CASE REPORT

Association of two rare entities: Kasabach–Merritt syndrome and littoral cell angioma
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Introduction

Kasabach–Merritt syndrome (KMS) is recognized by the involvement of a vascular tumor with severe thrombocytopenia and a potentially fatal coagulopathy.[1] Morbidity and mortality are high, as a result of profound bleeding, sepsis, or vital organ involvement when it is seen in infancy.[2] Interaction between platelets and endothelial cells, and the resulting proangiogenic phenotype, has been recognized as the major pathogenesis of KMS.[3]

Usually, it is not associated with infantile hemangiomas but may occur as a complication of rare lesions such as Kaposiform hemangioendothelioma and tufted angioma. Many of these arise in the soft tissue of the extremities.[4] Littoral cell angioma (LCA) is another benign vascular lesion of spleen rarely associated with and complicated by life-threatening KMS.[5] It originates from the splenic littoral cells.

Case Report

A 1-year-old male child was admitted to our hospital with abdominal distention and pain abdomen of 2 months duration.

Routine hematological parameters showed persistent severe thrombocytopenia, severe anemia, low red blood cell (RBC) count, high white blood cell count, raised prothrombin time, activated partial thromboplastin time, international normalized ratio, and increased bilirubin and lactate dehydrogenase levels with no improvement after transfusions.

Persistent anemia, thrombocytopenia, and deranged coagulation profile provoked surgeon to perform splenectomy.

Histopathological analysis of a grossly enlarged spleen [Figure 1] weighing 430 g revealed a vascular lesion involving the whole spleen, suggestive of LCA [Figures 2 and 3].

Child showed rapid clinical improvement and hematological profile also normalized within 2–3 days of post splenectomy [Table 1].

Discussion

LCA, first defined by Falk et al. in 1991,[5] is a rare, benign vascular tumor of the spleen. It originates from littoral cells that line the sinusoids of the red pulp. Usually, it occurs in adults and appears to be extremely rare in children. LCA is usually asymptomatic and is only discovered incidentally. The incidence of splenic hemangiomia varies from 0.03% to 14% at the autopsy.[6]

Clinically, LCA presents as a solid mass often along with splenomegaly, thrombocytopenia, or anemia, or as pyrexia of unknown origin but is often asymptomatic and is only discovered incidentally. The symptoms of KMS such as anemia, thrombocytopenia, and coagulopathy may well be associated with hypersplenism but is very rare in LCA.[7]

Grossly, two forms of LCA: Diffuse multinodular and solitary (rare).

Two subtypes of LCA with malignant potential are described: Littoral cell angiosarcoma and littoral cell...
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Hemangioendothelioma. Patients, if affected by either of these two, may present with metastatic disease, even after undergoing a splenectomy.

Histopathology of the expunged tissue is the only diagnostic test even in today’s era of advanced radiology as they cannot give a definite diagnosis due to similar appearances in other tumors but can suggest the vascular nature of the tumor. Our case showed diffuse involvement of spleen by unorganized anastomosing vascular channels lined by plump endothelial cells with hemophagocytosis or eosinophilic globules. Atypia and mitosis were notably absent. Few scattered megakaryocytes were seen. Micropapillary pattern in some areas and occasional lymphoid follicle were other features noted. This

