Oesophageal bleeding from aorto-oesophageal fistula due to aortic aneurysm

Case reports and a review of the literature

H. S. MYERS, W. SILBER

Summary

From pathology data it appears that aortic aneurysm may be the commonest cause of aorto-oesophageal fistula (AOF), but this entity is rarely diagnosed clinically. We report 6 patients, seen during a 5-year period, with aneurysms which initially caused chest pain and minor oesophageal bleeding. The diagnosis of AOF was made before death in only 1 case; surgery was not attempted. This patient and 4 others died when rupture into the oesophageal lumen or wall caused exsanguinating haemorrhage. The 6th patient, who died after prostatectomy without a major haemorrhage, had oesophageal fibrosis localized at the aneurysm; this type of lesion occurs in the development of a fistula. The therapeutic ideal is to forestall fatal rupture by prompt diagnosis and immediate surgery when mild oesophageal bleeding gives warning of fistula formation.

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Exsanguinating oesophageal haemorrhage due to an established aorto-oesophageal fistula (AOF) is almost invariably rapidly fatal. No record has been found of spontaneous recovery with long survival, and only a few patients have been salvaged by surgery.[1-4] Advances in this field must depend largely on prompt action at a relatively early stage of fistula formation, which might be revealed by minor haemorrhages and other premonitory signs.

Most AOFs can be classified as being due to injury, infection, malignant tumour or spontaneous aortic aneurysm. This article deals mainly with oesophageal fistulas and other lesions caused by aneurysms. Pathology studies have shown that AOFs due to aneurysms are not very rare, but most of the clinical papers up to 1981 have reported only single cases.

We report on 6 patients seen at Groote Schuur Hospital within 5 years, who presented with oesophageal bleeding due to aortic aneurysm which ruptured into the oesophageal lumen in 4 cases and caused wall lesions without intraluminal rupture in the other 2. The latest case is presented in detail, and the series is summarized in Table I to illustrate features of the natural history of aortic aneurysms progressing to form AOFs.

Department of Radiology and Gastro-intestinal Motility Clinic, Department of Surgery, Groote Schuur Hospital and University of Cape Town

H. S. MYERS, M.B. CH.B., M.MED. (RAD. D.), *Senior Specialist and Senior Lecturer*

W. SILBER, M.B. CH.B., F.R.C.S., F.A.C.S., F.A.C.G., *Associate Professor and Head of the Gastro-intestinal Motility Clinic*

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Case report

A woman in her 70s was admitted to a peripheral hospital with a complaint of retrosternal and interscapular pain for several days. A small haematemesis had occurred in that time. She had had a myocardial infarction 6 years previously, with complete recovery.

In hospital her general condition was satisfactory. The blood pressure was maintained around 160/85 mmHg, and the haemoglobin value remained steady at 13 g/dl. The stools contained occult blood. A chest radiograph showed a large aneurysm of the descending aorta (Fig. 1a). On the 7th day the patient was transferred to Groote Schuur Hospital for further investigation. On arrival she had a small haematemesis, and packed cells were immediately transfused. She then complained of dysphagia. On oesophagoscopy, at the 20 cm level, blood clot and debris, including food, obscured the mucosa, and the endoscope was not advanced further. A barium oesophagogram showed an erosive lesion in the oesophageal wall adjacent to the aneurysm (Fig. 1b). An AOF was then diagnosed. Shortly thereafter another haematemesis occurred, causing hypovolaemic shock. At that stage surgical opinion was opposed to thoracotomy. The patient was resuscitated, but she had a sudden exsanguinating haemorrhage 22 hours after admission and died almost immediately.

Autopsy findings

The entire aorta was severely affected by ulcerating atherosclerosis. An aneurysm of the descending part, 12 cm in length, had ruptured into the mediastinum, forming a large sac which communicated with the oesophageal lumen through an orifice 1 cm in diameter at the 30 cm level. The heart was mildly enlarged but the myocardium and valves appeared normal. Nephrosclerosis was the only other abnormality found. Histological examination showed extensive atherosclerosis but no aortic dissection, syphilis, tuberculosis or other arteritis. The oesophagus was necrotic at the fistula orifice but otherwise normal. No tumour or infection was detected in the mediastinum or elsewhere.

