Journal of International Case Reports (ICARE)
Case Presentation
Volume 2 | Issue 3 | Pages: 205-207
Published Date: August 07, 2023

ALK-Negative Inflammatory Myofibroblastic Tumour of the Ileum with Liver Metastasis & Paraneoplastic Syndrome- A Diagnostic & Therapeutic Challenge

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Received Date: July 20, 2023; Accepted Date: August 02, 2023; Published Date: August 07, 2023

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Citation: Ravel N, Rajan T, Sathiyamoorthy PN, Ramanan A, Chandralekha K, Dheeraj C, Meena K and Vijayalakshmi P. ALK-Negative Inflammatory Myofibroblastic Tumour of the Ileum with Liver Metastasis & Paraneoplastic Syndrome- A Diagnostic & Therapeutic Challenge. ICARE. 2023;2(3):1041.

Abstract

Background: Inflammatory Myofibroblastic Tumour (IMT) is also known as inflammatory pseudotumor and plasma cell granuloma. IMT is classified under the WHO category of soft tissue tumour - Fibroblastic and Myofibroblastic Tumour - borderline category. IMTs are locally aggressive, often occurring in the thoracic and abdominal cavities in young adults and children. Approximately 33% of patients with IMT develop paraneoplastic syndrome including fever, growth failure, malaise, weight loss, anemia, and thrombocytosis. ALK rearrangements occur in <70% of patients. ALK-negative IMTs may have ROS1 or PDGFRB mutations. Recent literature suggests NTRK gene fusion in a subset of the population. When complete resection is possible, it is curative in approximately 75% of cases. Metastasis is rare (<5%), but local recurrence is more common. The treatment of patients with advanced ALK fusion positive IMT is crizotinib. In patients without an ALK fusion, treatment for the locally aggressive disease is palliative with retrospective studies showing response with, taxanes and cisplatin-based chemotherapy. Few tumors have metastatic potential, and the disease is rarely fatal (5-year survival of 87%).

Case Description: Here we report a case of IMT arising from the ileum in a 62-year-old lady with liver metastasis and paraneoplastic syndrome. She presented with abdominal pain, leucocytosis, and anemia. Imaging features revealed an ileal mass and was taken up for laparotomy and resection. Post-op HPE was consistent with IMT, ALK-negative. PET CT showed multiple omental deposits and liver secondaries. She was started on imatinib and later was changed to paclitaxel following which she had a stable disease with symptomatic improvement.

Conclusion: IMT is a rare locally aggressive, recurrent borderline tumor with low metastatic potential and paraneoplastic association. ALK-negative metastatic tumors have no proven treatment options. Hence being a diagnostic and therapeutic challenge.

Keywords: Inflammatory myofibroblastic tumor; Paraneoplastic syndrome; ALK rearrangement

Introduction

Inflammatory myofibroblastic tumor (IMT) is also known as inflammatory pseudotumor and plasma cell granuloma. IMT is classified under the WHO category of soft tissue tumour – fibroblastic and myofibroblastictumour – borderline category [1,2]. It is seenin young adults and children [3]. Approximately 33% of patients with IMT develop a paraneoplastic syndrome including fever, growth failure, malaise, weight loss, anemia, and thrombocytosis [4]. Here we report a case of IMT arising from ileum in a 62 year old lady with liver metastasis and paraneoplastic syndrome.

Case Report

A 62-year-old post-menopausal lady k/c/o HTN, CAD presented with easy fatigability and significant weight loss of 6 months duration. Clinical examination revealed pallor and an abdominal mass felt in the right para umbilical region with restricted mobility. On evaluation, the patient had anemia, thrombocytosis, and neutrophilic leucocytosis (Hb: 8 g/dl PLT: 6L, TC: 38,700, N86%). Peripheral smear showed normochromic normocytic anemia, and neutrophilic leucocytosis with no atypical cells. USG abdomen showed mild splenomegaly with a hyperechoic lesion of size 6 cm × 6 cm in the paraumbilical region. Bone marrow aspirate showed hypercellular marrow, myeloid hyperplasia with an increase in precursors, and no atypical cells. PET CT showed FDG avid soft tissue dense nodules in the omentum and peritoneum with right hypochondrium showing a 5.8 cm × 2.3 cm × 5 cm lesion probably arising from the small bowel (SUV 6.35). The paraumbilical region shows a 1.6 cm × 1cm size FDG avid lesion. Similar lesions were seen in the infra umbilical, suprapubic region, and sacral subcutaneous plane. Subcapsular lesions were seen in the liver, the largest 2.8 cm × 2cm in segment 4 (SUV: 6) (Figures 1 and 2). There was diffuse uptake in the marrow (SUV 5.4) and splenomegaly with increased uptake. The patient was taken up for laparotomy and small bowel resection. Per operatively there was a large exophytic growth arising in the terminal ileum, with another large lesion in the sub-hepatic region abutting the abdominal wall. Post-op HPE showed features of IMT with IHC showing desmin, vimentin, and SMA positivity, CD117, DOG1, CK, synaptophysin & ALK-negative (Figures 3 and 4). Therefore, a diagnosis of ALK-negative IMT with liver Mets and paraneoplastic syndrome of anemia, leucocytosis, and thrombocytosis was made. The patient was started on Imatinib, and the patient was symptomatically better with the improvement of anemia, leucocytosis, thrombocytosis, and systemic symptoms. After 2 months patient progressed with worsening of symptoms and was challenged with single-agent paclitaxel and is having a stable disease after 4 cycles.

