

Penetrating Atherosclerotic Ulcer Possibly Originating from a Saccular Ductus Arteriosus Aneurysm: a Case Report

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Abstract

Ductus arteriosus aneurysm is a rare condition in adults and is sometime associated with lethal complications, including penetrating atherosclerotic ulcers. Here, we report the case of a 67-year-old patient who presented with persistent dry cough and hoarseness. Computed tomography revealed a saccular aneurysm 50 mm in diameter in the lesser curvature of the distal aortic arch opposite to the origin of the subclavian artery. The patient had no risk factors for atherosclerosis, such as hypertension, dyslipidemia, diabetes mellitus, or obesity. Imaging studies indicated no advanced atherosclerotic lesions in other arteries, including the abdominal aorta, or the coronary and cerebral arteries. Treatment with total arch replacement was successful. A follow-up of 17 months after the operation indicated a complete resolution of all symptoms, except for paramedian left vocal cord paralysis. Pathological examination showed the presence of ulceration in an atheromatous plaque penetrating the intima and media, and the formation of an underlying saccular aneurysm with rupture. This case suggests that penetrating atherosclerotic ulcers may form in ductus arteriosus aneurysms in adults regardless of major risk factors for atherosclerosis.

Key words: Ductus arteriosus aneurysm, penetrating atherosclerotic ulcer, pseudoaneurysm

Introduction

Ductus arteriosus aneurysm (DAA) is primarily described in infants and children and is rare in adults^[1]. DAA is thought to result from altered circulation after birth, weakening of the wall of the ductus arteriosus, or a combination of both factors^[2]. However, the pathogenesis of DAA in adults may not be the same as that in infants. A previous report indicates that adult-type DAA is frequently observed in patients older than 60 years of age, most of whom have a history of hypertension. This suggests that the aging of the arterial wall and atherosclerotic risk factors may be involved in DAA in adults^[3].

Penetrating atherosclerotic ulcer (PAU) was first described by Stanson et al. in 1986^[4] as an atheromatous

ulceration disrupting the internal elastic lamina and the underlying media with accompanying rupture, medial hemorrhage, or pseudoaneurysm formation. Patients with PAU are generally elderly individuals with hypertension and atherosclerosis^[4,5].

Here, we present the rare case of a PAU in a saccular aneurysm in the lesser curvature of the distal aortic arch in a patient with neither atherosclerotic risk factors nor advanced atherosclerosis in other arteries. We also discuss the possible mechanism of PAU formation in the context of adult-type DAA.

Clinical summary

A 67-year-old man was admitted to the Department of Respiratory Medicine at Fukuoka University Hospital for

evaluation of persistent dry cough and hoarseness. The patient had exhibited fever and cough several months ago. He received medication for a few days. Fever fell, but cough was gradually aggravated. Hoarseness had occurred since a few months ago. His past history revealed no hypertension, hyperuricemia, dyslipidemia, or diabetes mellitus. He had no smoking, but drinking habit. Thoracic computed tomography (CT) showed a saccular thoracic aortic aneurysm (Fig. 1A). The patient had 95.4% vital capacity and 88.0% forced expiratory volume in one second, as measured using spirometry. He was referred to the Department of Cardiovascular Surgery for treatment of the saccular thoracic aortic aneurysm. On admission, his height and weight were 161.2 cm and 56.4 kg, and his body mass index was 21.7. Blood pressure was 120/70 mmHg without a bilateral difference and pulse was 60/min regular. An ultrasound cardiogram indicated no asynergy of wall motion. However, there was hypertrophy of the apex, which suggested apical hypertrophic cardiomyopathy. No significant abnormalities were found in the cardiac valves. 3D-CT indicated the presence of a saccular aneurysm 50 mm in diameter in the lesser curvature of the distal aortic arch opposite to the origin of the subclavian artery (Fig. 1B). Clinical imaging examination, including magnetic resonance imaging, coronary and abdominal CT and cervical Doppler ultrasonography, revealed no evidence of advanced atherosclerotic changes in the abdominal aorta, the coronary, cervical, or cerebral arteries. No abnormalities were detected by hematological and biochemical examination. Syphilis examination was performed in

another hospital and had negative results. Total arch replacement with a synthetic graft was successfully performed. Postoperative laryngoscopic examination revealed paramedian left vocal cord paralysis. During the 17 months after the operation, the patient was free from symptoms with the exception of the paramedian left vocal cord paralysis. All tissue specimens were obtained in compliance with a protocol stipulated by the institutional review board of the Fukuoka University Hospital.

