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Case Report

Colon Leiomyosarcoma Diagnosed by Pleural Biopsy: A Case Study

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Abstract

Leiomyosarcomas (LMS) are rare tumors of the gastrointestinal (GI) tract with aggressive and poor prognosis. LMS metastasizes primarily to the liver but rarely to the lungs, and has hematogenous spreading from smooth muscle cells of the intestinal wall. Pulmonary LMS is encountered in one of 3000 pulmonary carcinomas. A 60-year male patient was admitted in the department of chest diseases in Meram Medical Faculty at Necmettin Erbakan University, Konya due to dyspnea, cough, fatigue, and weight loss, as well as massive pleural effusion findings on the left. His thoracentesis fluid demonstrated exudate characteristics, and cytological examination revealed lymphocytosis. On thoracic computerized tomography (CT), multiple nodules with irregular contours were observed in mediastinal pleura. Pleural biopsy was compatible with LMS. In primary focus scan, lesions showing increased fluorodeoxyglucose (FDG) involvement in the colon; (SUV max: 34.87-21.08) were seen on positron emission tomography/computed tomography (PET-CT). A 2-cm-polypoid lesion was observed on colonoscopy. Planned for chemotherapy due to the pleural metastasis of colon LMS, the patient died without receiving chemotherapy one month after the rapid impairment in general status. PET-CT is very valuable in the determination of the primary focus and the immunohistochemical examination and diagnosis of this rare malignancy of pleura. The fact that the malignant pleura with poor prognosis is diagnosed earlier will create wide treatment options and be beneficial to clinicians to know such a rare cause of exudative effusion. Based on literature, we are here reporting the case presented with massive pleural effusion due to pleural metastasis of colon LMS as the first case diagnosed with pleural biopsy at our set-up.
Key words: Massive pleural effusion, pleural metastasis, pulmonary leiomyosarcoma, thoracentesis.

Introduction

Pleural effusion is one of the common entities. Among its etiology, congestive heart failure is the most common and is followed by pneumonia and malignancy.[1] Soft tissue sarcomas originate from fibrous tissue, adipose tissue, striated and smooth muscles, vascular and nervous tissues, developing from mesenchymal cells during embryological development. Soft tissue cancers are one of the rare cancers, making up approximately 0.7% of adult cancers and 6.5% of childhood cancers.[2] Associated with poor prognosis, Leiomyosarcomas (LMS) are rare aggressive soft tissue tumors and affect 0.36/100,000 women per annum. Based on the literature, there is one pulmonary LMS per every 3000 pulmonary carcinomas.[3] LMS of the gastrointestinal (GI) tract is a tumor rarely encountered and originating from smooth muscle cells of the intestinal wall. It is often found in the stomach and accounts for less than 1% of malignant colorectal tumors.[4] It was reported that LMS was detected in the lung as multiple nodules, masses, or with the tree-in-bud pattern through different imaging methods.[5]

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Authors Contribution

CK & AZ conceptualized the project. CK, PD & HHE did the data collection and literature search. Drafting, revision & writing of manuscript were done by CK, AZ, PD & HHE.

Case Report

A 60-year-old male patient having massive pleural effusion findings and a smoking history of 60 pack/year was admitted to the Department of Chest Diseases, Meram Faculty of Medicine, Necmettin Erbakan University, Konya with complaints of cough, dyspnea, anorexia, fatigue, and weight loss. Thorax computerized tomography (CT) revealed massive pleural effusion and multiple nodular

Table 1: Immunohistochemical investigation.

