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Case Report / Olgu Sunumu

Low-grade fibromyxoid sarcoma in the mediastinum: a case report

Mediastinumda düşük dereceli fibromiksoid sarkom: Olgu sunumu

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Low-grade fibromyxoid sarcoma is a rare type of soft tissue tumor. A 25-year-old female patient was admitted to our clinic with the complaints of back pain and dyspnea. Thorax computed tomography revealed a mass lesion of 17x13x11 cm in its largest diameter in the posterior mediastinum. Total mass excision was performed through right thoracotomy. Histopathological examination revealed low-grade fibromyxoid sarcoma. Although low-grade fibromyxoid sarcoma rarely involves mediastinum, it should be considered in the presence of such mass.

Key words: Mediastinal neoplasm; mediastinum, sarcoma.

Düşük dereceli fibromiksoid sarkom, nadir görülen bir yumuşak doku tümörüdür. Yirmi beş yaşında kadın hasta sırt ağrısı ve nefes darlığı yakınmaları ile kliniğimize başvurdu. Toraks bilgisayarlı tomografide posterior mediastinumda, en geniş boyutları yaklaşık 17x13x11 cm çaplı kitle lezyonu saptandı. Sağ torakotomi ile total kitle eksizyonu yapıldı. Histopatolojik incelemede düşük dereceli fibromiksoid sarkom saptandı. Mediastinum, her ne kadar düşük dereceli fibromiksoid sarkomun nadir yerleşim yeri olsa da bu gibi kitle varlığında akla getirilmelidir.

Anahtar sözcükler: Mediastinal tümör; mediastinum; sarkom.

In 1987, Evans was the first author to label two different cases as having soft tissue tumors that appeared benign in nature but could metastasize. They were known as so-called "low-grade fibromyxoid sarcoma" (LGFMS).[1] It may be located in any of the locations where other soft tissue sarcomas have been described.[2] However, only two cases with mediastinal location have been reported to date.[3,4] This report presents a third case of LGFMS in the mediastinum.

CASE REPORT

A 25-year-old female patient was admitted with complaints of back pain and dyspnea. On computer tomography (Figure 1a), a lesion of 17x13x11 cm in diameter at its largest section was observed in the posterior mediastinum. In the fiberoptic bronchoscopy, the right main bronchus appeared to be narrower, starting from the carina, due to anterior compression. Because the diagnosis could not be established through non-invasive methods, the operation was started with a mediastinoscopy. Multiple biopsies were obtained from the lesion in the precarinal area. The frozen section analysis revealed a mesenchymal tumor; thus, a right thoracotomy was planned. In the explorative intervention, it was observed that a giant, lobulated, cream- colored, capsulated mass filled up the entire posterior mediastinum where it compressed on the anterior mediastinal formations. It extended to the hilum from over the diaphragm, ran under the azigos vein and the bronchial carina, and extended to the anterior mediastinum and opposite hemithorax. The mass was totally excised. The patient did not develop any postoperative complications and was discharged on the 11th day. In light of the postoperative medical and radiation oncology consultations, adjuvant chemotherapy or radiotherapy was not considered.

In the pathological evaluation, the sections were cream-to-yellow in color and bright, hard, and solid in structure. There were cystic degenerations in some areas. Microscopically, a tumor lesion formed by spindle-shaped cells was visible in the myxoid and hyalinized stroma. The tumor cells were formed by fascicules and whirlpools of cells which were slightly nuclear and spindle- and oval-shaped. The cells contained intranuclear structures at focal areas and were rarely mitotic. "Giant collagen rosettes" formed by the tumor

