tomography and computed tomography, fluoro-2-deoxy-D-glucose uptake was increased at the left chest wall, probably due to a previous procedure; multiple nodules were also observed in the right lung, suggesting possible lung metastasis.

After administering general anesthesia and single-lung ventilation, a wedge resection of the right upper lung, including the nodules, was performed under video-assisted thoracoscopic surgery conditions. The frozen sectional diagnosis of the lung nodules confirmed a benign pneumocyte hyperplasia. Subsequently, a wide left chest wall excision was performed, including partial resections of the 5th, 6th, and 7th ribs. The tumor free margin of the subcutaneous and muscle layers was

¹Department of Thoracic and Cardiovascular Surgery, Yonsei University College of Medicine, ²Department of Thoracic and Cardiovascular Surgery, Seoul National University Hospital, Seoul National University College of Medicine, ³Department of Pathology, Yonsei University College of Medicine

Received: July 19, 2011, Revised: August 24, 2011, Accepted: September 7, 2011

Corresponding author: In Kyu Park, Department of Thoracic and Cardiovascular Surgery, Seoul National University Hospital, Seoul National University College of Medicine, 101 Daehak-ro, Jongno-gu, Seoul 110-744, Korea
(Tel) 82-2-2072-2342 (Fax) 82-2-764-3664 (E-mail) ikpark@snu.ac.kr

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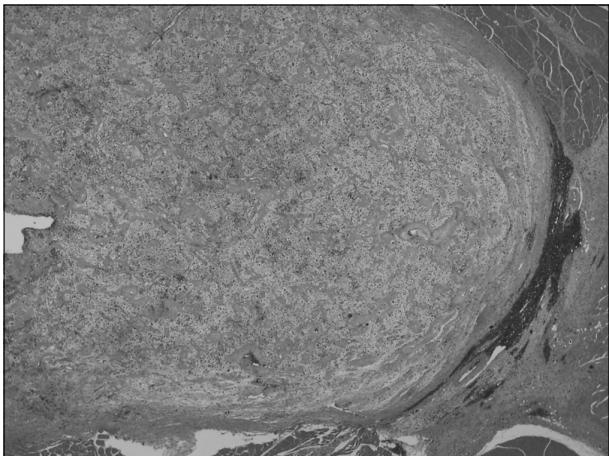


Fig. 1. A lower-power view of the inflammatory myxohyaline tumor illustrating a mixture of myxoid, hyaline, and inflammatory zones (H&E, $\times 12$).

