



AB199. A perioperative challenge: acquired von Willebrand disease secondary to IgG lambda monoclonal gammopathy of undetermined significance (MGUS) in oesophageal cancer

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Background: Acquired von Willebrand disease (AVWD) is a rare acquired bleeding disorder with 0.04% prevalence. It can be distinguished from von Willebrand disease (vWD) by lack of personal or familial history of bleeding diathesis. Resulting in reduction of plasma von Willebrand factor (vWF), it can be associated with a number of acquired conditions including haematoproliferative, cardiovascular, autoimmune disorders and neoplasia. Several mechanisms are described, including increased clearance of vWF secondary to autoantibodies, non-specific antibodies, adsorption onto malignant cells and shear stress.

Methods: We present the case of a 53-year-old female presenting with progressive dysphagia, weight loss and haemoptysis, subsequently found to have T3N2 squamous

cell oesophageal carcinoma (SCC). Background history included type I vWD diagnosed post bleed following dental extraction. However, following consultation and thorough work-up from coagulation team, including coagulation studies and serum protein electrophoresis showing a significant IgG paraprotein, was found to have an AVWD secondary to an IgG lambda monoclonal gammopathy of undetermined significance (MGUS) that posed a major perioperative challenge.

Results: Given the limited role for desmopressin (DDVAP) and vAF concentrate in AVWD, peri-procedural treatment with intravenous immunoglobulin (IVIG) and lenalidomide were initiated with good response in von willebrand studies and paraprotein levels. This allowed for safe progression to salvage en bloc 3-stage oesophagectomy post modified FOLFOX neoadjuvant regimen. There was no excess peri-operative blood loss and post-operative course was uneventful.

Conclusions: This case represents an example where close collaboration between specialist surgical and medical teams in a national centre is paramount in allowing for provision of the optimal treatment approach whilst minimising morbidity.

Keywords: Acquired von Willebrand disease (AVWD); oesophageal cancer; monoclonal gammopathy of undetermined significance (MGUS)

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