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(CASE REPORT)

A case of myxofibrosarcoma located on the chest wall

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Abstract

Myxofibrosarcoma is a variant of fibrosarcoma, a tumor consisting of malignant fibroblasts, exhibiting a variable myxoid stroma, pleomorphism and wide curved vascular pattern. The most common location site of the tumor is the extremities. Uncommonly it is located in the trunk, head and neck. Herein we present a very rare case of surgically treated chest-wall-myxofibrosarcoma.

Keywords: Myxofibrosarcoma; Chest wall; Surgical treatment

1. Introduction

Chest wall tumors can originate from bone, cartilage and soft tissue, and consist about 5% of all thoracic neoplasms. Soft tissue sarcomas, chondrosarcomas, fibrosarcomas and Ewing's sarcomas are among the most common malignant tumors of the chest wall. Myxofibrosarcoma is a variant of fibrosarcoma, a tumor consisting of malignant fibroblasts, exhibiting variable myxoid stroma, pleomorphism and large curved vascular pattern [1]. More commonly encountered during the 6th to 8th decades of life with a slight predominance of male and uncommonly detected under the age of 20. The most common site of the tumor is the extremities, whereas they are more rarely located in the trunk, head and neck. Patients often present with a complaint of slow growing, painless mass. Whilst superficially located neoplasms appear as multiple gelatinous or stiff nodules, those that are deeply located generally present as single infiltrative masses [1]. Herein, a case of myxofibrosarcoma with its very unusual location on the chest wall is presented in the light of pertinent literature.

2. Case Report

A 79-year-old male patient admitted with a complaint of swelling present on his chest for two months. He had a history of atrial fibrillation, hypertension and prostate adenocarcinoma. On physical examination, a palpable soft mass was observed in the pectoral region of the right hemithorax. Computed tomography (CT) (Figure-1a) and thoracic magnetic resonance imaging (MRI) (Figure-1b) of the thorax revealed a soft tissue mass in the right pectoral muscle of the with a craniocaudal length of 9 cm and having the transverse dimensions of 14 X 7 cm in its widest part. The mass had heterogeneous hyperintense signal feature on T2A image.

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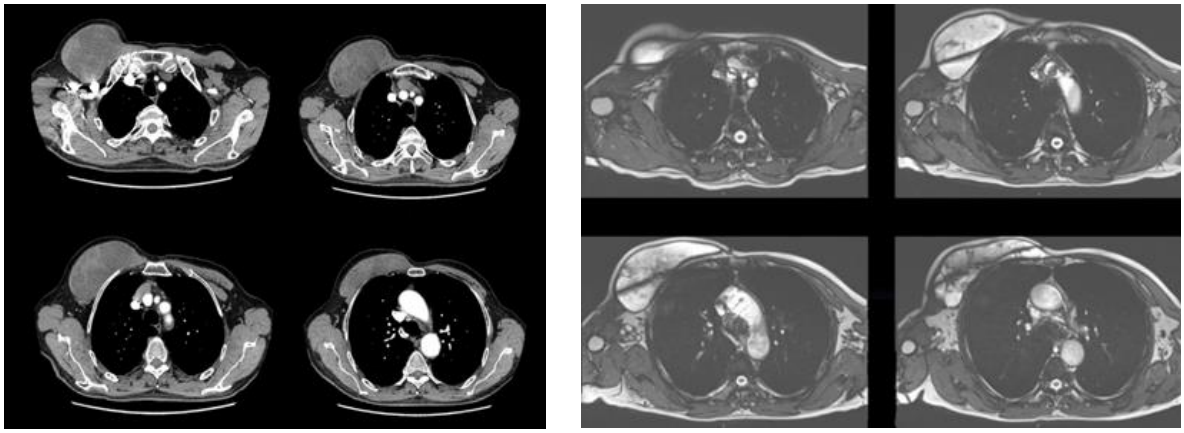

Figure 1 a. Mass image of the right hemithorax anterior chest wall in thorax CT. b. Mass image located in the anterior chest wall of the right hemithorax on thorax MR T2A images.

After preoperative preparations were completed, the patient was operated under general anesthesia. Above the most prominent region of the tumor, a 10-cm horizontal incision (Figure-2a) was made. The tumor was encapsulated, containing jelly-like material (Figure-2b) between muscle plans and was totally excised with a safety distance from the margins.

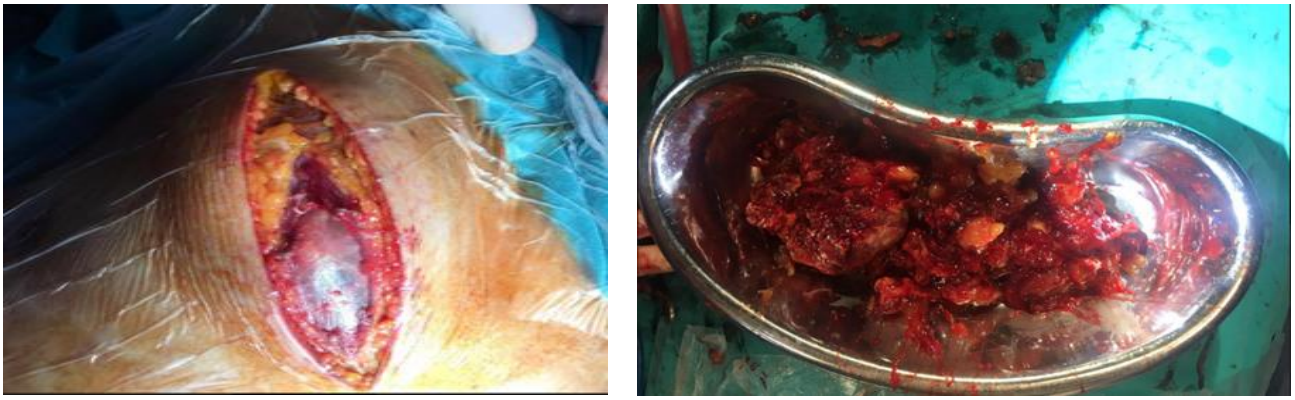

Figure 2 a. Horizontal incision. b. Encapsulated, containing jelly-like material

Pathological examination of the specimen revealed grade 2-3 myxofibrosarcoma (Figure-3).

