The Journal of Laryngology & Otology
April 2002, Vol. **116,** pp. 294–295

An unusual cause of epistaxis: a haemophilic pseudotumour in a non-haemophiliac, arising in a paranasal sinus

D. S. Stevenson, F.R.A.C.S., A. T. Keast, M.B., Ch.B.

Abstract

Most cases of epistaxis are due to simple causes and are easily treated on an out-patient basis. However, there are some cases where the origin of bleeding is not obvious or arises from an unusual pathological source. The authors describe a case of epistaxis due to a mass in the maxillary antrum that when biopsied showed the histological appearances of a haemophilic pseudotumour.

The patient was anticoagulated on warfarin for a cardiac valve replacement and this was thought to be the cause of the ongoing haemorrhage necessary for development of the pseudotumour. Even in haemophiliacs, pseudotumours are rare and we believe this case is unique in that the patient is a non-haemophiliac.

The epistaxis was eventually controlled by external beam radiotherapy to the pseudotumour. The management of this case is outlined as well as a review of the literature on haemophilic pseudotumour.

Key words: Epistaxis; Paranasal Sinus Diseases; Haemophilia; Pseudotumour, Inflammatory

Introduction

Haemophiliacs suffer a number of bone and joint changes with haemarthrosis and contractures being the most common. Pseudotumours are a rare complication first described by Starker in 1918.[1] Pseudotumours are part of a spectrum of haemophiliac cysts. Type I cysts are simple cysts confined solely to the soft tissue within the fascial envelope of a muscle. Type II cysts originate in the soft tissues but eventually give rise to changes in adjacent bones by interfering with the local periostial circulation. Type III cysts are haemophilic pseudotumours, characterized by a soft-tissue mass in proximity to bone with, or without, radiographical evidence of calcification or ossification within the cyst and with evidence of bone destruction and new bone formation.

Haemophilic pseudotumours are easily confused with primary bone sarcomas, metastases, myelomas, aneurysmal bone cysts and osteomyelitis. Untreated these lesions cause local destruction by pressure necrosis and may erode adjacent structures such as vessels, nerves and skin. This leads to pain, fistulae, sepsis and life-threatening haemorrhage. In some cases diagnostic uncertainty has lead to needle and open biopsy techniques, that can cause further complications. The mortality caused by pseudotumours prior to Factor VIII replacement therapy approached 50 per cent, but now the figure is less than 20 per cent.[2]

Pseudotumours are classically described as proximal and distal based on location. Proximal cases are primarily associated with the pelvis and femur and are more common in adults whereas distal cases occur in the hands and feet of children. Other sites have been described in other case reports and include the cranium, mandible, clavicle and orbit.[2,3] There has been only one other case involving the face – the bony nasal pyramid.[1]

The pathogenesis of pseudotumours is not fully understood, theories proposed include: (1) an extension from a haemarthrosis; (2) soft tissue or subperiosteal haemorrhage; (3) cortical or medullary haemorrhage. If there is a history of trauma, any of these mechanisms is possible and is propagated by the coagulopathy present. In our case there is no evidence of trauma and the 'injury' could have been an inflammatory process such as sinusitis.

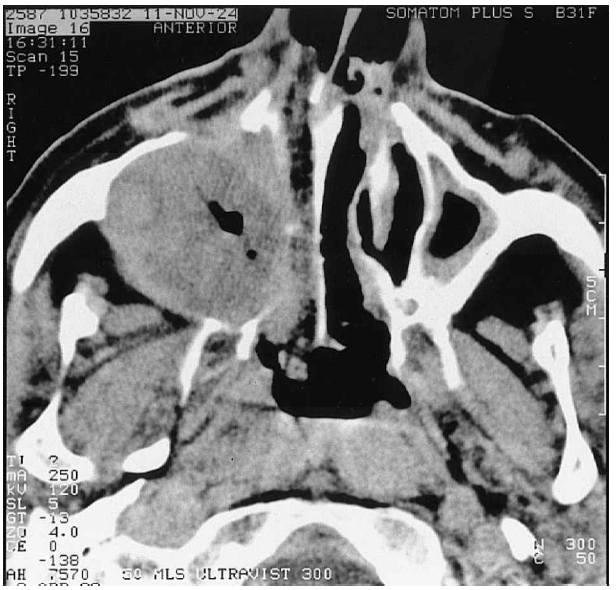
The histological appearance is of old coagulum with fibrous organization, new haemorrhage and regions of bone destruction and/or formation.

Treatment options depends on the ability to reverse the coagulopathy with agents such as Factor VIII replacement. For pseudotumours of the limbs the standard conservative approach of immobolizing the affected region and bed rest is used. A major limitation to this method is the presence of Factor VIII inhibitors which prevents the normalization of the coagulation cascade. Surgical intervention is usually only undertaken when the conservative approach has failed or there is imminent possibility of a life-threatening event. Current methods used include open and laparoscopic drainage of the pseudotumour coupled with filling the cavity with fibrin glue and/or bone grafts. Radiotherapy has been used in a number of cases even though the mechanism of action is not well understood.

