ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 9, Issue – 3, June – 2024 , Page No. : 10 – 13

From rarity to revelation: shedding light on gastric leiomyosarcoma in a small needle core biopsy – A case report.

[1]Dr. Neda Ahsan, Contractual Medical Practitioner (CMP), Department of Pathology, Central Hospital, South Eastern Railway, Garden Reach, Kolkata, West Bengal, India.

[2]Dr. Mahamaya Sharma, Additional Chief Health Director (ACHD) & Head of Department, Department of Pathology, Central Hospital, South Eastern Railway, Garden Reach, Kolkata, West Bengal, India.

Corresponding Author: Dr. Neda Ahsan, Contractual Medical Practitioner (CMP), Department of Pathology, Central Hospital, South Eastern Railway, Garden Reach, Kolkata, West Bengal, India.

Citation this Article: Dr. Neda Ahsan, Dr. Mahamaya Sharma, "From rarity to revelation: shedding light on gastric leiomyosarcoma in a small needle core biopsy – A case report", IJMSIR - June - 2024, Vol – 9, Issue - 3, P. No. 10 – 13.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Gastric leiomyosarcomas are exceptionally rare tumors, with only few documented cases to date. The introduction of Immunohistochemistry (IHC) has significantly improved the differentiation between gastrointestinal stromal tumors (GIST) and leiomyosarcomas, reducing the number of misdiagnosed cases. Here, we describe the case of a 63-year-old male who presented with a two-month history of abdominal pain and swelling. A computed tomography (CT) scan was done revealing a large poorly defined, heterogeneous mass (measuring 17x16.6 cm) in the abdominal cavity, with loss of distinct boundaries in the left liver lobe, the anterior wall of stomach, duodenum and colon along with multiple metastasis. A trucut biopsy was taken from the mass in epigastric region which on histopathological examination was suggestive of a gastrointestinal stromal tumor (GIST). However, on further detailed examination with Immunohistochemistry (IHC), a high grade leiomyosarcomas was confirmed.

Keywords: Gastrointestinal Stromal Tumor (GIST), Immunohistochemistry (IHC), Leiomyosarcomas, Stomach.

Introduction

Leiomyosarcoma (LMS), an aggressive tumor arising from smooth muscle cells or their mesenchymal precursors, is among the most prevalent types of sarcoma accounting for 10 – 20% of all sarcomas.[1] It commonly presents in the retroperitoneum and uterus, with fewer instances observed in the extremities and trunk. Gastric leiomyosarcomas is an exceptionally rare form of sarcoma with only 19 reported cases to date.[2]Prior to the identification of Immunohistochemical markers like C-KIT and DOG-1, most cases of Gastrointestinal stromal tumors(GIST) were misidentified as gastrointestinal leiomyosarcomas. Diagnosis of primary leiomyosarcomas now relies on a combination of Histomorphologicaly features, positive immunoreactivity to smooth muscle antigens (SMA), and non-responsiveness to GIST immunomarkers.[3]

Case Report

A 63-year old male presented to the OPD with a two-month history of abdominal swelling and two weeks of fever. He was a known alcoholic with no significant past medical history, comorbidities or family history of malignancy. He had no history of pain, jaundice, hematemesis or melena.

On physical examination, a large globular abdominal swelling was noted. Following which diagnostic workup was done.

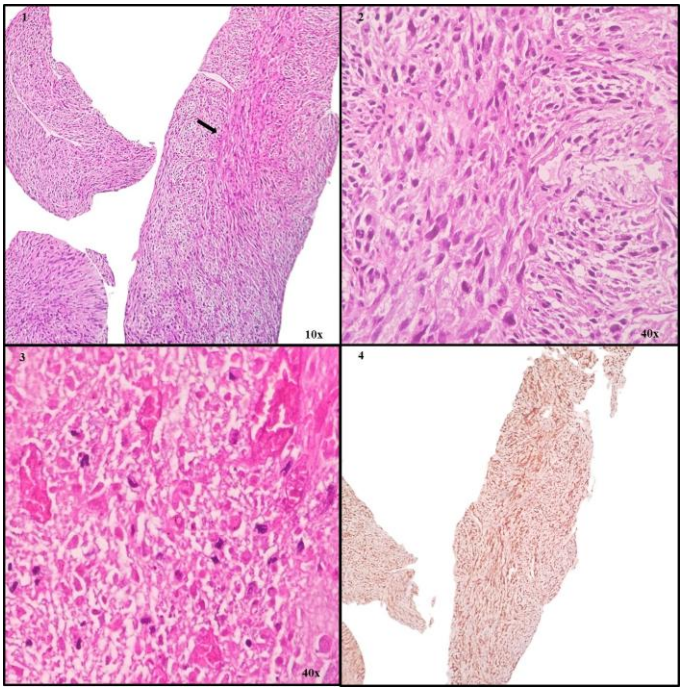
Laboratory investigations showed mildly increased WBC count (13.78 x 10^3/ml), anaemia (7.3 mg/dl), thrombocytopenia (73 x 10^3 / µl), raised PT/INR levels (15 seconds /1.37), elevated SGOT levels (128 IU/litre), raised CRP levels (24.23 mg/dl) and with normal levels of urea, creatinine, and electrolytes. Ultrasonography revealed a large (17 x 16.6 cm) ill-defined hetero-echoic mass involving the liver segment and anterior wall of the stomach.