Pathological findings

The surgical specimen comprised several pieces of aortic tissue. Figure 2A shows the aortic lesion that was partially excised by the surgeon. Irregular ulcer-like craters were found on the intimal surface upon gross evaluation (Fig. 2A, arrows). As shown in Fig 2B, microscopic observation showed ulceration of the atheromatous plaque associated with the disarray and partial disruption of the media. A saccular aneurysm with rupture was found under the disarrayed media (Fig. 2B). The intimal flap was composed of a disrupted fibrous cap infiltrated by numerous macrophages (Fig. 2B box a, Fig. 2C). The saccular aneurysm was filled with fresh thrombus containing atheromatous gruels and cholesterol clefts (Fig. 2D). The aortic media had disruption of the elastic lamella and fibers (Fig. 2B box b, Fig. 2E). Medial smooth muscle cells were preserved at the disruption site in the aortic media (Fig. 2F). On the other hand, the wall of the saccular aneurysm was composed of fibrous tissue with fragmented and aggregated elastin, although there

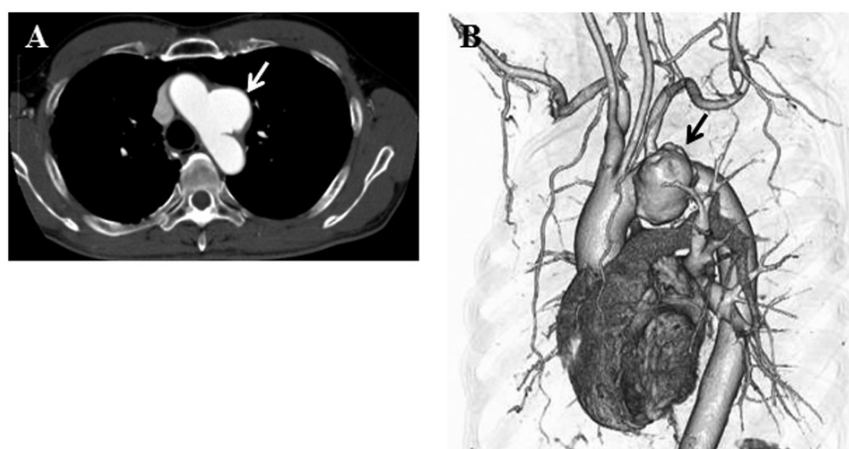


Fig. 1 Thoracic CT scan (A) and 3D CT scan (B). Thoracic CT shows an aneurysm projecting from the aortic arch region (arrow). According to 3D-CT, a 50-mm saccular aneurysm is located at the lesser curvature of the distal aortic arch opposite the origin of the subclavian artery (arrow).