Table 1: Pre- and post-operative hematological parameters

<table>
<thead>
<tr>
<th>Parameters</th>
<th>September 28, 2018</th>
<th>September 30, 2018</th>
<th>October 1, 2018</th>
<th>October 10, 2018</th>
<th>October 23, 2018</th>
<th>October 27, 2018</th>
<th>October 29, 2018</th>
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<tbody>
<tr>
<td>Hb (g/dL)</td>
<td>3.0</td>
<td>5.4</td>
<td>8.6</td>
<td>5.9</td>
<td>5.5</td>
<td>11.3</td>
<td>8.8</td>
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<tr>
<td>RBC count (million/cumm)</td>
<td>0.9</td>
<td>1.9</td>
<td>3.1</td>
<td>2.3</td>
<td>2.0</td>
<td>4.1</td>
<td>3.3</td>
</tr>
<tr>
<td>WBC count (cells/cumm)</td>
<td>25,000</td>
<td>7880</td>
<td>12,300</td>
<td>11,010</td>
<td>11,560</td>
<td>9728</td>
<td>13,380</td>
</tr>
<tr>
<td>Platelet count (lakhs/cumm)</td>
<td>0.11</td>
<td>0.39</td>
<td>0.08</td>
<td>0.07</td>
<td>0.05</td>
<td>0.66</td>
<td>1.51</td>
</tr>
<tr>
<td>nRBCs/100 WBC</td>
<td>50</td>
<td>400</td>
<td>1170</td>
<td>770</td>
<td>5380</td>
<td>14990</td>
<td>2230</td>
</tr>
<tr>
<td>PT(C)</td>
<td>-</td>
<td>28.0</td>
<td>-</td>
<td>-</td>
<td>11.5</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>PT(T)</td>
<td>-</td>
<td>36.5</td>
<td>-</td>
<td>-</td>
<td>&gt;120 s</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>APTT(C)</td>
<td>-</td>
<td>11.5</td>
<td>-</td>
<td>-</td>
<td>28.0</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>APTT(T)</td>
<td>-</td>
<td>25.9</td>
<td>&gt;190 s</td>
<td>-</td>
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</tr>
</tbody>
</table>

RBC: Red blood cell, WBC: White blood cell, PT: Prothrombin time, APTT: Activated partial thromboplastin time

Figure 1: (a) Splenectomy specimen m/s 15 cm × 9 cm × 3 cm. E/S – gray-brown, congested. (b) C/S – dilated areas not filled with blood gives the spongy appearance

Figure 2: Numerous, unorganized vascular channels, with an intervening red pulp-like disorganized stroma, with (a) or without lymphoid follicles (b) (H and E ×10). (c) Anastomosing channels lined by plump cells (H and E ×40). (d) Micropapillary architecture in some regions (H and E ×10)

Figure 3: (a) Dilated vascular spaces lined by plump cells (arrow) with the appearance of sinus lining (“littoral”) cells. Deposition of hemosiderin in some cells. (b) Megakaryocyte (Asterix). The channels lined by endothelial cells with hemophagocytosis (c) or eosinophilic globules (d) (H and E ×40)
emphasizes the need for considering a primary splenic disorder in children with chronic thrombocytopenia and splenomegaly.

Incidental discovery of thrombocytopenia, but no diagnosis despite investigations and absence of complications, in a 22 months child was reported. However, subsequent acute clinical and biological worsening with splenomegaly and severe consumption coagulopathy prompted a suspicion of myeloproliferative disease disproved by bone marrow examination. Splenectomy was performed to stop the failing status and risk of rupture. Histopathological analysis revealed LCA infiltrating whole spleen. In our case also LCA was not suspected in the associated KMS attributed to splenomegaly but was revealed by histopathological analysis.

In KMS, activation of the coagulation cascade is supposed to be due to platelet trapping, activation, and consumption within the abnormal vascular structure.

In our case persistent thrombocytopenia despite transfusions and coagulation abnormalities setting in later, prompted splenectomy for therapeutic reasons and subsequent histopathology revealed the associated finding of LCA as the cause for underlying KMS presentation. Post-splenectomy investigations showed rebounce in platelet counts.

Management of KMS is still controversial and treatment is usually empirical and response is unpredictable. Steroid therapy, interferon-alpha 2, vincristine, ticlopidine, radiation and embolization, and rarely splenectomy are the available options.

**Conclusion**

KMS diagnosis should be suspected in patients presenting with splenomegaly and profound thrombocytopenia. Our case, in a child with LCA of spleen is a very rare and benign vascular lesion of an internal organ with unusual presentation of KMS.

**References**