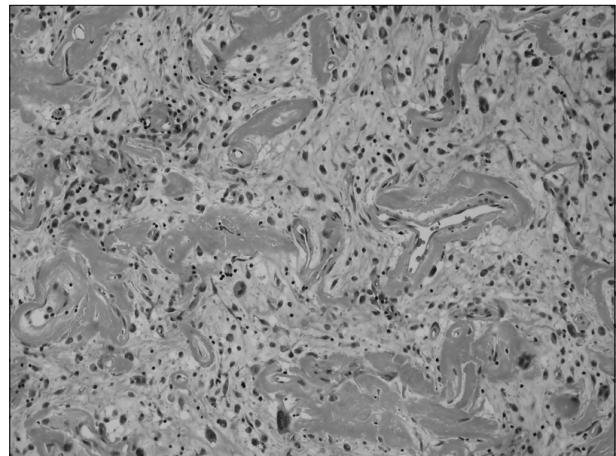
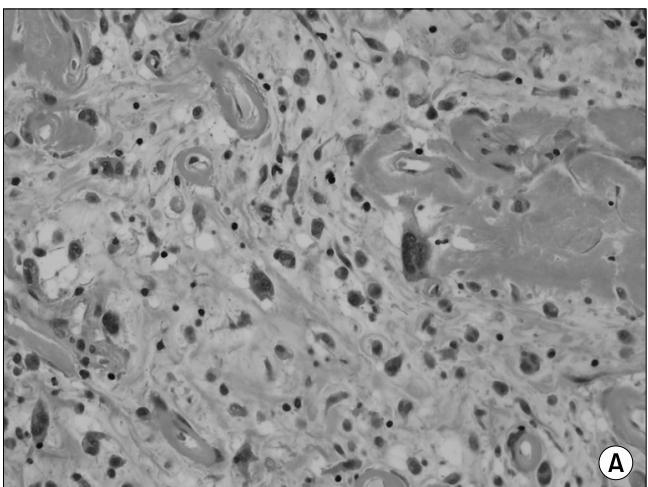
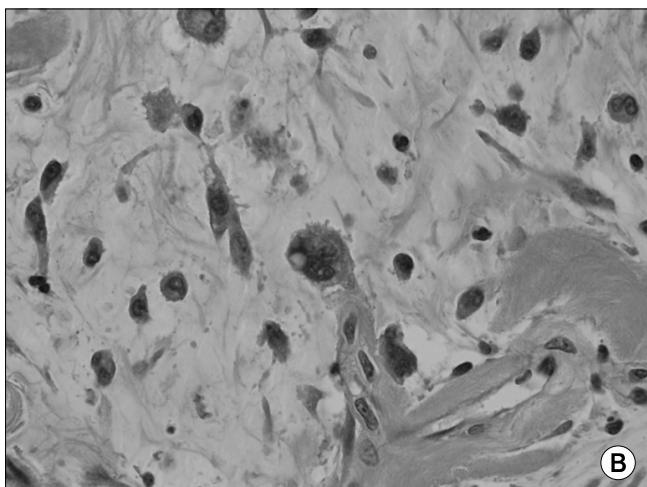


Fig. 2. An admixture of myxoid, hyaline, and inflammatory zones in an inflammatory myxohyaline tumor. Note the transition between myxoid and hyaline zones (H&E, $\times 100$).



A



B

Fig. 3. (A) A higher-magnification view of cells with smudgy nuclei in a myxoid zone of an inflammatory myxohyaline tumor (H&E, $\times 200$). (B) An enlarged tumor cell with a large eosinophilic nucleus resembling a virocyte in an inflammatory myxohyaline tumor (H&E, $\times 400$).

4 cm, and the skin margin was 2 cm. After the procedure, there was no gross evidence of a residual tumor. The chest wall was reconstructed with polypropylene mesh (Marlex, CR Bard Inc., Billerica, MA, USA) and bone cement. The patient was discharged on postoperative day 10.

DISCUSSION

MIFS, or acral MIFS, is a low-grade, malignant tumor that has only recently been defined pathologically. About 200 cas-

es have been reported since it was first described in 1998 by Montgomery et al. [1].

In 80% of cases, the lesion involves the subcutaneous tissues of the hands or feet [2], but it can be also be found on the elbows, thighs, or shoulders; thus the term "acral" was dropped in the 2002 World Health Organization classification [3].

Clinically, MIFS usually presents as a slow-growing, solitary, and painless mass that can sometimes infiltrate extensively into the surrounding soft tissues. Grossly, it is usu-

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ally visible as an ill-defined, multinodular mass, with myxoid areas, within the subcutaneous tissue, and has an irregular border and invasive margin [1,4]. Necrosis or hemorrhage may also occur. The tumors are 1–8 cm in size and discharge a clear, jelly-like substance when compressed [4].

The simple X-ray imaging of the affected area did not provide any specific findings. A magnetic resonance imaging also provided no pathognomonic patterns because of the mass' varied histological composition; however, the images did exhibit low signal intensity on the T1-weighted image and high signal intensity on the T2-weighted image.

Histologically, the mass was composed of dense, mixed acute and chronic inflammatory cells, and the hypocellular area was composed of hyaline and myxoid zones. Characteristic ganglion-like, lipoblast-like, and polygonal spindle cells that contained bizarrely shaped nuclei and inclusion-like nucleoli were present in an abundant eosinophilic cytoplasm. These cells were derived from the modified fibroblast cells and chronic inflammatory cells. A mitotic division was generally rare, and necrosis was often evident. Upon the immunohistochemical study, vimentin was always positive, CD68 and CD34 exhibited variable positivity, and HMB45 and CD45 were always negative [4].

