HAEMATEMESIS AND MELÆNA DUE TO RUPTURE OF A SACCULAR ANEURYSM OF THE AORTA INTO THE OESOPHAGUS

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RUPTURE of an aortic aneurysm is a rare cause of hæmatemesis and melæna. Rupture into the œsophagus is particularly rare, and when it does occur, the ætiology is usually syphilitic and the outcome immediately fatal. The following case is of interest because the patient survived for 10 days after the initial hæmatemesis and the underlying cause was shown to be medio-necrosis of the aorta.

Case History

A female patient, aged 63 years, was admitted as an emergency to Llandough Hospital on February 16, 1959. She had had sudden severe pain below the lower end of the sternum and epigastrium while eating her evening meal eight days previously. The pain radiated through to the back and persisted throughout the night. The following day she vomited a small clot of blood and during the week before admission she vomited 'coffee-ground' material on a number of occasions. Her motions were black for three days and on the morning of admission she passed a bulky black stool; after this there was dyspnœa with extreme weakness.

There was no previous history of indigestion, epigastric pain, heartburn, hæmatemesis or melæna. Her bowel actions had previously been normal and there was no history of any cardiac or vascular disorder. During a previous admission to hospital her blood pressure had been recorded as 160/110 mm. Hg.

On examination she was pale and her voice was feeble. She was not shocked and the extremities were warm. There was no clubbing, lymphadenopathy, jaundice, petechiæ, cyanosis or œdema. Temperature 98° F., respirations 20/min. Pulse 104/min., of fair volume and not collapsing. Arterial walls were moderately thickened and blood pressure 120/80 mm. Hg. No præcordial pulsation, apex beat not palpable; heart sounds clear, no murmurs. She had a frequent cough productive of mucopurulent sputum, occasionally streaked with bright red blood. Trachea central, chest expansion fair, with good air entry in all areas and a few scattered rhonchi at both bases. Tongue: furred and brown, with mild halitosis. Abdominal examination: slight tenderness and guarding in the epigastrium only; no viscera or masses palpable. Rectal examination confirmed a typical black tarry melæna stool. The pupils were equal, central, regular and reacted normally to light and accommodation. The plantar responses flexor, all tendon reflexes normal, no sensory loss.

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Laboratory Investigations. On admission the hæmoglobin was 71% (10.6 g.), W.B.C. 10,800/cu. mm. The blood film appeared normal. P.C.V. 32%; E.S.R. 62 mm./hr. (Wintrobe) (36 mm. after correction for anæmia). Blood group: A Rh (D) +, serum Wasserman and Kahn reactions negative. Bacteriological examination of sputum revealed no pathogenic organisms. The o-tolidine test for blood in the fæces was strongly positive.

Treatment. The patient had been treated at home by her general practitioner by bed rest and a light diet. In hospital she was confined to bed, placed on a fluid diet and was given morphine. During the first 24 hours she was transfused slowly with 2 pints of whole blood after which she appeared to be very much improved. On the second evening in hospital she suddenly vomited 16 fluid ounces of fresh blood and collapsed. In spite of all resuscitative measures, including the rapid infusion of plasma, she died—ten days after the onset of her first symptoms.

Autopsy Findings

The body was that of an elderly woman, of good general nourishment. Rigor mortis was present in all limbs, but apart from marked pallor there was no external evidence of disease or injury.

The heart weighed 320 g. and the endocardium and myocardium appeared normal. The valves were likewise normal and the left and right coronary arteries and their main branches were patent and free from atheroma. There were several atheromatous plaques in the ascending aorta, especially so immediately distal to the cusps of the aortic valve and at the origin of the great vessels. In addition there were several circumscribed translucent areas where the thickness of the aorta was greatly reduced. The smallest of these were merely indentations in the intimal surface while the largest measured 1½ x 1 cm. and was situated in the ascending aorta.