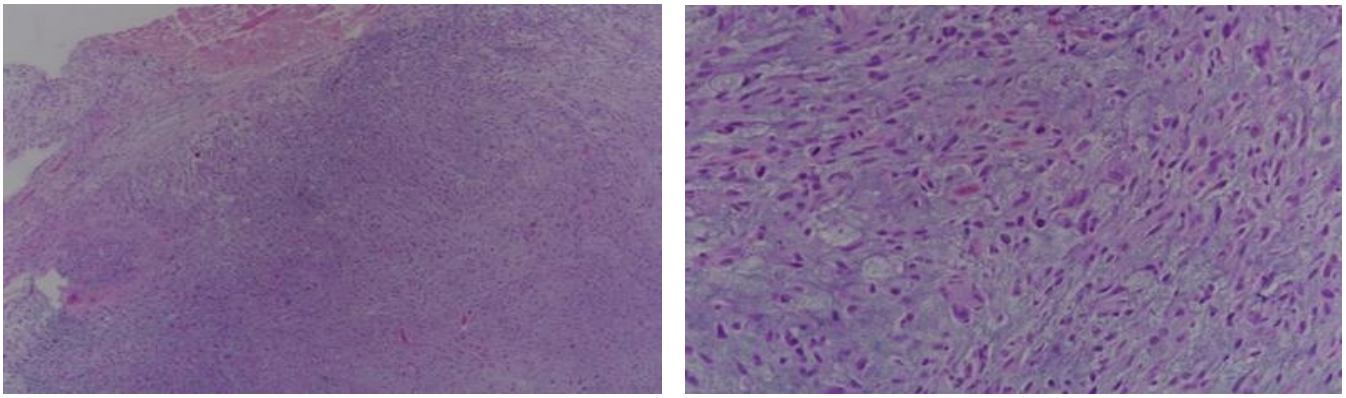

Figure 3 a. Myxofibrosarcoma consisting of pleomorphic cells on myxoid floor (H&E x40). b. Numerous mitotic figures are observed on cellular floor in myxofibrosarcoma (H&E x200).

After an uneventful postoperative period, the patient was discharged by cure. During the follow-up period, an appearance compatible with recurrence was observed in the operation area 3 months later. Thereupon, the patient was taken to a joint operation with the Orthopedics Clinic. An enlarged chest wall resection was performed, including partial resection of 4th rib and total resection of 1st, 2nd and 3rd ribs. Then, chest wall reconstruction with aortic graft was performed. The result of the pathological examination of the surgical material was reported as "Recurrence of Myxofibrosarcoma, grade 3".

Written informed consent was obtained from the patient for publication of his data.

3. Discussion

Myxofibrosarcoma is more commonly encountered in older men and patients often admit to the hospital due to slow-growing, painless mass which is located in the extremities [1]. It has been reported that myxofibrosarcoma is located in extremities with a percentage of 77%, trunk with 12%, mediastinum or retroperitoneum with 8% and head with 3% [2]. In the present case, myxofibrosarcoma was located on the chest wall, which is one of the very rare site for the development of the condition. There are quite a limited number of data in the literature regarding myxofibrosarcoma cases located on the chest wall. These data are limited to a few case reports and small series [3-5]. An electronic search of the literature in PubMed revealed only 3 publications by giving the “myxofibrosarcoma” and “chest wall” keywords for the title.

McMillan et al. reported 22 myxofibrosarcoma cases in a study of 192 patients in which they investigated the factors affecting the recurrence after resection in chest wall soft tissue sarcomas, while Kachroo et al. reported 1 myxofibrosarcoma case in a series of 51 patients in which they shared their surgical experience in primary chest wall sarcomas [3,4]. Klingbeil et al. presented a case of myxofibrosarcoma that developed after trauma and located in the chest wall [5]. In our case, there is no trauma history. This tumor, which appears regularly in subcutaneous tissues [6], less frequently seen in deeper tissues such as in our case. In patients with myxofibrosarcoma, local recurrence tends to occur in 50-60% of cases, irrespective of histological grade. Metastasis and tumor-related mortality are closely linked with tumor grade. While none of the low grade metastases metastasizes, in intermediate and high grade neoplasms, metastasis can develop in 20-35% of the cases [1]. In these group of patients, who can be encountered frequently with recurrent relapses, larger limited resection constitutes the basis of treatment. If possible, the limits of resection should be at least 4 cm from the macroscopic border of the primary tumor. It is suggested that postoperative chemotherapy and/or radiotherapy are associated with better survival outcomes.

4. Conclusion

Myxofibrosarcoma may rarely be localized in the trunk, head and neck. This tumor, which is frequently encountered with recurrent relapses, may become higher grade with recurrences. In these group of patients, larger limited resection constitutes the basis of treatment.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Statement of ethical approval

The procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Declaration of Helsinki.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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