Discussion

In the course of this century a wide variety of causes of AOF has been reported (Table II). In some cases several factors act together. Sepsis might complicate injury, or might combine with haemodynamic or degenerative processes in causing rupture of the aneurysm. Irradiation, cytotoxic drugs, infection and malignant tumours might all be co-factors.

TABLE I. CLINICAL, OESOPHAGOSCOPIC AND AUTOPSY DATA IN 6 CASES

Patient	Sex	Age	Pre-admission: Pain	Pre-admission: GI bleed	Bleeding (hours after admission)	Oesophagoscopy	Autopsy — oesophagus; mediastinum
1	F	70+	9 d	8 d	1/2, 12, 22 (EH, D)	Blood and food at 20 cm	Rupture at 30 cm; mediastinal haematoma
2	F	69	2 wks	7 d; 12 h	26 (EH,D)	'Ulcer', blood, pulsations, compression	Rupture in lower third
3	F	75	3 mo.	36 h; 6 h	12 (EH,D)	—	Rupture mid-region; mediastinal and retropharyngeal haematoma
4	M	70	4 d	12 h	48 (EH,D)	—	Kinked around aneurysm, mucosa congested, lumen blood-filled
5	M	65	2 mo.	2 mo.	—	—	Local mural fibrosis at aneurysm site
6	F	44	3 d	3 d	30 (EH, D)	'Fungating' pulsatile mass at 15 cm	Rupture at 20 cm

Notes:
Pain — retrosternal and/or interscapular in all cases; patient 5 also had angina.
Dysphagia in patients 1 and 3; cervical pain and hoarseness in patient 3.
All pre-admission bleeds were small.
All chest radiographs showed aneurysm of descending aorta; arch also involved in patient 6.
All aneurysms were arteriosclerotic; additional syphilitic aortitis present in patient 3.
EH, D = exsanguinating haemorrhage, death. (Patient 5 died after prostatectomy.)