Immunohistochemical Staining	*Clinical Practice*	*Effects on Patients*
Desmin	Striated and smooth muscle; leiomyosarcoma; rhabdomyosarcoma	+
Vimentin	Mesenchymal neoplasms; malignant melanoma and subsets of carcinoma and lymphoma	+
Pancytokeratin	In distinguishing epithelial and mesothelial cells from mesenchymal cells	-
TTF-1	Lung adenocarcinoma; thyroid carcinoma; a subset of small cell carcinoma	-
P63	Prostatic basal cells; breast myoepithelial cells; squamous cell carcinoma	-
Calretinin	In distinguishing mesotheliomas from lung adenocarcinomas	-
S-100	Neural tumor; melanoma; chondrocytes; epithelial neoplasm subset	-
Actin	Skeletal, smooth, and cardiac muscles	-
Cytokeratin-LMW	Epithelial neoplasm with simple epithelium	-
AFP	Hepatocellular carcinoma; yolk sac tumor	-
MART-1	Melanoma-associated marker	-
EMA	Epithelial marker; a subset of sarcoma, meningioma, and hematolymphoid neoplasm	-
Myogenin	Rhabdomyosarcoma	-
CD34	Blasts; stem cells; endothelium; GIST; soft tissue tumor subset	-
CD20	Pan B-cells	-
DOG-1	GIST in particular CD117-negative lesion	-

AFP: Alpha-fetoprotein, EMA: Epithelial membrane antigen, GIST: Gastrointestinal stromal tumor, LMW: Low molecular weight, MART-1: Melanoma-associated marker, TTF: Thyroid transcription factor

lesions with irregular contours (the largest was 3 cm in diameter). There was no pathological finding in bronchoscopy. The fluid drawn via thoracentesis was exudative. In the cytological examination of the fluid, although there was 75% lymphocyte dominance, no malignant cells were seen. In the pathological examination of the closed pleural biopsy, performed with the Cope's pleural biopsy needle, atypical cells with small solid islands in multiple parts were observed. The nucleoli of the cells were distinct, oval, round, and with pleomorphic core and pale eosinophilic cytoplasm. When these cells were immunohistochemically stained, desmin and vimentin were found positive, while other markers were seen negative (Table-1).

The Ki67 labeling index was 60-70%. Typical and atypical mitosis in the tissue was also noteworthy, and all these findings supported LMS (Figure-1).

The lesions presenting with increased fluorodeoxyglucose (FDG) involvement in the left pleura [standard uptake value (SUVmax: 4.46-15.76), the lymph nodes with increased FDG involvement in the anterior mediastinum (SUVmax: 13.25) and the subcarinal (SUVmax: 5.69) were observed on positron emission tomography/ computed tomography (PET-CT) ordered primarily for focus screening of LMS. In addition, other lesions showing focal increased FDG involvement in the descending colon and sigmoid colon junction in size of 20x15 mm (SUVmax: 34.87) and the sigmoid colon in size of 15x11 mm (SUVmax: 21.08) were also observed (Figure-2).