Upper Gastric Endoscopy showed multiple stomach erosions in the body. Contrast-Enhanced Computed Tomography (CECT) of abdomen was done which showed an extensive ill-defined heterogeneous mass involving the left liver lobe, stomach, duodenum, colon, omental and mesenteric deposits, along with multiple liver nodules. PET scan revealed multiple bone metastases.

Atrucut biopsy was performed and two cores measuring 0.4 cm and 1 cm were received for histopathological examination (HPE). The histopathological examination revealed a tumor comprising of bland spindle cells arranged in short fascicles with eosinophilic cytoplasm, elongated nuclei and inconspicuous nucleoli. Few moderately pleomorphic cells at the periphery of necrotic tissue were noted along with occasional mitotic figure; hence a diagnosis of metastatic Gastrointestinal Stromal Tumor (GIST) was offered. To confirm this diagnosis, an Immunohistochemical (IHC) panel was carried out, including markers like DOG-1, C-KIT, CD34, and S100.The tumor was negative for these markers.

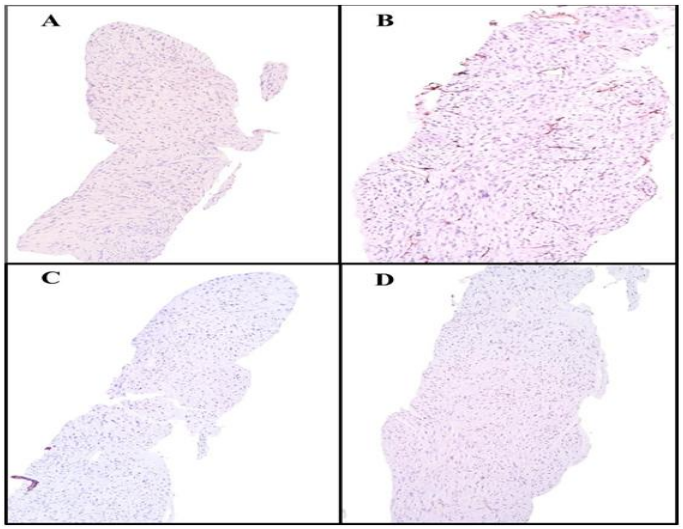
Given the negative results, Smooth Muscle Actin (SMA) was performed to exclude the possibility of leiomyosarcomas as a differential diagnosis for his disseminated disease. IHC revealed strong and diffuse staining for Smooth Muscle Actin (SMA), confirming the diagnosis of Leiomyosarcomas.

Unfortunately, the patient succumbed to the illness before any medical intervention could be initiated, due to his advanced stage of disease.



Figure

1. Low power view shows a predominantly benign tumor with bland spindle cells. Arrow shows presence of leiomyosarcomas.
2. High power view shows focal area with moderately pleomorphic cells.
3. Shows presence of necrosis.
4. Shows strong and diffuse SMA positivity.



Figure

Tumor shows negativity for GIST markers.

A. Negative for DOG-1
B. Negative for CD34
C. Negative for CKIT (CD117)
D. Negative for S100

Discussion

Leiomyosarcomas (LMS), a common subtype of soft tissue sarcomas accounts for Upto 10% to 20% of all sarcomas.[1]Before the emergence of KIT, GISTs were often misdiagnosed as leiomyomas and leiomyosarcomas. However, since its establishment in the late 1990s, it has been recognized that primary gastric leiomyosarcomas is extremely rare, comprising less than 1% of gastric tumors.[4]They typically originate from the muscularispropria or muscular is mucosa layers at different sites within the stomach and may either remain within the gastric wall as intramural lesions or extend primarily into the submucosa, possibly infiltrating the mucosa and forming endogastric masses, sometimes with ulceration. Alternatively, growth toward the subserosa results in exogastric lesions, often diagnosed only after significant enlargement.[5]The cause of gastric leiomyosarcomas remains uncertain, but potential risk factors include radiation exposure, chemical exposure, Epstein-Barr virus infection, and immunosuppression.[6]