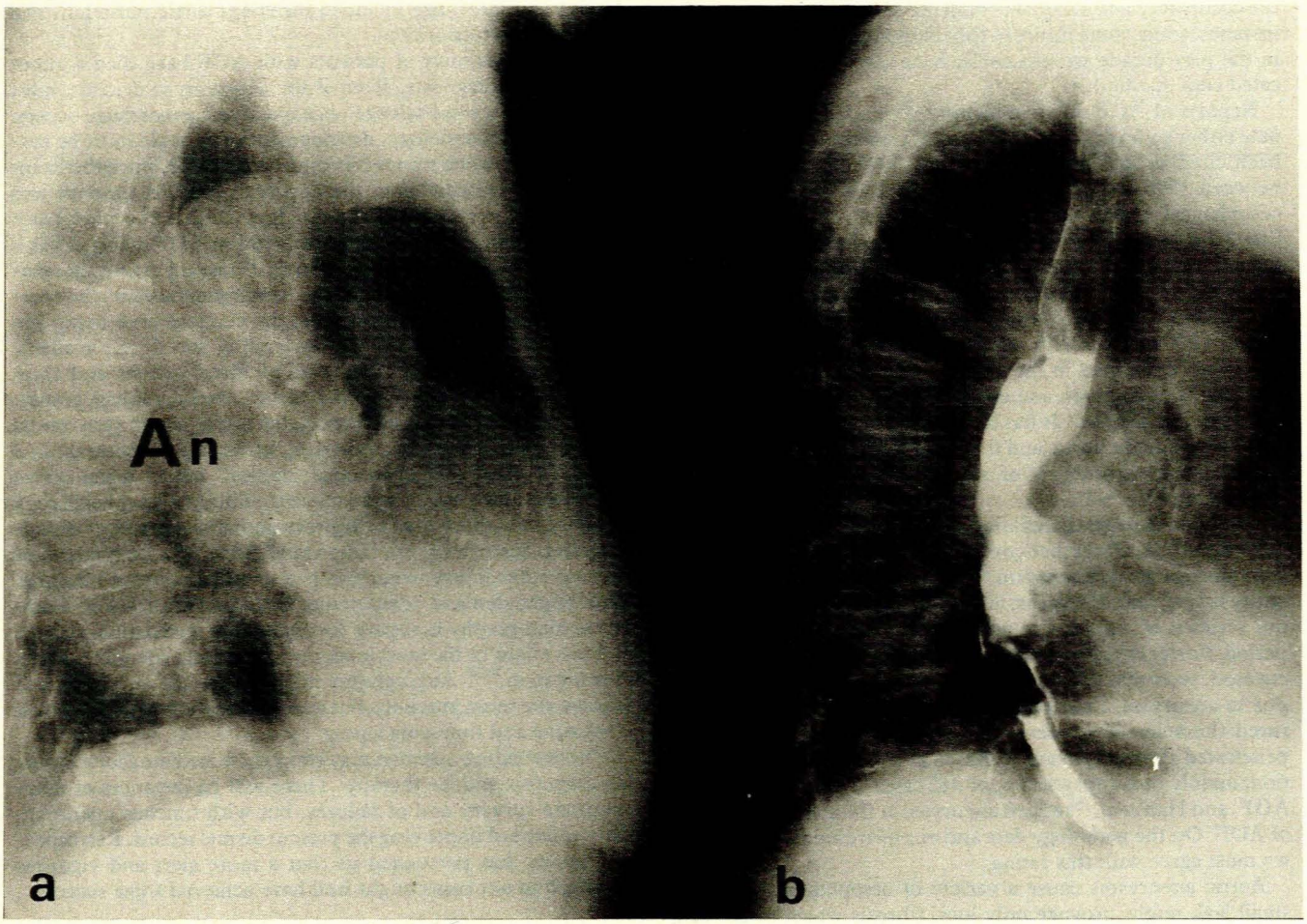

Fig. 1. a — lateral chest radiograph showing large aneurysm (An) of descending aorta; b — barium oesophagogram 8 days later showing erosion of the oesophagus by the aneurysm. At autopsy an AOF was found at the 30 cm level.

TABLE II. CAUSES OF AORTO-OESOPHAGEAL FISTULA, ACTING SINGLY OR IN COMBINATION

Non-traumatic
- **Infection**
  - **Aorta — syphilis,[15,32] tuberculosis ± aneurysm,[26] mycotic,[16] *Mycoplasma*,[27] *Candida*[28]**
  - **Oesophagus — with foreign body, corrosive,[39] peptic ulcer[4,27]**
  - **Mediastinum — tuberculosis,[26] pyogens, *Mycoplasma*,[27] *Candida*[28]**
- **Aneurysm of aorta — arteriosclerotic, infective, dissecting, medial necrosis, idiopathic**
- **Malignancy — carcinoma of lung, oesophagus[5-7]**
- **Peptic ulcer of oesophagus with hiatal hernia,[4,28] with achalasia**
- **Diverticulum of oesophagus[34]**

Traumatic
- **Non-iatrogenic — foreign body ingestion (bone, wood, metal, dentures)[1]**
- **Iatrogenic**
  - **Surgery — oesophageal anastomosis leak;[37] aortic graft leak;[29,30,38] coarctation, ductus, vascular ring resection;[2,31]**
  - **Hufnagel valve erosion into oesophagus[36]**
  - **Instrumentation — Celestin tube, endoscopy**
  - **Irradiation — for malignant tumour, cytotoxic agents[5,6]**
  - **Angiography — aortography[33]**

Authors differ in their personal experience and in calculating the occurrence and order of prevalence of the various causes. For instance, some quote injury as the commonest cause of AOF, but in the past decade we personally have seen only one authenticated case due to this.

