CASE REPORT

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Myxofibrosarcoma of The Abdominal Wall at Moewardi Hospital, Surakarta: A Rare Case Report

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ABSTRACT

Introduction: Myxofibrosarcoma (MFS) is a fibroblast-derived sarcoma, which accounts for approximately 5–10% of all soft tissue malignant tumors. The mean age in patients with MFS is between the fifth and seventh decades. Around 77% of MFS cases occur in the upper extremities. Other areas of the body including the trunk (12%), retroperitoneum or mediastinum (8%), abdominal wall, and heart have also been reported.

Case Presentation: A 57-year-old Indonesian female visited our center with complaints of a lump in the left lower abdomen for 4 months. The lump has grown rapidly in the last two months. A contrast abdominal MRI examination found a soft tissue mass on the left lumbar abdominal wall to the left iliac with a size of 15 x 11,4 x 13,4 cm and bordered by part of the left external oblique muscle and the left internal oblique muscle pushing the intestinal system medially. The biopsy of the mass revealed a high-grade myxofibrosarcoma. The patient underwent a wide excision with an elliptical incision design, with in-toto removal of the lump. The immunohistochemical result of the tumor showed high-grade myxofibrosarcoma with free margins at the incision margin. Follow-up was performed at 1 month, 6 months, and 12 months postoperatively. The patient did not receive radiotherapy.

Conclusions: This report describes a rare case of MFS of the abdominal wall. Recognizing the histopathological features of MFS and applying the appropriate immunohistochemical examinations are crucial in establishing the correct diagnosis. This case provides a diagnosis and treatment experience of MFS that occurs in the abdominal wall.

INTRODUCTION

The World Health Organization (WHO) classifies sarcomas into two main categories: soft tissue sarcoma (STS) and bone sarcoma [1]. STS is one of the top 10 cancer cases in Indonesia. STS can appear at any age and affect any location on the body. The incidence of STS is less than 1% of all malignant tumors. The estimated number of new cases of STS in the United States in 2016 was 12,310, and the estimated number of deaths was 4,990 [2]. Approximately 60% of soft tissue sarcomas affect the extremities (most commonly the thighs), 19% of the trunk wall, 15% of the retroperitoneum, and 9% of the head and neck [3]. Histologically, the most common type of sarcoma was malignant fibrous histiocytoma (MFH) (28%).

Myxofibrosarcoma (MFS) is a connective tissue neoplasm and a rare form of sarcoma. It is about 5–10% of all soft tissue malignant tumors are myxofibrosarcomas (MFS), which are fibroblast-derived sarcomas. Patients with MFS range from fifth to seventh decades in age on average. The upper extremities account for about 77% of MFS patients. There have also been reports of the heart, abdominal wall, retroperitoneum or mediastinum, and the trunk (12%), among other bodily parts [4]. MFS involves the dermis or subcutaneous tissue in over two-thirds of cases. Tumors are divided into three grades, with grade I being locally aggressive and grades II and III showing metastatic potential as well as having more complex cytogenic aberrations [5].

CASE PRESENTATION

A 57-year-old Indonesian female visited our center with complaints of a lump in the left lower abdomen for 4 months before being admitted to the hospital. The lump has grown rapidly in the last two months. The skin was intact, and she denied pain. Physical examination

revealed a lump in the left lumbar region, tenderness (-), venectation (+), reactive zone (+), Carnett's test (-), solid (+), mobile, and no enlarged inguinal lymph nodes (**Figure 1**). A contrast abdominal magnetic resonance imaging (MRI) examination found a soft tissue mass on the left lumbar abdominal wall to the left iliac with an approximate size of 15 x 11,4 x 13,4 cm, bordered by part of the left external oblique muscle and the left internal oblique muscle pushing the intestinal system medially. No lymph node metastases were found in the MRI (**Figure 2**).

The biopsy of the mass, performed in our hospital, revealed high-grade myxofibrosarcoma (**Figure 3**). A wide excision was performed with an elliptical incision design, with in-toto removal of the lump. A resected lump measured 20 x 15 x 10 cm with a margin of 2 cm (**Figure 4**). The tumor was above the fascia, not infiltrating the left external oblique abdominis muscle. Postoperative defects can be treated with primary sutures without reconstruction. The histopathological results of the tumor showed a high grade with free margins at the incision margin. Follow-up was performed at 1 month, 6 months, and 12 months postoperatively with good condition, and no relapses or metastases were found. The patient did not receive radiotherapy.

