HMB-45 Negative Solitary Mesenteric Lymphangioleiomyoma: A Case Report

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Abstract

Lymphangioleiomyomas are typically associated with lymphangioleiomyomatosis a multisystem disease, affecting predominantly women in the reproductive period. We present a case of a 44 year old African American female with pain in the left upper quadrant of the abdomen. The CT scan of the abdomen and pelvis showed a large left abdominal and pelvic mass encasing mesenteric vessels and causing displacement of the bowel loops to the right. Grossly, the specimen consisted of a segment of bowel with a 20 cm tan brown and cystic mass arising from the mesentery and containing milky fluid. The histological findings were typical of lymphangioleiomyoma and included ramifying network of lymphatic spaces lined by a single layer of endothelium surrounded by fascicles and bundles of abnormal smooth muscle-like cells(LAM cells), occasional lymphoid follicles, and congested vascular spaces. However, HMB-45 staining which is typically positive in the tumor cells was negative. Most commonly, this disease process involves the lungs, which were not involved in our patient. Extrapulmonary lymphangioleiomyomas are rare and mainly occur in the pelvis,mediastinum and retroperitoneum. The mesentery as an involved site of extra pulmonary lymphangioleiomyomas is an extremely rare location. Only four cases of mesenteric lymphangioleiomyomas have been described in the literature, of these two also had pulmonary involvement. Our case is unique in that it is not only the largest solitary mesenteric lymphangioleiomyoma, but it is also the first report of an HMB-45 negative lymphangioleiomyoma involving the mesentery.

Case Report

A 44 year old African American woman presented with pain in the left upper quadrant of the abdomen. The CT scan of the abdomen and pelvis showed a large left abdominal and pelvic mass encasing mesenteric vessels and causing displacement of the bowel loops to the right. Grossly, the specimen consisted of a segment of bowel with a 20 cm tan brown and cystic mass arising from the mesentery and containing milky fluid. The histological findings were typical of lymphangioleiomyoma and included ramifying network of lymphatic spaces lined by a single layer of endothelium surrounded by fascicles and bundles of abnormal smooth muscle cells(LAM cells), occasional lymphoid follicles and congested vascular spaces. However, the tumor cells were negative for HMB-45 staining. The tumor was diagnosed as a solitary mesenteric lymphangioleiomyoma. The patient had an uneventful postoperative course and has been free of recurrence till now.

Figure 1. CT Scan

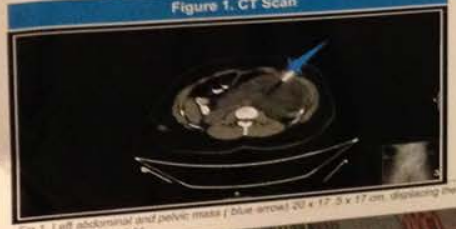


Fig 1. Left abdominal and pelvic mass (blue arrow) 20 x 17. 5 x 17 cm, displacing the bowel loops to the right

Figure 2. Gross Pathology

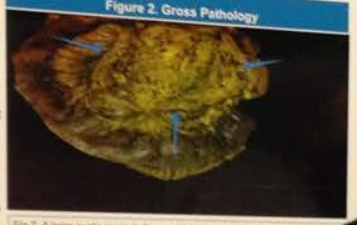
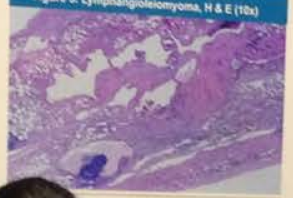


Fig 2. A large cystic mass in the small bowel mesentery(blue arrow). The cut surface showed a cystic mass with abundant milky content in spaces

Figure 3. Lymphangioleiomyoma, H & E (10x)