Reports of AOF due to a malignant tumour usually incriminate either pulmonary or oesophageal carcinoma. The latter has been cited as causing an AOF in just over 100 cases reported between 1896 and 1978.[5-9] In one group of 145 patients with oesophageal carcinomas 4 developed AOFs.[9] Carter *et al.*[8] found that 3 out of 24 AOFs seen at their medical centre were due to this tumour. On the other hand, AOF was not mentioned in two reviews covering 5 423 cases of oesophageal carcinoma,[10,11] and we have not to our knowledge encountered one instance of AOF in over 1 200 patients with oesophageal carcinoma whom we have personally examined in the past 15 years. Indeed, oesophageal bleeding is uncommon and exsanguinating haemorrhage rarely occurs in this disease.[6] It has been suggested that these tumours seldom invade the aorta, and that fistulas might be due to necrosis following thrombosis of vasa vasorum, infection and/or radiotherapy.[5]

Twenty-one papers containing clinical data, published between 1929 and 1981, altogether covered only 25 cases of AOF due to spontaneous aortic aneurysm.[12-22] This might suggest that this entity is very rare. However, in 1981 Baron *et al.*[23] added 5 cases; our series is of similar size and was encountered over a 5-year period. Carter *et al.*,[8] without stating clinical details or the period covered, recorded that 16 of the 24 AOFs in their patients were due to aneurysm. Four pathology reviews, covering 1 938 ruptured thoracic aortic aneurysms, showed that 187 (9,6%) had penetrated the oesophagus.[12] Baron *et al.*[23] found documentation, mainly in autopsy series, of 242 cases of aneurysm causing AOF, and Han *et al.*[22] ranked aneurysm as the commonest cause of AOF. On the pathology data and our own clinical experience, we must agree with this rating.

Aortic aneurysms cause a variety of oesophageal lesions. A small leak might provoke only local fibrosis which might heal permanently. Alternatively, aortic pressure and pulsations, perhaps with sepsis or injury, may erode the tethered oesophagus at the fibrotic site, thus forming a short fibrous channel between the two lumens. More diffuse lesions also occur, including aortic and mediastinal dissection, with haematoma in a false aneurysm causing ischaemic necrosis and erosion of the oesophageal wall. Blood either penetrates directly from adventitia to the lumen of the oesophagus or dissects the muscular layer, forming a submucosal haematoma before ultimately perforating the mucosa.

The variable natural history of fistula formation accounts for different manifestations. Often catastrophic bleeding and sudden death occur, apparently without premonitory signs. In other cases a clinical triad can be recognized: (*i*) midthoracic pain with involvement of the aortic, oesophageal or mediastinal tissues; (*ii*) single or intermittent mild 'herald' or 'sentinel' haematemesis and/or melaena caused by oesophageal lesions short of complete penetration; (*iii*) after a variable latent interval, which might be due to thrombosis or the tamponade effect of a haematoma, sudden torrential loss of bright red blood occurs, signifying complete fistulization. Massive bleeding also occurs with extensive submucosal dissection.

Our 6 cases illustrate the clinical and pathological features delineated above. All patients had initial chest pain. One developed cervical pain, hoarseness and dysphagia associated with mediastinal haematoma extending into the retropharynx. All patients had initial 'sentinel' bleeding between 36 hours and 2 months before admission to hospital. Five died of exsanguinating haemorrhage 12 - 48 hours after admission. Four of these patients had complete perforation of the oesophageal wall, and 1 had submucosal haemorrhage with blood filling the lumen. The 5th patient, who died following a prostatectomy after a long latent interval without recurrence of oesophageal bleeding, had only localized oesophageal fibrosis. Oesophagoscopic findings in 3 cases at a late stage included bleeding, aortic pulsations and ulceration (fistula orifice).