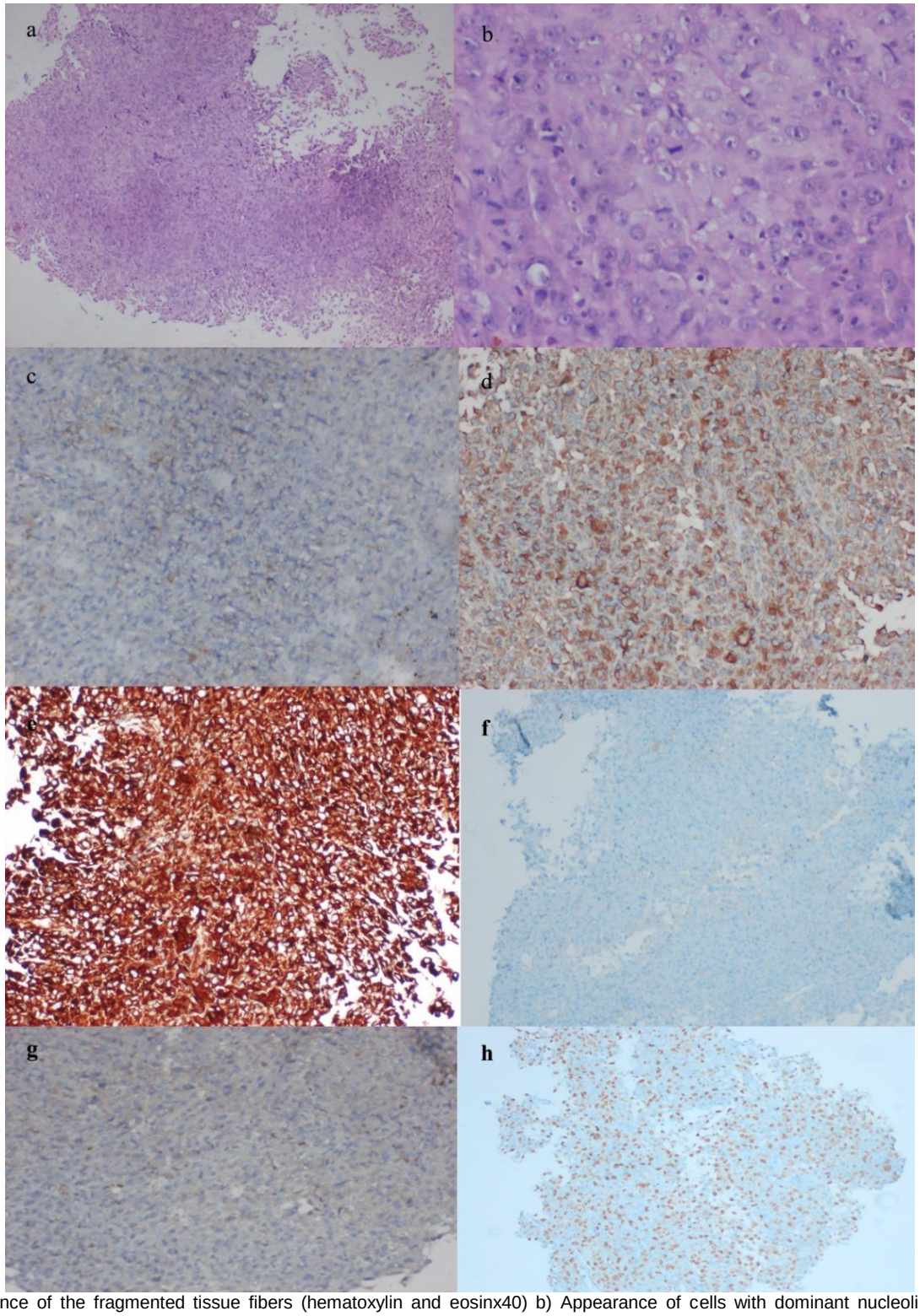
In the colonoscopy, a polypoid lesion of 15-20 mm in diameter was observed in the sigmoid colon (Figure-3).

Polypectomy could not be performed because the patient could not tolerate it. While the supportive treatment was continued, despite the planned chemotherapy. The patient received no chemotherapy because of the rapid impairment in his general status and died one month after diagnosis.

Discussion

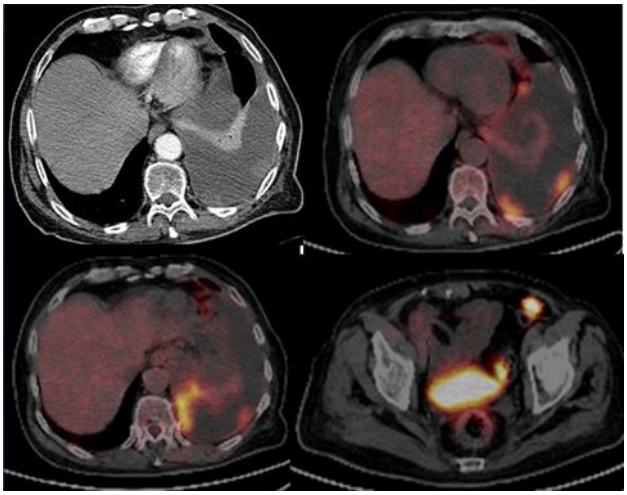
LMS of the GI tract is among the rarely diagnosed entities. To the best of our knowledge, several cases were reported in the literature between 1990-2015.[6] LMSs are mesenchyme-originated tumors capable of metastasizing from the primary sites, including the retroperitoneum, head, and neck. A case of massive pleural effusion was presented with the case of metastatic pulmonary LMS for the first time in 2015.[5] In light of the literature, no other cases of colonic LMS with the development of massive pleural effusion leading to pleural metastasis have so far been published, apart from the case we have reported. Therefore, we considered that our case is the second one related to the topic, except for the first case of colon LMS diagnosed with pleural biopsy.