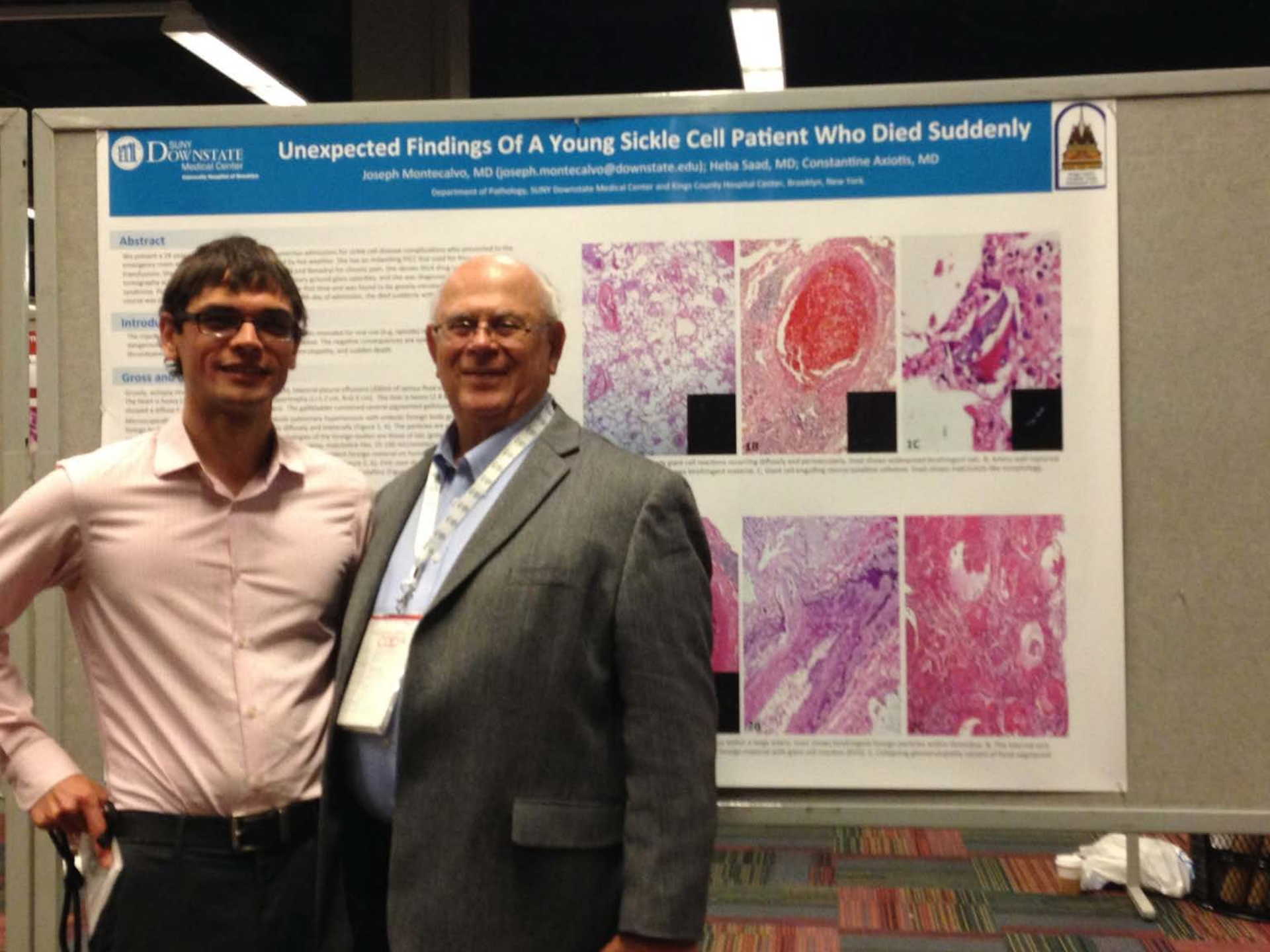


shows ramifying network of lymphatic spaces of abnormal smooth muscle- like cells(LAM cells), and congested vascular spaces

Figure 4 ,5. Ly

Fig 4,5. The individual cells

Figure 6(A


SUNY Downstate Medical Center
Unexpected Findings Of A Young Sickle Cell Patient Who Died Suddenly
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Abstract
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HEALTH SCIENCE CENTER AT BROOKLYN
STATE UNIVERSITY OF NEW YORK
ARID1A Expression in Uterine Carcinosarcomas
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Background
ARID1A (AT-rich interactive domain 1 A gene), is a recently identified tumor suppressor gene which plays a central role in endometrial carcinogenesis [1-3]. Its loss of expression has been shown to correlate with progression of atypical hyperplasia of endometrium to low grade and high grade endometrial carcinoma. By molecular profiling ARID1A has been associated with endometrioid-type in contrast to serous-type of endometrial carcinoma [4,5]. Expression of ARID1A in carcinosarcoma has been very briefly explored in the literature [6]. The purpose the study is to assess the frequency of loss of ARID1A in uterine carcinomasarcoma (Malignant Mixed Mullerian tumors, MMMT) and whether this loss can be correlated with any discernible clinicopathological implications or the subtype of its epithelial component.
Results
Materials and Methods
The expression of ARID1A in uterine malignant mullerian mixed tumor was studied by immunohistochemistry using a mouse anti-human ARID1A polyclonal antibody (Sigma). The secondary rabbit anti-mouse antibody and detection system were from EnVision™+ Dual Link System (DAKO, Atlanta, GA). Immunostaining was performed on a DAKO autostainer. Briefly, after rehydration, antigens were retrieved in a Decloaking Chamber (Pressure Cooker) using the Dako Target Retrieval Solution (DAKO, Atlanta, GA). Primary anti ARID1A antibody was diluted to 1:200. Secondary antibody and HRP color-developing agents were applied as instructed by the supplier (DAKO, Atlanta, GA). Finally, the slides were counterstained with hematoxylin. Controls are appropriate.
Fifty four cases of uterine malignant mullerian mixed tumor were retrieved from the pathology archives. . Cases with both endometrioid ()and serous component ()were included. Expression of ARID1A was assessed by immunohistochemistry (human polyclonal antibody, by Sigma) and clinical data was collected by a retrospective medical record review. Positive expression was defined as >25% of positive strong expression in tumor cells. Clincopathological correlation (age, tumor stage, lymphovascular invasion, lymph node status, sarcoma histological subtypes), and with the expression of ARID1A was studied.
Figure 1. Carcinomatous component of carcinosarcoma with negative AIRD1A stain. The left panels are H&E staining at 100 X (A) and 400 X magnification (C). The right panels are immunostaining using the ARID1A antibody at 100 X (B) and 400 X magnification (D).
Figure 3. Carcinosarcoma with positive ARID1A stain. The left panels are H&E staining at 100 X (A) and 400 X magnification (C). The right panels are immunostaining using the ARID1A antibody at 100 X (B) and 400 X magnification (D).
Conclusion
•ARID1A is lost or has low expression in a subset of MMMT cases, in which both carcinomatous and sarcomatous components showed low expression.
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SUNY Downstate Medical Center
University Hospital of Brooklyn
Epstein Barr Virus-associated Atypical B-Cell Lymphoproliferation in Adult T-cell lymphoma/Leukemia – A Diagnostic Dilemma
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