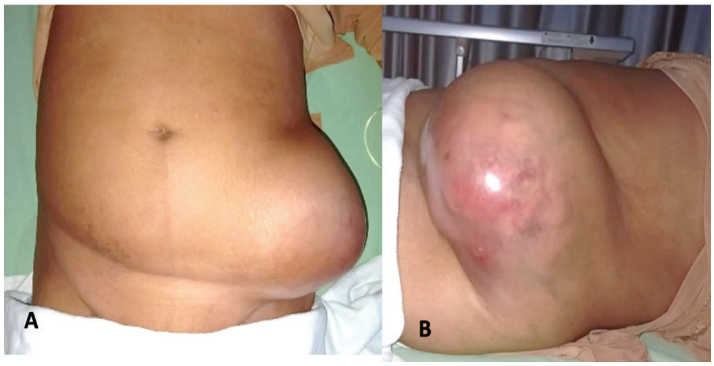


Figure 1. (A) Anterior view; (B) Lateral view. There was a lump in the left lumbar region with venectation (+), reactive zone (+), and Carnett's sign (-) which is solid and mobile.

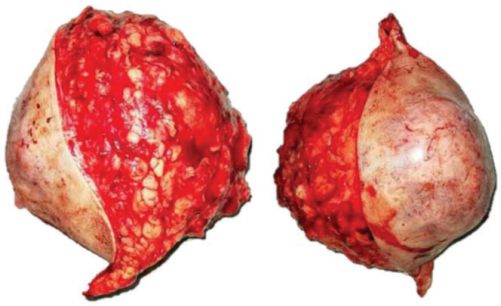


Figure 4. The resected lump, measuring 20 x 15 x 10 cm

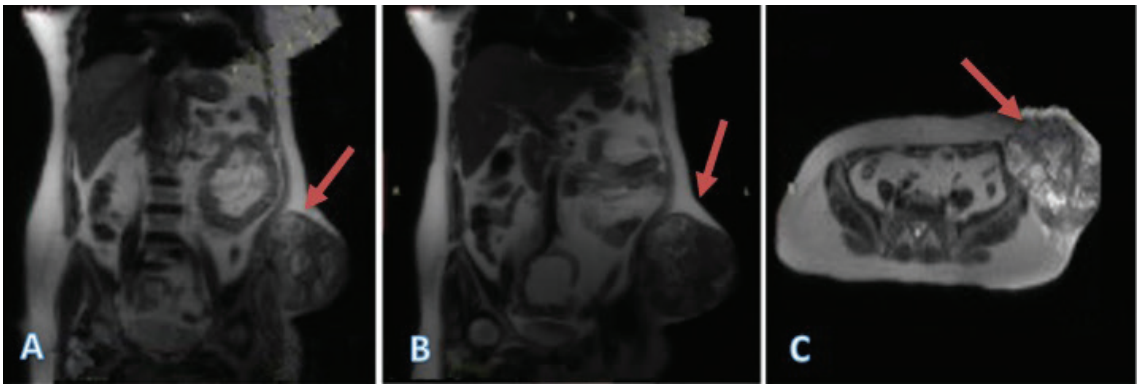
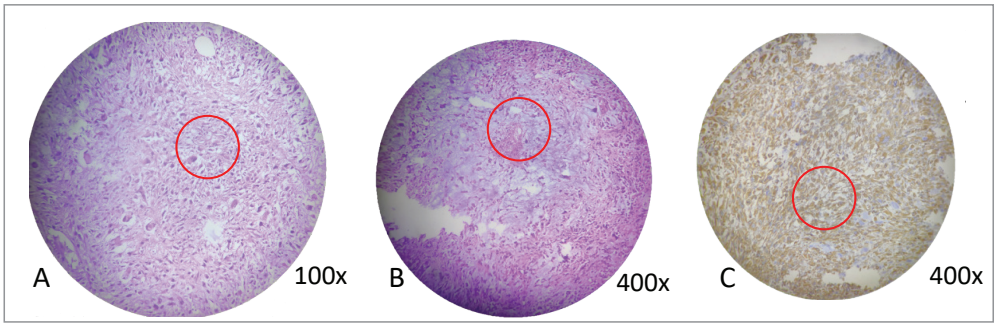


Figure 2. (A) and (B) Coronal section; (C) Axial section. On contrast abdominal MRI, a soft tissue mass was found on the left abdominal wall from the left lumbar to the left iliac region bordered by a portion of the left external oblique muscle and the left internal oblique muscle pushing the intestinal system medially. No invasion to intraperitoneal. No lymph node metastases were found.

