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Microangiopathic Haemolytic Anemia Associated with Bone Marrow Metastasis in a 12 Year Old Child

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Microangiopathic Haemolytic Anemia

A 12 year old female child presented to the emergency department of our institute with complaints of generalized weakness and lethargy for three months and rapidly progressive breathlessness of 15 days duration. There was history of episodes of hematemesis and melena. Examination was notable for presence of tachypnea, tachycardia, severe pallor, mild pedal edema, right sided pleural effusion, mild hepatomegaly and splenomegaly. Investigations revealed elevated serum lactate dehydrogenase levels, 2127 IU/L, unconjugated hyperbilirubinemia, 3.3 mg/dl, haemoglobin 8.3 g/dl, total leucocyte count, 18×10^9/L, platelet count, 30×10^9/L, and a reticulocyte count of 23.3%. Peripheral blood smear showed numerous schistocytes, polychromasia, circulating nucleated red blood cells (Figure 1A). Coagulogram was normal. Bone marrow aspiration with biopsy showed presence of clusters of non-hematopoietic cells, with signet ring morphology (Figures 1B and 1C) positive for periodic acid Schiffand pan-cytokeratin stains (Figure1C insets). Alfa-fetoprotein (AFP) and carcinoembryonic antigen (CEA) levels were normal. Abdominal ultrasound showed bilateral enlarged ovaries, with solid and cystic components and maintained shape indicative of Krukenberg's tumor. The patient died due to sudden cardiac arrhythmia and further investigations to ascertain the primary site could not be carried out. Autopsy was not performed. A plausible diagnosis of occult gastric primary with dissemination to the ovaries and bone marrow was considered, which is extremely rare at this age.

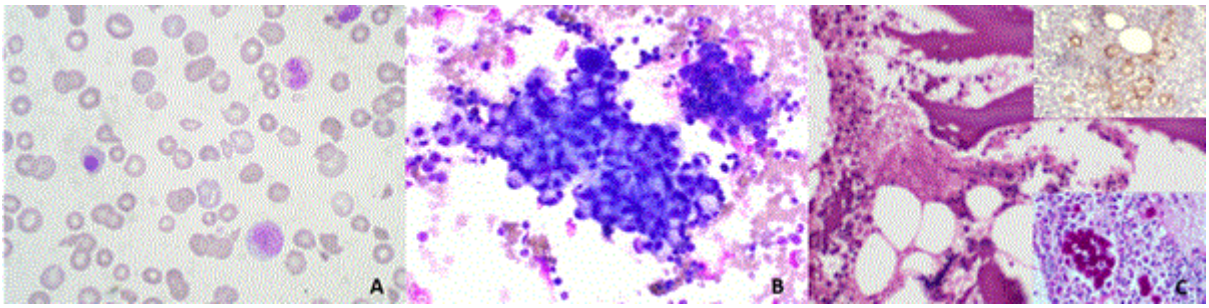

Figure 1: Peripheral blood smear showing numerous schistocytes, nucleated red blood cells, and polychromasia (GiemsaX 1000), (A) bone marrow aspiration and biopsy smears showing presence of clusters of oval shaped cells with eccentric nuclei, abundant vacuolated cytoplasm, consistent with signet ring cells (Giemsa 400X and H and E 1000X), (B and C), inset of C, highlighting positivity for Per-iodic acid Shiff (PAS) stain and pan cytokeratin in the malignant cells.

MAHA is an uncommon yet a fatal para-neoplastic symptom of disseminated malignancies originating from gastro-intestinal tract, breast and prostate [1-3]. In an unusual clinical scenario, presence of MAHA requires a detailed evaluation including a bone marrow examination to detect a malignancy.

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