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Letter to Editor

Paediatrics Section

Fatal Chronic Immune Thrombocytopenia with Co-existent Type-1 Neurofibromatosis

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Dear Editor,

We would like to update about the follow-up outcome of the child, whose case report was published in this journal [1]. The child was previously managed by the primary unit with consultation of paediatric haematology team. A clinical diagnosis of immune thrombocytopenia with underlying neurofibromatosis was considered and she was managed with oral steroid and discharged on oral prednisolone. She responded well and her platelet count improved (Investigations dated 3rd September 2019 showed Heamoglobin (Hb), total leucocyte count and platelet count being 11.3 g/dL, 8.18×10^3 µL, 256×10^3/µL, respectively). She was discharged but she could not attend haematology clinic on follow-up due to COVID-19 related nationwide lock down. She stopped all her medication and no follow-up blood counts were available for the next 14 months. She was doing fine for nearly one year. She presented again on 27th november 2020 with menorrhagia, severe anaemia and thrombocytopenia (Hb-3.2 g/dL, total leucocyte count- 11.27×103/mL, platelet 2×103/mL). She was admitted under same primary unit and started on injection methylprednisolone 500 mg intravenous (i.v.). In view of poor response to the oral steroid therapy and persistent menorrhagia soon her condition worsened and she developed severe headache and raised Intra Cranial Pressure (ICP).

Intracranial bleed was suspected and she was shifted to paediatric haematology unit for further management and was admitted to paediatric intensive care unit. Her Non Contrast Computed Tomography (NCCT) revealed acute-on-chronic intracranial bleed along with midline shift. She was started on i.v. methylprednisolone 1 gm/day along with i.v. immunoglobulin 2 gm/kg over 24 hours. But her platelet did not improve over next 72 hours. Her blood group was B Rh positive, hence she was also considered for anti-D immunoglobulin but could not be given due to its non-availability in the market [2,3]. She was also given multiple single donor platelet in view of persistent severe thrombocytopenia and persistent headache over next one week, but her platelets never crossed 10000/µL and rapidly fell down to the range of 2000-3000 µL. She was also started on injection romiplastin for persistent thrombocytopenia. Her hepatitis B and C and Human Immunodeficiency Virus (HIV) serology was negative, Antinuclear Antibody (ANA), anti-ds Deoxyribonucleic Acid (DNA) were also negative.

Other causes like malignancy or aplastic anaemia were ruled out with bone marrow examination which were reported as hypercellular marrow with all haematopoietic components and prominence of erythroid precursors and megakaryocytes [4]. In view of persistent thrombocytopenia, she was started on azathioprine and considered for emergency splenectomy [5], but her clinical condition worsened further with recurrence of severe intracranial bleed and succumbed to her illness.

This case highlights the severe nature of the illness and the need to ensure adequate compliance and regular follow-up to avoid catastrophic complication. This case also highlights the untoward effect of COVID-19 related problem faced by patients with chronic illnesses. The association of chronic immune thrombocytopenia with type-1 neurofibromatosis is co-incidental or does it have any true association of such severe immune thrombocytopenia still needs to be established.

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