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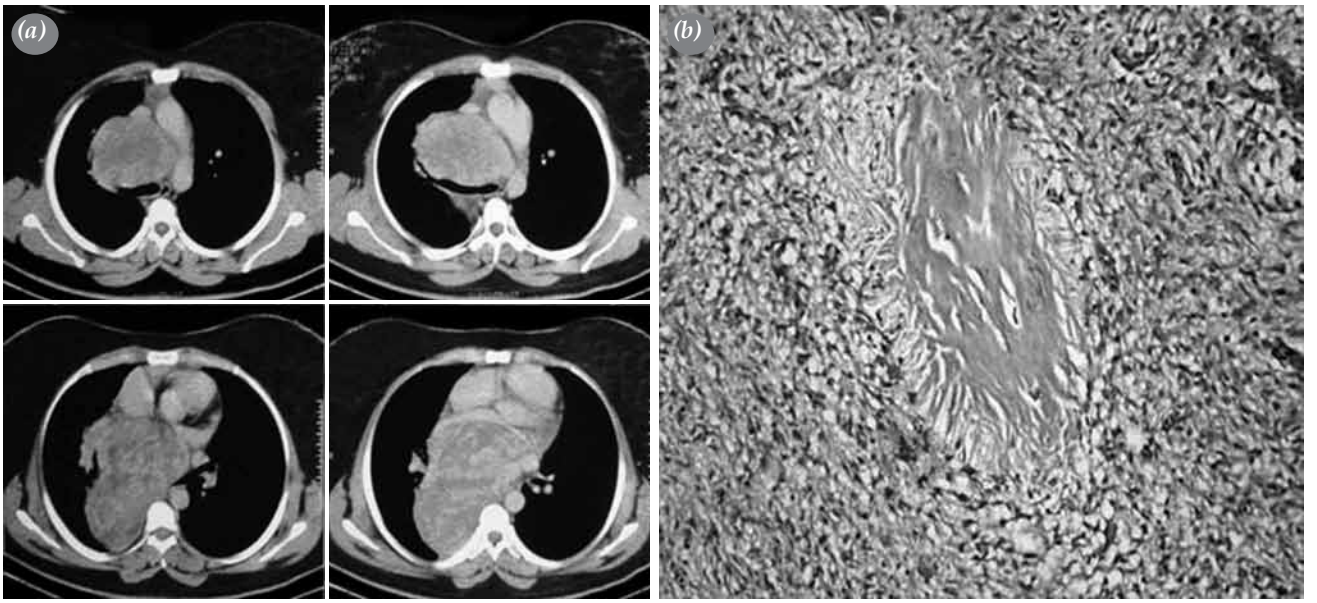

Figure 1. On the thorax computed tomography image, a lesion of nearly 17x13x11 cm in diameter at its largest section was observed in the posterior mediastinum. **(a)** The lesion compressed the neighboring bronchial and mediastinal structures and extended to the anterior mediastinum and **(b)** microscopically giant collagen rosette is seen (H-E x 200).

cells that surrounded the collagen nodules in myxoid areas were striking (Figure 1b). There were wide areas of hyaline degeneration in the stroma, but no necrosis was detected. In the immunohistochemical evaluation, the neoplastic cells were diffusely and markedly positively stained with vimentin. There was no staining for antibodies to S100 protein, the CD34 human hematopoietic progenitor cell antigen, the CD99 protein antigen, calretinin, cytokerotin (CK)7, the epithelial membrane antigen (EMA), pankeratin, actin, desmin, or neuron-specific enolase (NSE). However, the tumor was immunoreactive to the CD57 protein antigen and factor XIIIA. The neoplastic cells were not stained with anti-Ki-67 antibody. According the morphological and immunohistochemical findings, a diagnosis of LGFMS (Hyalinizing spindle-shaped cell tumor with giant rosettes) was established.

DISCUSSION

Low-grade fibromyxoid sarcoma is a rare type of soft tissue neoplasm found in young and middle-aged adults.[5] Most frequently, these lesions are located in the deep soft tissues of the lower extremities and thighs. They are situated less frequently in the chest wall, axilla, shoulder area, inguinal area, hip and neck.[3]

Low-grade fibromyxoid sarcoma is usually macroscopically well-contoured but presents microscopic infiltrations to adjacent soft tissues. The tumor sections have bright yellow-white color in focal areas due to myxoid matter accumulation. No necrosis or hemorrhage exists.[3,5] Microscopically, the tumor is characterized by whirlpool and focal linear formations of spindle-shaped cells. The myxoid zone may extend into an immediate fibrous zone, or there may be a step-wise passage between the zones. The curved or branched net, which often dominates the myxoid zone, is composed of blood veins of capillary size.[3] These macroscopic and microscopic findings are similar to the histopathological findings in our case.

In differential diagnosis of LGFMS, benign soft tissue tumors, such as desmoid fibromatosis, neurofibroma, and fasciitis with myxomatous degeneration, should be kept in mind.[3,5,6] Although LGFMS is deceptively benign in appearance, it has a high local recurrence rate, and, in a majority of patients, it recurs frequently and metastasizes into the lungs. Local recurrence has been reported in nearly 65% of LGFMS patients. In the literature, the recurrence has been reported to occur as early as six months after the excision of the first lesion and as late as 50 years after the excision.[5]

Low-grade fibromyxoid sarcoma is usually seen in young adults in the form of large masses characterized by fibrous, myxoid areas and may be located in the mediastinum. The treatment involves the excision of the primary lesion, recurrent lesions, or metastases.[3,4,6] Close, long-term follow-up is essential.[3]

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