The differential diagnosis can be broad and varied depending on whether the inflammatory, myxoid, or bizarre atypical component predominates within the lesion. Tenosynovitis, ganglion cyst, and myxoma (for the inflammatory and benign entities), and myxoid malignant fibrous histiocytoma, myxoid liposarcoma, and epithelioid sarcoma (for the malignant tumors) are the main components of the histopathological differential diagnosis of a giant cell tumor in the tendon sheath or of a myxoid origin [2,5]. Because of the difficulty of an accurate diagnosis from the lesion's clinical presentation or radiological characteristics, the diagnosis is often delayed and a surgical excision is more likely to be suboptimal.

The local recurrence rate of the tumor is high (range, 22% to 67%) when a local excision alone has been performed, usually being reported between 3 months and 5 years post-operatively [4]. The distant metastasis rate is low (6.5%); however, cases have reported metastasis to the lung, neck, and skull [6,7]. One tumor-related death has been reported [6].

Although the treatment process has not been well-defined, a complete excision with clear margins is recommended. Postoperative radiotherapy may be a good additional therapy for patients who have a large tumor or a positive resection margin [2,8]. Some papers also report chemotherapy as an option [4].

There has only been one other reported case of MIFS involving the chest wall. Premalata et al. [5] reported upon a MIFS of the upper back, over the scapula. They performed a wide excision of the tumor, without rib resection, and reported no recurrence within 10 months after surgery. We performed a wider local excision with a tumor free margin of 4 cm, including an adjacent rib resection and the reconstruction of the chest wall, in accordance with the guidelines for a malignant tumor of the chest wall.

In summary, MIFS is a very rare tumor that predominantly affects the distal extremities. It has not been previously described in the Korean literature. We report the first case of MIFS in Korea, and the second such tumor to be described in the chest wall. The patient underwent a wide local excision and a chest wall reconstruction.

REFERENCES

- Montgomery EA, Devaney KO, Giordano TJ, Weiss SW. *Inflammatory myxohyaline tumor of distal extremities with virocite or Reed-Sternberg-like cells: a distinctive lesion with features simulating inflammatory conditions, Hodgkin's disease, and various sarcomas*. Mod Pathol 1998;11:384-91.
- Cordoba-Fernandez A, Juarez-Jimenez JM, Mazuecos-Blanca J, Illanes-Moreno M. *A report of a rare case of myxoinflammatory fibroblastic sarcoma*. J Am Podiatr Med Assoc 2010;100:497-501.
- Kindblom LG, Meis-Kindblom JM. *Myxoinflammatory fibroblastic sarcoma*. In: Fletcher CD, Unni KK, Mertens F, eds. *Pathology and genetics of tumors of the soft tissue and bone*. World Health Organization classification of tumours; vol. 4. Lyon: IARC Press. 2002. 96-7.
- Meis-Kindblom JM, Kindblom LG. *Acral myxoinflammatory fibroblastic sarcoma: a low-grade tumor of the hands and feet*. Am J Surg Pathol 1998;22:911-24.
- Premalata CS, Rama Rao C, Padma M, Vijaykumar M. *Myxoinflammatory fibroblastic sarcoma: report of a rare case at an unusual site with review of the literature*. Int J Dermatol 2008;47:68-71.
- Hassanein AM, Atkinson SP, Al-Quran SZ, Jain SM, Reith

- JD. *Acral myxoinflammatory fibroblastic sarcomas: are they all low-grade neoplasms?* J Cutan Pathol 2008;35:186-91.
7. Sakaki M, Hirokawa M, Wakatsuki S, et al. *Acral myxoinflammatory fibroblastic sarcoma: a report of five cases and review of the literature.* Virchows Arch 2003;442:25-30.
8. Tejwani A, Kobayashi W, Chen YL, et al. *Management of acral myxoinflammatory fibroblastic sarcoma.* Cancer 2010; 116:5733-9.