There was a small saccular aneurysm measuring 1.3 cm. in diameter which communicated with the aorta through a well-defined circular orifice measuring 7 mm. in diameter. The latter was situated at the junction of the aortic arch and the descending part of the thoracic aorta immediately below and posterior to the origin of the left subclavian artery (Fig. 1). The sac was lined with intima and the aorta was entirely free of atheroma.

The aneurysm had ruptured into the œsophagus through a longitudinal tear 2 cm. in length in the œsophageal wall (Fig. 2). The stomach was distended by a recent blood clot which formed a cast of the viscus. The stomach mucosa was perfectly healthy and there

FIG. 1.—*Anterior View*: The heart has been removed and the aortic arch opened up showing the origins of the arterial branches above and the opening of the aneurysm below and directed posteriorly.

FIG. 2.—*Posterior View*: Showing the longitudinal tear in the œsophagus and the position of the aorta in relation to the œsophagus.

FIG. 3.—*Neck of Aneurysm* ($\times 150$): Elastic stain of entrance to the aneurysmal sac showing the abrupt ending of the elastic fibres of the media.

was no ulceration or scarring of the duodenum. The small intestine was filled with fresh blood and the large bowel contained a considerable quantity of altered blood. The gall bladder showed no abnormality and

contained a small amount of bile only. The liver was pale and the spleen soft and shrunken. Pituitary, thyroid and adrenal glands were normal. Both kidneys were pale but otherwise normal. The uterus contained

several small fibroids showing hyaline degeneration and calcification. The lungs were congested and oedematous and there was blood in the branches of the right main bronchus due to aspiration at the time of the terminal bleed. There was no atelectasis or aspiration pneumonia.

Histological Examination

No significant abnormality was found in the heart, lungs, liver, spleen or kidneys.

Examination of the Aorta. Blocks were taken from ascending aorta, aortic arch, descending thoracic and abdominal aorta (14 in all). Sections were stained with Hæmatoxylin-eosin, Weigert's elastic stain, Mucicarmine and Periodic-acid-Schiff reaction (PAS).

The intima showed a moderate degree of atheroma only, but marked degenerative changes were found in the media throughout the entire length of the aorta. The lesions were focal in distribution and consisted of hyaline and cystic degeneration of the media, resulting in partial and occasionally complete destruction of the elastic, collagen and muscle fibres. In addition, mucoid material stained by Mucicarmine and giving a positive PAS reaction could be detected in the cystic areas and to a lesser degree in the interlamellar spaces of the media. In areas with destruction of the entire media there was definite thinning of the vessel wall. A section taken from the aortic wall near the opening of the aneurysm exhibited a similar extensive area of medio-necrosis with more or less complete destruction of the elastic, collagen and muscle fibres (Fig. 3). None of the numerous sections examined showed any evidence of syphilis.

Differential Diagnosis

The differential diagnosis included gastric or duodenal ulceration, portal hypertension with œsophageal varices, acute œsophagitis and neoplasm of the upper gastro-intestinal tract. The unusual feature in this case was the sudden onset of severe pain during a meal without previous history of heartburn, indigestion or other gastro-intestinal upset. Ramseyer (1956) reported a similar case in which the onset of pain and dysphagia had been attributed by the patient to swallowing a chicken bone. In the present case the sudden pain while eating could also have been due to swallowing a

fragment of bone. Frequent cough was a prominent symptom and the patient's general practitioner had considered influenza as a possible diagnosis when he first saw her. In large aneurysms of the arch of the aorta cough is often a presenting symptom and in this instance it is also probable the cough was due to compression of the left main bronchus by the pulsating aneurysm even though the aneurysm was small. Unfortunately straight X-ray of the chest was not performed, but it would probably have proved of little help in diagnosis in view of the small size of the aneurysm and the absence of general dilatation of the aorta. In the absence of cardiovascular signs, of palpable liver, spleen or other masses in the abdomen, bleeding from a peptic ulcer was the tentative diagnosis until the fatal hæmorrhage. Indeed the signs, symptoms and presentation of this case fully supports the statement made by Osler over half a century ago that 'there is no disease more conducive to clinical humility than aneurysm of the aorta' (Osler, 1909).

