TTP and PNH: 2 cases demonstrating presentation in the peripartum setting and overlap with symptoms of PET/HELLP. Early recognition of these rare mimics can be lifesaving. Both presented in a regional hospital 3 hours from a tertiary unit.

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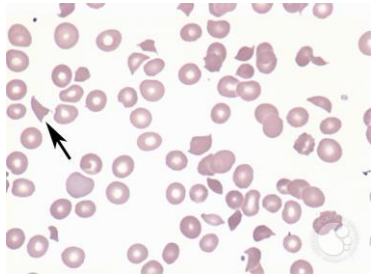
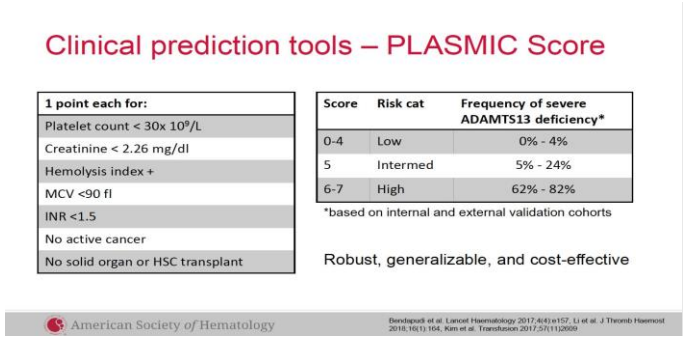
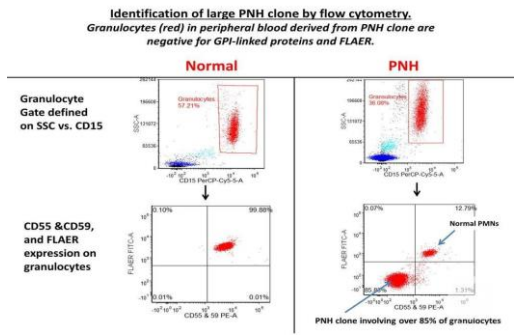
Case 1
Nov 2018, 38 year old primigravida delivered DCDA twins at 36+3 via LSCS for threated premature labour secondary to E.coli UTI. Well initially. Day 4 post partum had small coffee ground vomit. Hyperreflexia noted on examination. Slight elevation GGT. Renal function normal. Haemoglobin 70 g/L, platelet count 9 x 10^9/L. MCV84fl. LDH1900 iu/L. Initial concerns regarding evolving severe PET and given magnesium bolus and infusion awaiting blood film. Urgent blood film, in house, consistent with microangiopathic haemolytic anaemia (MAHA). PLASMIC score 7 out of 7. Working diagnosis TTP. Urgent HEMS transfer to tertiary unit for plasmapheresis (PLEX). Also given pulse methylprednisolone and rituximab. Through first day of illness rapidly deteriorated with mental obtundation thence coma. Intubated and ventilated with high inotrope requirement. PLEX continued. By 48 hours could facetime twins in the regional hospital. Made full recovery and received 2 years maintenance rituximab. Remains well and relapse free. Bloods confirmed initial ADAMTS-13 <1% which normalised with therapy.

Case 2
Previously well 22 year old primigravida. Vaginal delivery, epidural with low vacuum extraction for failure to progress. Post partum developed generalised abdominal pain and gross haematuria within 6 hours. CT IVP negative for injury. Unanticipated anaemia (Hb 60 g/l) with abnormal liver function studies, high LDH and negative coombs. Initial diagnosis HELLP with possible bladder trauma. Transfer to tertiary hospital for urology review. Continued with severe refractory abdominal pain and high transfusion requirement. Renal tract trauma excluded with repeat imaging but found to have portal vein thrombosis. Triggered PNH screen which was positive (78% PNH clone in granulocyte series). Commenced eculizumab with improvement in all symptoms, resolution of haematuria and stabilisation of haemoglobin without need for further transfusion. The entire presentation was thought primarily driven by PNH.



Blood film demonstrating Schistocytes consistent with MAHA. (ASH film archive)





Discussion :
TTP more commonly occurs an autoimmune condition with antibody induced severe reduction in ADAMTS-13 which is a metalloprotease vital for post translation modification of high molecular weight VWF. This leads to microvascular thrombosis and MAHA. Diverse neurologic complications are common. Significant renal impairment is unusual early in the disease as is coagulopathy.

PNH is an acquired mutation in the PIG-A gene that leads to the absence and loss of function on blood cells, including CD59, leading to loss of resistance to native complement, with subsequent intravascular haemolysis and free haemoglobin scavenging of nitic oxide with downstream vasospasm, smooth muscle dysfunction and thrombosis (including unusual sites). The clinical presentation can be acute such as in this case, or chronic with abdominal pain and intravascular haemolysis leading to iron deficiency and a tendency to thrombosis.

Practice point:
Although uncommon, TTP can present denovo in the peripartum period and can easily be misdiagnosed as PET/HELLP. All patients with thrombocytopenia, especially severe and associated with any abnormal neurology should have a blood film made and reported urgently to look for schistocytes consistent with MAHA. This can be organised in most local laboratories by the scientists. The PLASMIC score should then be applied. Although not entirely diagnostic, the score does guide which patients should be transferred for urgent plasmapheresis pending definitive diagnosis. TTP is often fatal if misdiagnosed and plasmapheresis not initiated. Conversely, early recognition can lead to excellent outcomes. PNH, also uncommon, can present denovo in the peripartum period with similar overlap in symptoms with PET/HELLP. PNH should be considered in the setting of DAT negative haemolysis associated with any of iron deficiency, unexplained abdominal pain and thrombosis, including unusual sites. Diagnosis can be easily made by ordering flow cytometry to exclude PNH (CD55&CD56). Effective treatment with eculizumab is readily available.