

ISOLATED RIGHT HAND COMPARTMENT SYNDROME, AN UNUSUAL PRESENTATION OF EVAN SYNDROME

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Introduction: Acute compartment syndrome in Evan syndrome is rare . Acute compartment syndrome is infrequently reported as a complication following bleeding disorder. Evan syndrome is an uncommon hematological disorder that typically associated with anemia and affected individual commonly susceptible to spontaneous bleeding and recurrent infection. There are several case report stated the complication of thrombosis in Evan Syndrome that lead to limb amputation and deep vein thrombosis. However , there are lack of published data regarding acute compartment syndrome in Evan syndrome.

Discussion: This is a case report of 54 years old Malay lady with the underlying Evan syndrome presented with history of spontaneous right hand swelling for 2 days. There were no history of preceding trauma or infection. Clinical examination revealed multiple bruises all over the body. Right hand examination shown swollen over dorsum up to forearm area, tense and blister 1cmx1cm over dorsum area. Tender upon palpation. Passive stretch test positive over dorsal compartment. Range of motion limited over all finger. Otherwise, radial pulse still palpable and comparable, capillary refill time immediate, pulse oximetry all finger all above 95%. Emergency fasciotomy of right hand was done and intraoperatively noted blood clot at subcutaneous layer and surrounding muscles. No sign of infection and distal pulse palpable with good volume. Procedure was uneventful and postoperatively wound was clean and healthy. Tablet prednisolone and cyclophosphamide prescribed during admission. Long term plan is wound closure and split skin graft.

Conclusion: Emergency fasciotomy for acute compartment syndrome remain the standard management even in Evan syndrome case. Although it is not reported elsewhere, Evan syndrome can be presented with acute compartment syndrome. Hence, prompt evaluation and management is crucial to evade permanent damage or loss of the affected limb. High index of suspicion for possibility of acute compartment syndrome are required to avoid delay in diagnosis.