POST-TRANSPLANT EPITHELIOID INFLAMMATORY MYOFIBROBLASTIC SARCOMA: A CASE REPORT

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Introduction:
Inflammatory myofibroblastic tumor (IMT) is a mesenchymal neoplasm of intermediate biological potential which might recur or rarely metastasize. They usually present as solitary or rarely as multiple masses in abdominal/pelvic region, lung, mediastinum and retroperitoneum of children and young adults. IMT's post hematopoietic stem cell transplantation (HSCT) has been reported in only eight patients till date. In post solid organ transplant, their incidence is even rarer with only few case reports to support the evidence

In 2011, Mariño-Enríquez et al. described a novel variant of IMT, Epithelioid Inflammatory Myofibroblastic Sarcoma (EIMS). They are characterized by an epithelioid morphology and prominent neutrophilic inflammatory infiltration. Clinically, it has a more aggressive behavior with short disease-free survival. Approximately 50% of IMTs aberrantly express ALK protein triggered by clonal rearrangements of ALK gene located on chromosome 2p23.

Case report:
An 18 month old female received a liver transplant in October, 2015 for decompensated chronic liver disease with clinical and biochemical features consistent with “progressive familial intrathoracic cholestasis”. Her post operative period remained uneventful and was discharged in stable condition. No intra abdominal mass lesions were detected on radiology before and soon after the transplant. In May, 2016, she presented with fever and distended abdomen. CT abdomen and PET scan showed FDG avid multiple lesions in omentum, subdiaphragmatic area and attached to the serosa of small bowel (figure 1). Their trucut biopsies revealed a mesenchymal neoplasm, positive for vimentin and SMA. All other lineage markers such as caldesmon, C-kit, CD3, S100, and CD31 were negative. Since the MIB-1 index was 15-20%, the suspicion of malignancy was high. More representative sample was required for final tumor typing. The patient was operated upon, with a plan to resect all the tumors. Frozen section confirmed the mesenchymal neoplasm.

Two large masses measuring 16 and 8 cm in their maximum dimensions and 2 smaller nodules (0.5 cm, 1 cm) attached to the bowel were excised. Their external surface was uniform smooth, grey white. Cut section was vaguely lobulated, grey white and myxoid. Tumor masses from omentum and small bowel showed large areas of necrosis & hemorrage. Figure 2 for Histomorphology.

Discussion:
De novo neoplasms account for almost 30% of deaths 10 years after liver transplantation and are the most common causes of mortality in patients surviving at least 1 year after transplant. The risk of malignancy is two to four times higher in transplant recipients than in an age- and sex-matched population. Skin cancers, lymphomas and Kaposi sarcomas occur most frequently after organ transplantation. The incidence of malignancies is increased as a consequence of chronic immunosuppression after solid organ transplantation. EBV is implicated in Post Transplant Lymphoproliferative Disorders and Myogenic tumors. IMT with prominent epithelioid cytology transformation is very rare. In a study with 73 cases of IMT, Cook et al. found 4 cases that exhibited round cell transformation characterized by large polygonal cells with large nuclei and prominent nucleoli, in a loose, pale staining background. One of these 4 cases showed distinctive nucle membrane staining pattern of ALK.

In 2010, Butrynski et al. reported sustained partial response to the ALK inhibitor crizotinib in a patient with ALK-translocated IMT with epithelioid cytology. Only 20 cases have been documented in English literature, the clinical, morphological and immunohistochemical features of which are summarized in Table below. All of these showed positive signal for ALK translocation resulting from RANBP2-ALK fusion. Accordingly, we infer that RANBP2-ALK fusion gene might be a potential molecular mechanism for the rapid growth and recurrence of EIMS.

<table>
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<td>Age incidence</td>
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<td>72% adults</td>
<td>34</td>
<td>5:1</td>
<td>84% intra abdominal</td>
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2. Asli Tufan, Gulistan Bahat Inflammatory pseudotumors after stem cell transplantation [Hematology Reports 2015; 7:5848-5850].