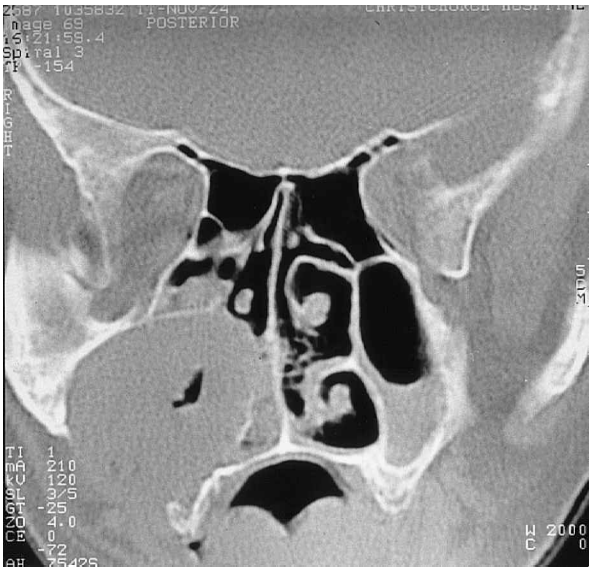
Case report

A 75-year-old Caucasian male presented with persistent right-sided epistaxis of spontaneous onset. He was antic-

From the Department of Otolaryngology – Head and Neck Surgery, Christchurch Hospital, Christchurch, New Zealand.
Presented at The New Zealand Society of Otolaryngology, Head and Neck Surgery, Annual General Scientific Meeting, Christchurch, October 2000.
Accepted for publication: 28 September 2001.



(a)



(b)

FIG. 1

CT scans a) axial and b) coronal of the paranasal sinuses demonstrating the extensive haemophilic pseudotumour centred in the right maxillary antrum.

oagulated on warfarin following mechanical aortic valve replacement and chronic atrial fibrillation, his INR was within therapeutic norms at 2:1. His pertinent medical history included long-term chronic obstructive airway disease (COAD) with progression to cor pulmonale, he required 500 mg of frusemide to control his heart failure.

His first bleed settled spontaneously, examination of his nose was normal and he was discharged. During a second admission three days later an area of abnormal tissue was seen beneath the inferior turbinate, this was biopsied. Histology showed mucus, blood and inflammation with no evidence of malignancy. The bleeding did not settle, a computed tomography (CT) scan (Figures 1 (a) and 1(b)) showed a soft tissue mass of mixed density almost completely filling the right maxillary antrum. There was evidence of bone destruction involving the medial, lateral and anterior walls of the maxilla. Radiologists reported this as consistent with a slowly progressive malignant process, possibly a lymphoma.

Repeated biopsies, and clearance of the abnormal tissue, were then taken via a Caldwell-Luc approach, the specimens yielded tissue suggestive of an organizing haematoma centred on the anterolateral antral wall. The antrum was packed at the end of this procedure to attempt to tamponade the bleeding. This failed and multiple blood transfusions were required. Selective embolization of the external carotid branches was attempted, this was of no benefit.

Following transfer of his care a full review of the histology was undertaken and the diagnosis of a haemophilic pseudotumour was raised. After discussions with colleagues and a literature review, treatment by radiotherapy was felt to be the best option. Following 30 Gy centred on the right maxilla, the bleeding ceased.

Our patient is being followed in the out-patient clinic with no further bleeds over an 18-month period and the antral mucosa looks normal. He has a persistent oro-antral fistula that is closed by his upper dental plate, repair of this has not been attempted.

Discussion

This case shows that not all cases of epistaxis are easy to diagnose and treat. Our patient spent a considerable time in hospital and underwent a number of procedures all with their own morbidity and mortality. We, as the treating physicians, were rapidly running out of options and if the seemingly unuseful histology had been accepted then the outcome may have been potentially fatal. This case demonstrates the need for a review of any facts that do not seem to fit with the clinical picture.

This case is unique in being the only documented case of a haemophilic pseudotumour occurring in a non-haemophiliac and the only case report of a pseudotumour occurring in a paranasal sinus. It shows that this condition should be included in the differential diagnosis for any expanding soft tissue mass in a patient with haemophilia or any other coagulopathy.

References

1 Raj P, Wilde JT, Oliff J, Drake-Lee AB. Nasal haemophilic pseudotumour. *J Laryngol Otol* 1999;**113**:924–7
2 Liu SS, White WL, Johnson PC, Gauntt C. Haemophilic pseudotumour of the spinal canal. *J Neurosurg* 1988;**69**:624–7
3 Merchan ECR. The haemophilic pseudotumour. *International Orthopaedics* 1995;**19**:255–60

Address for correspondence:
Mr D. S. Stevenson,
Department of Otolaryngology–Head and Neck Surgery,
Christchurch Hospital,
Private Bag 4710,
Christchurch,
New Zealand.

Fax: +03 3640273

Mr D. Stevenson takes responsibility for the integrity of the content of the paper.
Competing interests: None declared

https://doi.org/10.1258/0022215021910573 Published online by Cambridge University Press