Patients with gastric leiomyosarcomas may exhibit a range of symptoms, with the most common being upper gastrointestinal (UGI) bleeding and abdominal pain. Additional symptoms can include the presence of a palpable mass, weakness, weight loss, nausea, and vomiting as observed in our case. The specific symptoms experienced often depend on factors such as the location and size of the tumor, as well as the presence of ulceration. In cases where tumors are particularly large, the primary clinical sign may be the discovery of a sizable abdominal mass of unknown origin.[7] Leiomyosarcomas often spread hematogenously, with around 40% of patients exhibiting distant metastases at the time of initial diagnosis, as was seen in this patient with majority eventually developing metastatic disease. The liver and lungs are frequently affected by metastases, although metastasis to other organs is also observed. [7]Metastatic GIST should be considered as a strong differential as they present with similar symptoms. The most common site of metastasis for GIST is the liver and peritoneum, with bone metastasis being extremely rare. [8] Leiomyosarcomas often remains asymptomatic until they reach a significant size and extend beyond the stomach into the neighboring organs at the time of diagnosis, as noted in the indexed case. Radiological imaging including Ultrasound (USG), Computed tomography (CT) and Magnetic Resonance Imaging (MRI), are essential for the assessment of leiomyosarcomas. CT scans are particularly sensitive, as they not only help in the detection of tumors but are also useful in identifying metastases.[9]While these imaging techniques offer valuable insights into the tumor's origin and spread, the definitive diagnosis still relies on histopathological examination.

Microscopically, distinguishing leiomyosarcomas from other spindle cell tumors like Gastrointestinal Stromal tumor (GIST) and nerve sheath tumors is crucial. Features such as high mitotic count, nuclear atypia, and pleomorphism which usually aid in differentiating leiomyosarcomas from benign leiomyomas, were absent in this case. Notably, Metastatic Gastrointestinal Stromal tumor (GIST),a primary differential diagnosis, should be excluded in all cases of disseminated disease, as was done here. Immunohistochemical markers like CD117, CD34, and DOG1 help in this distinction, with DOG1 being the best marker for GIST.[9] Conversely, leiomyosarcomas typically shows negative staining for C-KIT or DOG1 and positive staining for α-SMA, desmin, and other markers. This differentiation is essential as GIST responds to KIT-directed therapy, while leiomyosarcomas does not. Neurogenic tumors are excluded by S100 negativity. Therefore, for an accurate diagnosis, histopathological examination accompanied by Immunohistochemical staining is essential, particularly in a small core needle biopsy.

Treatment of leiomyosarcomas depends on a number of prognostic factors including histological grade, tumor type, size and metastasis. Surgical excision is the most curative option for its treatment.[6] But in large inoperable cases where excision alone isn't sufficient, chemotherapy and radiation are also used, having limited impact.

Conclusion

In summary, while Gastrointestinal Stromal tumor (GIST) is the predominant tumor in the gastric region, the presence of disseminated metastasis in a patient's clinical presentation warrants consideration of gastric leiomyosarcomas as well while examining a small needle core biopsy. In such cases, thorough Immunohistochemical analysis is essential for accurate diagnosis and appropriate management.

References

1. Mangla A, Yadav U. Leiomyosarcoma. PubMed. Treasure Island (FL): StatPearls Publishing; 2021.
2. Wang T, Zreik R, Leng B. Primary Gastric Leiomyosarcoma: A Rare Case. Curēus. 2023.
3. Aggarwal G, Sharma S, Zheng M, Reid MD, Crosby J, Chamberlain S, et al. Primary leiomyosarcomas of the gastrointestinal tract in the post–gastrointestinal stromal tumor era. 2012;16(6):532–40.
4. Kang WZ, Xue LY, Tian YT. Leiomyosarcoma of the stomach: A case report. World J Clin Cases. 2019;7(21):3575–82.
5. Takagi T, Saito S, Yokota S, Kaneko Y, Takahashi K, Rihito K, et al. Laparoscopic and endoscopic cooperative surgery for leiomyosarcoma of the stomach: a case report with a review of the literature. J. Surg. Case Rep. 2021;7(1).
6. Mehta V, Rajawat M, Rastogi S, Phulware RH, Mezencev R. Leiomyosarcoma of the stomach with metastasis to the liver: a case report with review of the literature. Future Sci. OA. 2018;4(2):264–4.
7. Garg R, AlRajjal A, Berri R, Barawi M. Primary Gastric Leiomyosarcoma: a Case Report and Review of the Literature. J. Gastrointest. Cancer.2020; 51(1):335–40.
8. Suresh C, Chaudhuri T, Babu KG, Lakshmaiah KC, Lokanatha D, Jacob LA, et al. Metastatic gastrointestinal stromal tumor: A regional cancer center experience of 44 cases. South Asian J. Cancer. 2017;6(3):118–21.
9. Al-Yousofy F, Alshargabi G, Ahmed F, Almohtadi A, Fazea M, Altam A. Primary huge gastric leiomyosarcoma with multiple metastases in a 60-year-old female: a case report. Pan Afr. Med. j. 2022;42(223).