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A retrospective review of suspected thrombotic thrombocytopenic purpura cases from a tertiary referral centre

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INTRODUCTION

- Thrombotic thrombocytopenic purpura (TTP) is a rare and life-threatening diagnosis with a high rate of mortality without prompt treatment.
- Definitive diagnosis relies on demonstrating severe deficiency of the metalloprotease ADAMTS13 using an activity assay .
- This assay is not available in many centres therefore history, examination and standard laboratory investigations guide the decision to commence immediate treatment for suspected TTP.
- Diagnosis is challenging; the 'classic pentad' is present in only 35% of cases, and the clinical presentation can overlap with a number of other conditions.
- Over the past 2 years, University Hospitals Bristol NHS Foundation Trust (UHBT) has seen an increase in referrals for patients with suspected TTP from across the South West of England.
- A retrospective review of referrals for patients with suspected TTP was performed to order to establish final diagnoses and outcomes.

AIM

- The aim of this retrospective review was to gain a better understanding of the actual diagnoses and outcomes of patients who are referred with suspected TTP.
- This information will help the unit estimate what resources will be required for the liaison and laboratory elements of a TTP service in the South West in future.

METHOD

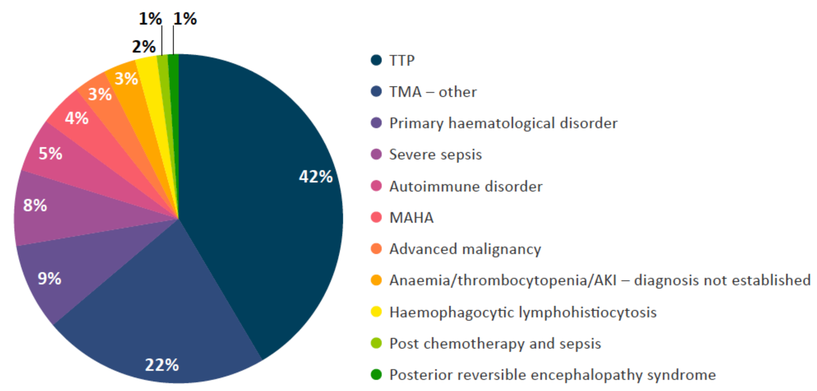
- A retrospective review was conducted of all patients who had an ADAMTS13 activity assays at UHBT over from 01/11/2017 – 01/11/2019.
- Electronic patient notes were reviewed and the following information was gathered:
  - ADAMTS13 activity result
  - Requesting NHS trust
  - Final diagnosis
  - Place of care e.g. intensive care, transfer to tertiary centre
  - Death within 6 months of result
- For those patients with confirmed TTP, further information on clinical status was gathered (e.g. first presentation, relapse, remission status) and aetiology (e.g. immune or congenital)
- Significant effort was made to establish a final diagnosis in all patients, particularly those treated at other NHS trusts.

RESULTS

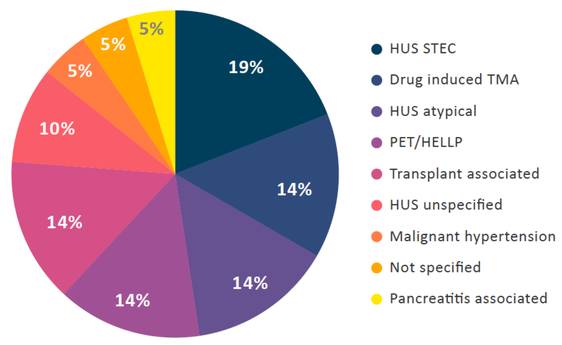
ADAMTS13 assays
- 378 assays were performed on 99 patients (final diagnosis established in 87%):
  - 89% adults
  - 63% females
  - 55% of the tests performed on new patients came from 11 acute NHS trusts, 45% were from patients at UHBT
  - Median number of tests per patient = 1
  - Mean number of test per patient = 3.82.

Outcomes
- 14/33 (42%) of new patients on different sites were transferred to UHBT to commence plasma exchange prior to a result
- 2/14 (14%) were subsequently found to have normal ADAMTS13 activity
- 24/52 (46%) of patients required admission to an intensive care unit during their acute illness.
- 6/88 (7%) patients died within 6 months of the ADAMTS13 request.

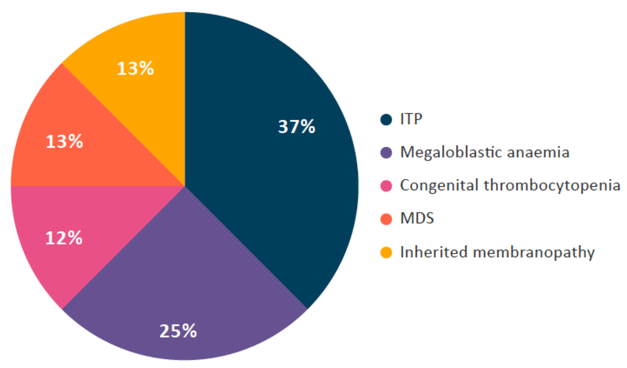
Graph 1: Final diagnosis of patients referred to UHBT with suspected TTP (n=86)



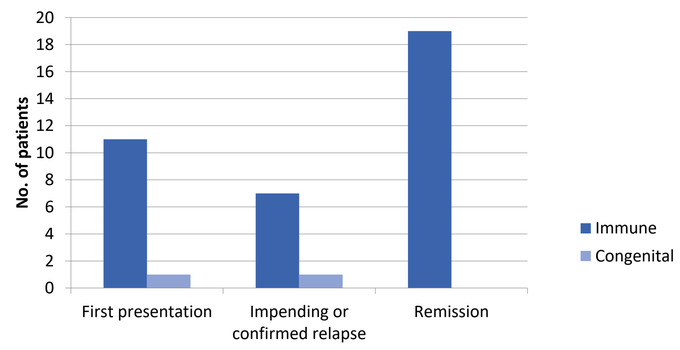
Graph 2: Final diagnosis in patients who were found to have an alternative thrombotic microangiopathy (n=18)



Graph 3: Final diagnosis in patients who were found to have a primary haematological disorder (n=8)



Graph 4: Clinical status of patients with confirmed TTP (n=38)



CONCLUSIONS

- The results reflect the complexities around diagnosis of acute TTP and the plethora of differential diagnoses that need to be considered by specialist treating centres when consulted about these challenging patients.
- As the model of care for TTP moves towards centralisation, clear pathways to initiate dialogue, access diagnostic investigations and link to other specialty disciplines will be essential.
- The team at UHBT will respond to this study by educating all clinicians involved in the diagnosis and management of TTP on the results, strengthening local networks by sharing results and reaffirming referral pathways and supporting resources to provide a regional liaison service.

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REFERENCES

CONTACT INFORMATION

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