The Hong Kong Society of Haematology
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Title	**Incidence and treatment outcome of acquired thrombotic thrombocytopenic purpura in a single institution in Hong Kong**
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Abstract

Introduction

Acquired thrombotic thrombocytopenic purpura (aTTP) is a rare potentially fatal microangiopathic anemia. Hemolytic anemia and organ ischemia in aTTP are caused by acquired severe deficiency of ADAMTS13 (a disintegrin and metalloprotease with thrombospondin type 1 repeats, member 13) due to the presence of autoantibody. In the past, diagnosis of aTTP was made by the presence of clinical pentad. The availability of measurement of ADAMTS13 antigen and activity level and detection of anti-ADAMTS13 antibody significantly improves our accuracy in making the diagnosis of aTTP in recent years. Reported incidence ranges from 1.5 to 6 cases per million adults per year in European and American populations. [1-4] Similar data in Asians is scarce. We therefore conduct a retrospective review on local incidence and treatment outcome of aTTP in one teaching Hospital in Hong Kong over a period of ten years.

Method

Patients' data are collected from our haematology laboratory, apheresis centre and outpatient clinic during the period from January 2014 to January 2024. Patient fulfilling clinical criteria of aTTP (fever, microangiopathic haemolytic anemia, thrombocytopenia, neurological manifestation and/or renal impairment) supported by laboratory investigation results including ADAMTS 13 assay will be included.

Results

There were seven patients diagnosed aTTP over a period of ten years. As we are serving a population of one million, the calculated incidence would be 0.7 case per million per year, which is lower than that reported in Caucasians and other Western populations. Male to female ratio was 5:2 and median age at presentation was 46 (range 35 to 59). Four patients had no precipitating factor identified, while the remaining three had underlying conditions (Sjogren's syndrome, HHV6 viral myocarditis and ischemic heart disease on ticagrelor). Six of the patients had ADAMTS13 assay and all of them had ADAMTS13 activity level below 10 IU/dL (normal range 60.6 -130.6 IU/dL) at presentation. Five of the six patients with ADAMTS13 antibody measured upon diagnosis were positive. Median platelet and haemoglobin at presentation were $15x10^9$ /dL and 8.5g/dL respectively. Two patients had fever, three had neurological manifestations but only one of them had renal impairment. All of them were treated with corticosteroids, plasma exchange and anti-CD20 monoclonal antibody. Median number of sessions of plasma exchange was 10 (range 1-30). Among two of them, splenectomy was performed because of suboptimal response to corticosteroid and anti-CD20 monoclonal antibody. One patient died from fulminant cardiac failure within one day of commencing treatment. The median progression free survival and overall survival of this cohort of patients are 53 months and 57 months respectively.

Limitations

This is retrospective analysis of a single centre. Recall and misclassification bias may affect accuracy of our data analysis.

Reference:

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