'Angiodysplasia' of the tongue with acquired von Willebrand's disease

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Summary

A case of acquired von Willebrand's disease is reported in which the patient developed a swelling on the dorsum of the tongue. Histology of this lesion closely resembled that described in angiodysplasia of the colon.

Introduction

Several reports have described an association between von Willebrand's disease and vascular malformations in the stomach and bowel (Ahr *et al.*, 1977; McGrath, Johnson and Stuart, 1979; Ramsay *et al.*, 1976; Rosborough and Swaim, 1978). There are no previous reports of such lesions occurring elsewhere in the gastrointestinal tract.

The authors present a case suggesting that other sites may be affected in a similar manner.

Case report

A 54-year-old man was referred with a 5-year history of episodic melaena. Repeated investigations by gastroduodenoscopy, colonoscopy, barium studies, coeliac axis arteriography and laparotomy had failed to reveal the source of bleeding. Serum electrophoresis 3 years previously had revealed a monoclonal band of IgG specificity and light chain κ ; however, serum immunoglobulin levels were normal, Bence-Jones protein was not detected in the urine and repeated bone marrow examinations had not shown evidence of myelomatosis. For 18 months, the patient had noticed that on the dorsum of the tongue there was a small swelling which was not increasing in size but which occasionally bled.

An inguinal hernia repair 18 years previously had passed without incident but 2 laparotomies in the last year had been complicated by severe bruising around the wounds; in the preceding year, he required repeated suturing following tooth extraction and had abandoned shaving with a razor because he bled easily. There was no history of a bleeding tendency in his family.

Findings on examination were normal apart from a small soft non-ulcerated swelling (6 mm diameter) on the mid-dorsum of the tongue. Investigation

revealed normal platelet count and morphology, prothrombin time 12 sec (control 12 sec), kaolin-cephalin clotting time 31 sec (normal range 30-45 sec), thrombin time 14 sec (control 13 sec), factor VIII coagulant (VIII:C) assay 40% (normal range 50-200%), factor VIII-related antigen (VIII R:Ag) assay 32% (normal range 50-200%) and factor VIII-related von Willebrand factor (VIII:wF) assay 20% (normal range 50-200%). Bleeding time by template was 7.5 min (normal <7 min); platelet aggregation was normal with collagen but defective with ristocetin. Infusion of Factor VIII was followed by a rapid fall of factor VIII:C and factor VIII R:wF to pre-infusion levels by 4 hr with no late rise in activity and the bleeding time shortened from 7 to 4.5 min. These findings are characteristic of acquired von Willebrand's disease.

The appearance of the lesion on the tongue raised the possibility of malignancy and excision was thought advisable. Following preparation by factor VIII infusion the papule was removed without complications from bleeding. Histology showed an angiomatous malformation consisting of large blood-filled chambers lined by endothelium and roofed by squamous epithelium.

Discussion

The patient gave a history strongly suggesting a coagulation defect of recent onset. Acquired von Willebrand's disease is a condition diagnosed by a constellation of features which typically include a mild to moderate bleeding tendency, decreased platelet adhesion to glass beads, reduced or absent platelet aggregation in response to ristocetin, low levels of factors VIII:C and VIII:Ag and absence of a secondary rise in factor VIII:C following the infusion of factor VIII concentrate (McGrath *et al.*, 1979; Rosborough and Swaim, 1978). In most cases of this disorder, an inhibitor molecule (usually IgG) appears to interfere with factor VIII levels (McGrath *et al.*, 1979; Rosborough and Swaim, 1978) and a similar mechanism is likely here. Vascular malformations in the stomach and intestine such as telangiectasia (Ahr *et al.*, 1977;

McGrath *et al.*, 1979; Rosborough and Swaim, 1978) and angiodysplasia (Ramsay *et al.*, 1976) have been described in association with acquired von Willebrand's disease, often associated with severe, recurrent gastrointestinal bleeding. The association may represent a basic endothelial defect as factor VIII is manufactured at this site (McGrath *et al.*, 1979).

This is believed to be the first report of such a malformation on the tongue where the histology closely resembled that seen in vascular ectasia of the colon (Boley *et al.*, 1977). The differential diagnosis included a squamous cell carcinoma. However, recognition of bleeding diathesis and pre-operative correction of factor VIII levels allowed removal of the angioma without incident from an organ noted for its vascularity.

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