Discussion

The incidence of aortic aneurysm is less than it was a few decades ago since the introduction of antibiotic therapy for syphilis. Saccular aneurysms of the aorta are most frequently syphilitic in origin or due to rare causes such as idiopathic medio-necrosis cystica, septic embolus, tuberculosis or trauma.

Rupture of an aortic aneurysm anywhere into the gastro-intestinal tract, without being immediately fatal, is a rare cause of hæmatemesis and melæna. Calenda and Uricchio reviewed the literature in 1953 and found that a total of 55 cases had been reported, the majority of which were secondary to rupture of an *abdominal* aneurysm into the stomach or small bowel. Since then a few more cases have been reported but these also were either immediately fatal or were ruptures at sites other than the œsophagus. Rupture of an aneurysm of the *thoracic* aorta into the œsophagus presenting as hæmatemesis and melæna, with survival for any

TABLE I
CASES REPORTED SINCE 1929 OF RUPTURE OF AN AORTIC ANEURYSM INTO THE ŒSOPHAGUS, AND WHICH WERE NOT IMMEDIATELY FATAL

Author and Date	Sex	Age	Ætiology	Symptomatology
Koppisch (1933)	Male	47	Syphilis	Dyspnœa, pain and fever—2 months; hæmatemesis—several hours.
Watanabe (1936)* Spearman (1936)	Male	19	Trauma	Fatal bleeding of short duration 3 years after a shot in the chest.
Kaplan (1942)	Male	58	Syphilis	Pain—2-3 weeks; hæmatemesis—7 days.
Bernstein (1944)	Male	43	Syphilis	Pain—6 days; hæmatemesis—4 days.
Lutembacher (1949) ..	Female	48	Syphilis	Severe dyspnœa—3 days; hæmatemesis—3 days.
Calenda and Uricchio (1953)	Male	65	Syphilis	Pain of unrecorded duration; hæmatemesis—11 days.
Krudy (1955)	Male	48	Syphilis	Pain—1 day; hæmatemesis—5 days.
Brunner (1956)	Female	37	Septicæmia	Fatal bleeding of short duration following septic abortion.
Ramseyer (1956)	Male	59	Arteriosclerosis	Dyspnœa—8 months; pain—7 days; hæmatemesis—3 days.

* Described in Japanese literature—details not available. Cited by Calenda and Uricchio.

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TABLE 2
PUBLISHED SERIES OF THE OVERALL INCIDENCE OF RUPTURE OF ANEURYSMS OF THE THORACIC AORTA, TOGETHER WITH PERCENTAGES OF ALL SUCH RUPTURES INVOLVING THE OESOPHAGUS

Author	No. of Cases of Thoracic Aneurysm Studied	Number of Aneurysms which Ruptured (Any Site)	Number which Ruptured into the Oesophagus	Percentage of All Ruptures Rupturing into the Oesophagus
Boyd	4,000	1,197	112	9.4%
Lucke and Rea ..	263	51	5	9.8%
Kampmeier ..	633	98	20	20.4%
Lemann	Ruptures only	592	50	11.8%

length of time, appears to be exceedingly rare. Only 10 cases have been reported in the last 30 years (Table 1).

Prior to that, Blumenstätt in 1928 had reviewed eight cases in the literature and reported two cases of his own, making a total of 20 cases in the literature of survival after gastro-intestinal bleeding from a thoracic aneurysm. Blumenstätt described two types of aneurysmal rupture:—

(1) The immediately fatal type in which all layers of the wall of the aneurysm and the oesophagus rupture at the same time causing a massive hæmorrhage. The majority of ruptures are of this type.