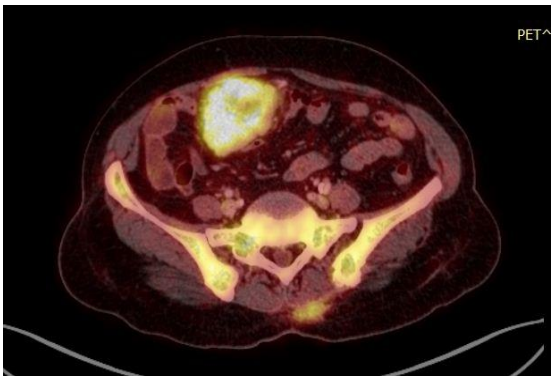

Figure 1: PET CT showing metabolically active ileal Lesion.

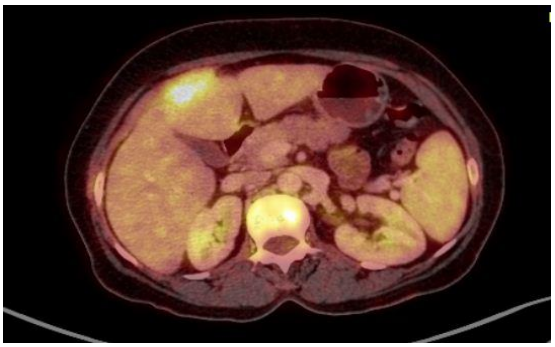

Figure 2: PET CT showing metabolically active subcapsular liver lesion.

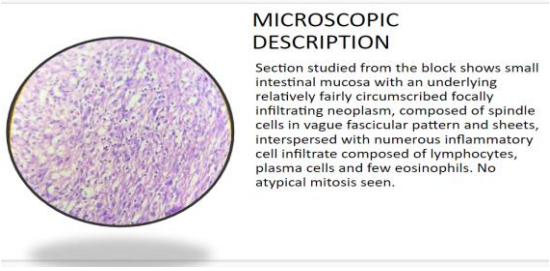

Figure 3: Microscopic picture of IMT.

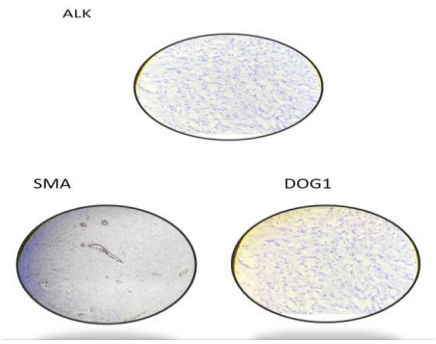

Figure 4: IHC showing ALK negative, DOG1 negative, SMA positive tumour.

Discussion

Inflammatory Myofibroblastic Tumor (IMT) is also known as inflammatory pseudotumor and plasma cell granuloma. IMT is classified under the WHO category of soft tissue tumour – fibroblastic and myofibroblastictumour – borderline category [1,2]. IMTs are locally aggressive, often occurring in the thoracic and abdominal cavities in young adulIts and children [3]. Approximately 33% of patients with IMT develop a paraneoplastic syndrome including fever, growth failure, malaise, weight loss, anemia, and thrombocytosis [4]. Its histology is heterogeneous, with variable grouping of spindle, fibroblastic- myofibroblastic, and inflammatory-type cells. ALK rearrangements occur in <70% of patients [5]. ALK negative IMTs may have ROS1 or PDGFRB mutations [6]. Recent literature suggests NTRK gene fusion in a subset of population [7]. When complete resection is possible, it is curative in approximately 75% of cases. Metastasis is rare (<5%), but local recurrence is more common. Treatment of patients with advanced ALK fusion positive IMT is crizotinib [8]. In patients without an ALK fusion, treatment for locally aggressive disease is palliative with retrospective studies showing response with taxane& cisplatin based chemotherapy. Few tumors have metastatic potential and the disease is rarely fatal (5-year survival of 87%) [4].

Conclusion

IMT is a rare locally aggressive, recurrent borderline tumor with low metastatic potential and paraneoplastic association. ALK-negative metastatic tumors have no proven treatment options. Hence being a diagnostic and therapeutic challenge.

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