Figure 3. Myxoid istopathology.
(A) and (B) Mesenchymal tumor, extensive necrosis, and myxoid. Some cells are pleomorphic, large cells. Some cells are lipoblasts; (C) Immunohistochemical examination: vimentin (positive diffuse in tumor cells), according to myxofibrosarcoma.



DISCUSSION

Sarcoma is divided into two main parts: soft tissue sarcoma (STS) and bone sarcoma. In STS that occurs in adults, the most common histopathological types are liposarcoma, leiomyosarcoma, and undifferentiated pleomorphic sarcoma [2]. Based on the records of Moewardi Hospital Surakarta in 2021, there were 134 clinical cases and 175 anatomical pathology cases of sarcoma, and myxofibrosarcoma was only 3% of all sarcoma cases.

Myxofibrosarcoma, known as myxoid MFH, is a cohesive, distinct, and identifiable form despite having a broad histological spectrum [7]. Clinically, MFS has a higher rate of local recurrence and a lower rate of distant metastasis when compared to other forms of sarcoma [8]. According to their cell density, cytopleomorphism, and frequency of mitotic images, tumors are typically divided into four malignancy grades during pathological examination [9]. Grade I and II tumors in MFS are characterized by a clear atypia; fibroblast-like cells predominate, while histiocytic cells dominate grade III and IV tumors [10]. MFS exhibits a polynodular development pattern with alternate hypocellular and myxoid zones. It also has multicellular fibrous patches and long, curving blood vessels in different mucinous stromal 20–70% of instances involve tumors, which can be shallow or deep and typically develop beneath or near the skin [11]. For fibrosarcoma, imaging examinations such as computed tomography (CT), MRI, and positron emission tomography (PET)-CT are frequently used. These tests also have significant clinical significance. MRI is regarded as an essential examination technique for the identification of soft tissue tumors, including myxofibrosarcoma, because of its high resolution, which makes it possible to depict the location, size, shape, and range of tumor invasion as well as its pathological components [7].

In this case report, a progressively enlarged tumor mass was found; clinically, an operable abdominal mass was found, and an accurate diagnosis is required. A supportive examination in the form of a contrast abdominal MRI was carried out to see the boundaries of the tumor. In this case, a soft tissue mass was found on the left lumbar abdominal wall to the left iliac region that partially infiltrated the left external oblique abdominis muscle, bordered by the left internal oblique abdominis that forces the intestinal system medially. The resected lump was measured 20 x 15 x 10 cm with a margin of 2 cm. No lymph node metastases were found. The biopsy of the mass showed high-grade myxofibrosarcoma. Abdominal wall sarcoma surgery involves two stages. First, the tumor must be resected, and second, the abdominal wall needs to be reconstructed. For tumor removal, there is no consensus on the width of surrounding normal tissue that should be removed to improve outcomes [4].

Management of fibrous myxoids requires handling by various disciplines, including surgeons, oncologists, and radiotherapists. The diagnosis of this disease is mainly made through histopathological examination. Wide excision is the standard therapy locally. In general, when surgery is performed, a wide excision should be performed with a margin of 2 cm from the tumor margin, and the tumor should not remain at the incision margin [10]. Postoperative radiotherapy may be performed if tumor-free margins cannot be obtained at surgery [11]. In this case report, a wide excision was performed with a margin of 2 cm. The tumor was still limited to the fascia, not infiltrating the left external oblique abdominis muscle. Patients with myxofibrosarcoma have a 5-year survival rate of 60–70% [10]. Due to the recurrence of this tumor type, patients need to be monitored and followed up after surgery. This patient was followed up in the past year, and no recurrence or metastasis was found.

CONCLUSIONS

This report describes a rare case of MFS of the abdominal wall. Recognizing the histopathological features of MFS and applying the appropriate immunohistochemical examinations are crucial in establishing the correct diagnosis. This case provides a diagnosis and treatment experience of MFS that occurs in the abdominal wall.

DECLARATIONS

Competing interest

The author(s) declare no competing interest in this study.

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