The vast majority of patients with AOF have died without antemortem diagnosis. Even if the diagnosis is evident, most patients are beyond salvage once massive haemorrhage occurs. Irrespective of aetiology, development of a fistula must be suspected early, when minor bleeding occurs, and, since the latent interval is unpredictable, the approach must be prompt and vigorous to forestall disastrous bleeding which may occur 'at any moment'. Immediately the possibility of an AOF is suspected, a decision must be made about investigations and urgent thoracotomy. This entails assessing the patient's general state, the operability with regard to the aneurysmal site (the arch being less suitable for surgery than the descending aorta), the available facilities and skills, and the value of investigations and their dangers, including the hazard of delay. Oral oesophagography might be useful, rapid and innocuous but to detect early mural involvement it must be thorough, including air contrast and rotational views. An erroneous 'normal' report made on an inadequate study may trap an unwary clinician. Oesophagoscopy might be valuable but has the grave potential dangers of dislodging clot, disturbing the tamponade effect of a haematoma or causing other injury which can precipitate disastrous bleeding. This might demand temporizing balloon compression. Computed tomography is superb for evaluating mediastinal lesions, and this might be the most innocuous, rapid and adequate special investigation.[24,25] Angiography might define the aneurysm and rupture site more precisely, but it sometimes fails to do this and it is invasive and time-consuming.

The overriding question may concern immediate thoracotomy for diagnosis and/or therapy. There will be instances of inappropriate surgery and of failures, but with timeous operation, there is hope of improving the present dismal record. Retrospective review has persuaded us that a more alert and vigorous approach in our cases might well have achieved some success.