(2) The type in which, on the first occasion, the wall of the oesophagus is dissected longitudinally by the force of the blood, and massive hæmorrhage occurs subsequently when the oesophageal mucosa ruptures. Patients who survive for days, weeks or even months after the first symptom of rupture, and who may present with hæmatemesis and melæna, are in this category.

Our patient experienced severe pain behind the sternum with dysphagia when the aneurysmal sac ruptured into the outer wall of the oesophagus. This was followed during the next week by several small hæmorrhages through the mucosa, culminating in a massive hæmorrhage on the tenth day after the first episode. The smaller episodes of bleeding probably occurred from minute erosions through the oesophageal mucosa, and the final massive hæmatemesis resulted from the longitudinal tear in the mucosa which was found at autopsy.

Several authors have analysed series of cases of thoracic aortic aneurysm and their subsequent outcome (Table 2).

Kampmeier (1938) studied the notes of 633

patients with aneurysm of the thoracic aorta and found that pain was the presenting symptom in 40% of cases, dyspnœa in a further 40% and the remaining 20% presented with cough, palpitations, hoarseness or dysphagia. Of the cases in Table 1 pain and/or dyspnœa preceded hæmatemesis by weeks and even months in some instances. In the majority, rupture occurred in large aneurysms in grossly diseased portions of the thoracic aorta which could be detected by X-ray examination of the chest. Our patient had an unusually small aneurysm, the cause of which was revealed only on microscopy, namely cystic medio-necrosis of the aorta. Although the exact nature of this condition is not fully understood, it is not related to syphilis or to atherosclerosis. It is generally accepted that cystic medio-necrosis can be a predisposing factor in dissecting aneurysms but the case here described could in no way fall into that category. With the present day advances in thoracic surgery it may well have been that this aneurysm could have been treated surgically if the condition could have been diagnosed earlier.

Summary

A patient with a ruptured saccular aneurysm of the aorta, involving the oesophagus, is presented. Death occurred 10 days after the initial gastro-intestinal bleeding. The post-mortem findings and the histological appearances are described. The literature is reviewed.

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THROMBOSIS-IN-SITU OF THE MAIN PULMONARY ARTERIES

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MASSIVE thrombotic occlusions of the main pulmonary arteries is a rare condition occurring perhaps once in five thousand post-mortem examinations. Its clinical recognition is even rarer (Magidson and Jacobson, 1955). Posselt (1909) could find only three examples of complete occlusions of the pulmonary artery in its main branches, and Brenner (1931), six cases of organizing thrombus in the main pulmonary arteries.

Case Report

F.M.L., 51-year-old female patient, was admitted to Guy's Hospital on 4.5.61 with a history of progressive breathlessness and palpitations of four years' duration. A heart murmur had been heard at the age of 7 but although she attended a special school she had led a normal life up to the age of 41. She then sustained an injury to her left leg and had suffered from recurrent swelling in it since. She also suffered accidental injury to the front of her chest and thereafter complained of pain in the chest. For the past ten years she had noticed gradually increasing breathlessness and occasional palpitations and suffered from frequent chest colds. Prior to admission her breathlessness was of such a degree that she was confined to bed.

Examination revealed a thin patient with cyanosed mucous membranes and early clubbing of the fingers. The jugular venous pressure was raised 4 cm. above the sternal angle and well marked 'a' and 'v' waves were present. The apex beat was in the 6th space 1 in. outside and the mid-clavicular line and was diffuse in character. There was a moderate left parasternal pulsation. A pansystolic murmur was audible at the apex and left sternal edge with a soft ejection murmur in the pulmonary area. The second sound in the pulmonary area was accentuated and rather widely split. There were no abnormal physical signs present in the respiratory system. The liver was palpable, 1 finger's breadth below the costal margin.

FIG. 1

There was no peripheral œdema present and no tenderness over the deep veins of the legs.