LMS of the GI tract usually occurs in the fourth and sixth decades of life. The most common symptom is abdominal pain. In addition, such complications as an abdominal palpable mass, altered bowel rhythm, hemorrhage, perforation, or congestion are also encountered in LMS.[7] While



a) Apearance of the fragmented tissue fibers (hematoxylin and eosinx40) b) Appearance of cells with dominant nucleoli, including pleomorphic nuclei (hematoxylin and eosinx200) c) Calretinin staining negative (x100) d) Desmin staining positive (x100) e) Vimentin staining positive (x100) f) Alpha smooth muscle actin staining negative (x100) g) Pancytokeratin staining negative (x100) h) Ki-67 labeling index (x100)

Figure 1: Samples of Immunohistochemical staining.



Lesions demonstrating focally increased FDG involvement in left pleura (SUVmax: 4.46-15.76) and colon (SUVmax: 38.87) on PET-CT

Figure 2: Sections obtained through thorax CT and PET-CT scans.

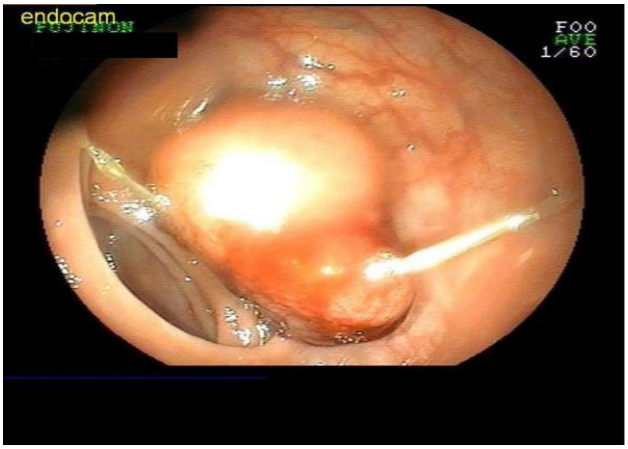


Figure 3: Colonoscopic appearance of colon leiomyosarcoma.

none of such findings were determined in our case, the case displayed the common symptoms of cancer, such as weakness and weight loss. Since most studies related to LMS-induced pulmonary metastases are in the form of case reports in the literature, the rates and features of LMS have yet to be fully characterized. Despite their nonspecific characteristics, the clinical symptoms may include cough, shortness of breath, hemoptysis, or chest pain. Our patient also revealed dry coughs and shortness of breath. Sarcomas have high F-18-FDG affinity in general.[2] In a study investigating 493 patients with different histological subtypes of bone and soft tissue sarcomas, there were more than five values of SUVmax in histologically high-grade tumors(8). It was reported that the anatomical origin of sarcomas affects SUVmax value, and sarcomas of gynecological origin tend to have higher SUVmax values than those with non-gynecological origin.[8] Although LSM in our case was out of gynecological origin, SUVmax value was observed to be very high (SUVmax: 34.87-21.08). Even if the diagnosis of LSM can also be performed morphologically through hematoxylin-eosin (HE) staining, the immunohistochemical investigation confirms the diagnosis.

If the patient is considered to be appropriate for operative treatment in the treatment process, surgical resection is the basis of the treatment, while chemotherapy and radiotherapy are available as adjuvant modalities.[9] However, if the tumor is unresectable, LMS is sensitive to doxorubicin, ifosfamide, and trabectedin, and the average survival rate is estimated to be 12 months.[10] Advanced primary pulmonary LMS is treated with chemotherapy, and the prognosis is quite poor. In the cases of pulmonary LMS metastasized from other systems, long-term survival can be achieved through metastasectomy.[5]

Pulmonary LMS - whether to be primary or metastatic - is a type of extremely rare and difficult to diagnose cancer with a poor prognosis. Our case is the first colon LMS diagnosed by pleural biopsy. Since early diagnosis is of crucial importance in determining treatment options and in prognosis, such cases may be considered among the causes of exudative pleural effusion, even if rare.

Conflict of interest: None declared.

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