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REFERENCES

1. Sloop RD, Thompson JC. Aorto-esophageal fistula: report of a case and review of literature. *Gastroenterology* 1967; **53:** 768-777.
2. Yonago RH, Iben AB, Mack GB. Aortic bypass in the management of aorto-esophageal fistula. *Ann Thorac Surg* 1969; **7:** 235-237.
3. Ctercteko G, Mok CK. Aorto-esophageal fistula induced by a foreign body: the first recorded survival. *J Thorac Cardiovasc Surg* 1980; **80:** 233-235.
4. Montgomery ACV, Chilvers AS. Oesophago-aortic fistula. *Postgrad Med J* 1981; **57:** 380.
5. Söreide O, Janssen CW Jr, Kvam G, Hartveit F. Aorto-oesophageal fistula complicating carcinoma of the oesophagus. *Scand J Thorac Cardiovasc Surg* 1976; **10:** 79-84.
6. Alrenga DP. Fatal haemorrhage complicating carcinoma of the esophagus. *Am J Gastroenterol* 1976; **65:** 422-426.
7. Emamy H, Mehzad M. Carotidoesophageal fistula complicating carcinoma of the esophagus: report of a case. *Am Surg* 1976; **42:** 376-378.
8. Carter R, Mulder GA, Snyder EN, Brewer III LA. Aortoesophageal fistula. *Am J Surg* 1978; **136:** 26-30.
9. Taquino GJ, Joseph GF. Carcinoma of the esophagus: an analysis of 145 cases with special reference to metastases and extensions. *Ann Otol Rhinol Laryngol* 1947; **56:** 1041-1058.
10. Vinson PP. Malignant disease of the esophagus: study of 1 000 cases. *Northwest Med* 1933; **32:** 320-323.
11. Appelqvist P. Carcinoma of the esophagus and gastric cardia. *Acta Chir Scand* 1972; suppl 430: 1-92.
12. Hooper WL. Haematemesis and melaena due to rupture of a saccular aneurysm of the aorta into the oesophagus. *Postgrad Med J* 1962; **38:** 297-301.
13. Patrick CV, Taylor JF. A case of dissecting aneurysm of the aorta. *Lancet* 1929; **1:** 181-182.
14. Pewters JT. Rupture of aortic aneurysm into esophagus. *JAMA* 1955; **158:** 587.
15. Gresham GA, Johnson HRM. Rupture of aortic aneurysms into the gut. *Br Heart J* 1959; **21:** 518-522.
16. Ferguson MJ, Arden MJ. Gastrointestinal hemorrhage secondary to rupture of aorta. *Arch Intern Med* 1966; **117:** 133-140.
17. Komorowski J, Machura B. Spontaneous rupture of aortic aneurysm into the esophagus. *Bull Polish Med Sci Hist* 1966; **9:** 24-25.
18. Schramek A, Weisz GM, Erlik, D. Gastro-intestinal bleeding due to arterio-enteric fistula. *Digestion* 1971; **4:** 103-108.
19. Clinicopathological Conference. Case of leaking aortic aneurysm with oesophageal fistula. *Br Med J* 1976; **2:** 567-571.
20. Sinar DR, DeMaria A, Kataria YP, Thomas FB. Aortic aneurysm eroding the esophagus: case report and review. *Am J Dig Dis* 1977; **22:** 252-254.
21. Roth JA, Parekh MA. Dissecting aneurysms perforating the esophagus. *N Engl J Med* 1978; **279:** 776.
22. Han SY, Jander HP, Ho KJ. Aortoesophageal fistula. *Southern Med J* 1981; **74:** 1260-1262.
23. Baron RL, Koehler RE, Gutierrez FR, Forrest JV, Weyman PJ. Clinical and radiographic manifestations of aortoesophageal fistulas. *Radiology* 1981; **141:** 599-605.
24. Faling LJ, Pugatch RD, Robbins AH. Diagnosis of unsuspected esophageal perforation by computed tomography. *Am J Med Sci* 1981; **281:** 31-34.
25. Baron RL, Levitt RG, Sagel SS, Stanley RJ. Computed tomography in the evaluation of mediastinal widening. *Radiology* 1981; **138:** 107-113.
26. Robbs JV, Bhoola KD. Aorto-oesophageal fistula complicating tuberculous aortitis. *S Afr Med J* 1976; **50:** 702-704.
27. Champion MC, Sullivan SN, Coles JC, Goldbach, M, Watson WC. Aortoenteric fistula. *Ann Surg* 1982; **195:** 314-317.
28. Lefkowitz M, Elsas LJ, Levine RJ. Candida infection complicating peptic esophageal ulcer: infection in an aortic-esophageal fistula. *Arch Intern Med* 1964; **113:** 672-675.
29. Seymour EQ. Aortoesophageal fistula as a complication of aortic prosthetic graft. *Am J Roentgenol* 1978; **131:** 160-161.
30. Smaha LA, Klima T, Leatherman LL. Aortoesophageal fistula: late complication after repair of thoracic aortic aneurysm. *JAMA* 1978; **240:** 2077-2078.
31. Bigge T, Rothnie NG. Aorto-oesophageal fistula: a late complication of a resected coarctation. *Br J Surg* 1974; **61:** 545-546.
32. Calenda DG, Uricchio JF. Gastrointestinal hemorrhage due to rupture of aortic aneurysm into the esophagus. *JAMA* 1953; **153:** 548-549.
33. Dale HT, Thomson K, DeWeese JA, Popio KA. Aortic rupture into the esophagus during angiography. *JAMA* 1978; **239:** 1880-1881.
34. Powell MEA. A case of aortic-oesophageal fistula. *Br J Surg* 1957; **45:** 55-57.
35. MacPherson DJ, Thompson WR. Aortic esophageal fistula secondary to achalasia accompanied by megaesophagus and esophageal ulceration. *N Engl J Med* 1958; **259:** 1027.
36. Kittle CF. Aorto-esophageal fistula: a late complication following insertion of a Hufnagel valve. *J Thorac Surg* 1958; **36:** 44-48.
37. Ullman AS, Shire KJ, Horn RC. Aorto-esophageal fistula: an unusual complication of esophago-gastrostomy following resection for carcinoma of the esophagus. *Can Med Assoc J* 1961; **85:** 27-31.
38. Strug BS, Saltzman DA, Feldman MI, Sabbagh AH. Aortoesophageal fistula. *Cardiovasc Res Cent Bull* 1979; **18:** 34-38.
39. Ottosson A. Late aortic rupture after lye ingestion *Arch Toxicol* 1981; **47:** 59-62.