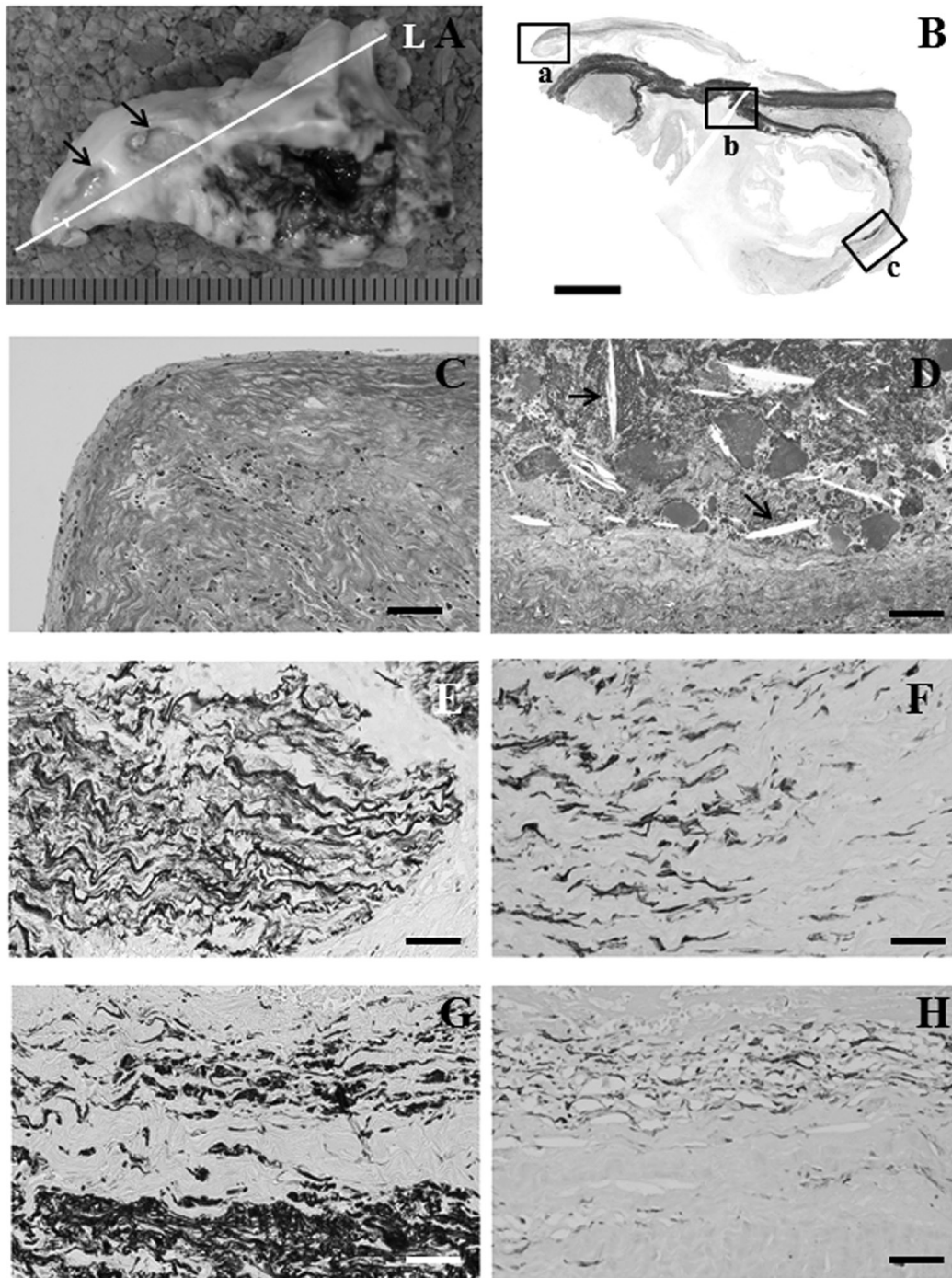


Fig. 2 Pathological examination of the specimen of the aneurysmal wall partially excised by the surgeon. A) Ulcer-like craters (arrows) are noted on the intimal surface upon macroscopic observation. B) The loupe view, indicated by line L in A, showing a plaque ulcer with partially disrupted media and the formation of a saccular aneurysm. Bar = 5 mm. C) Intimal flap, indicated by box a in B, comprises a fibrous cap and is infiltrated by lymphocytes and macrophages. Bar = 100 μ m. D) Fresh thrombus containing atheromatous gruels and cholesterol clefts (arrows) is found within the saccular aneurysm which is made of fibrous tissue. Bar = 100 μ m. E and F) Partially disrupted media of aorta, indicated by box b in B, shows the disruption of the elastic lamella and fibers. Smooth muscle cells are noted between the elastic lamella. Bar = 50 μ m. G and H) The wall of the aneurysm dome, indicated by box c in B, had fragmented and aggregated elastic fibers, but no elastic lamella or smooth muscle cells. Bar = 50 μ m. B, E, and G; Elastica Van Gieson staining; C and D; Masson's trichrome staining; F and H; Immunostaining using anti- α smooth muscle actin antibody.

was no elastic lamella (Fig. 2B box c, Figs. 2D and 2G). Smooth muscle cells were atrophic and disappeared in the wall of the saccular aneurysm (Fig. 2H).

Discussion

Saccular DAA is a rare condition, particularly in adults, and arise at the site of a ductus arteriosus along the lesser curvature of the aortic arch^[6]. On the other hand, saccular aneurysms are commonly found in the cerebral arteries in individuals over 40 years of age^[7]. Degeneration of media, including elastic and smooth muscle cell components, is considered to be a common pathology in saccular aneurysm regardless of its etiology^[8,9]. In the present case, the saccular aneurysm was found in the lesser curvature of the distal aortic arch opposite to the origin of the subclavian artery, and could be considered to be within a ductus arteriosus. Microscopically, the wall of the sac was composed of dense collagenous tissue and was devoid of elastic lamella and medial smooth muscle cells. On the other hand, disrupted elastic lamella and medial smooth muscle cells were found in the disrupted aortic media. These findings suggest that saccular DAA may predispose to the generation of PAU.

PAU develops in advanced atherosclerotic lesions and penetrates through the internal elastic lamina and into the media. In fact, patients with PAU have extensive atherosclerosis in the aorta and other arteries^[5,10]. Atherosclerosis risk factors, such as hypertension, dyslipidemia, obesity, and diabetes mellitus, are often recognized in patients with PAU^[5]. Here we report the case of a patient with an atheromatous ulcer characterized by disruption of media and formation of pseudoaneurysm in the distal aortic arch, which suggests the presence of a PAU. However, the patient had no atherosclerosis risk factors or advanced atherosclerotic lesions in other arteries, including the abdominal aorta, or the coronary, carotid, and cerebral arteries. It is well known that atherosclerotic lesions are frequently found in association with aneurysms, which suggests a role of the hemodynamic stress in atherosclerosis^[9]. Thus, PAU may develop in advanced atherosclerosis found in predisposing saccular DAA regardless of major risk factors for atherosclerosis.

Figure 3 presents the proposed pathological mechanism underlying PAU formation in the present case. An incompletely obliterated ductus arteriosus results in the formation of the diverticulum (Fig. 3A).

Elastic tissue components of the media are disrupted in the wall of the diverticulum^[11]. Together with this medial malformation, hemodynamic stresses may contribute to cause the formation of an aneurysm at the diverticulum (Fig. 3B). The mural thrombus and atherosclerotic lesion are formed within the aneurysm due to alterations in hemodynamic conditions (Fig. 3C). Based on the above-described course for aneurysmal disease, PAU may have developed on the wall of the saccular DAA and led to a pseudoaneurysm (Fig. 3D).

In summary, the present case indicates that PAUs might occur in saccular DAAs and lead to their rupture in adulthood regardless of the presence of major risk factors for atherosclerosis, including hypertension, dyslipidemia, and diabetes mellitus.

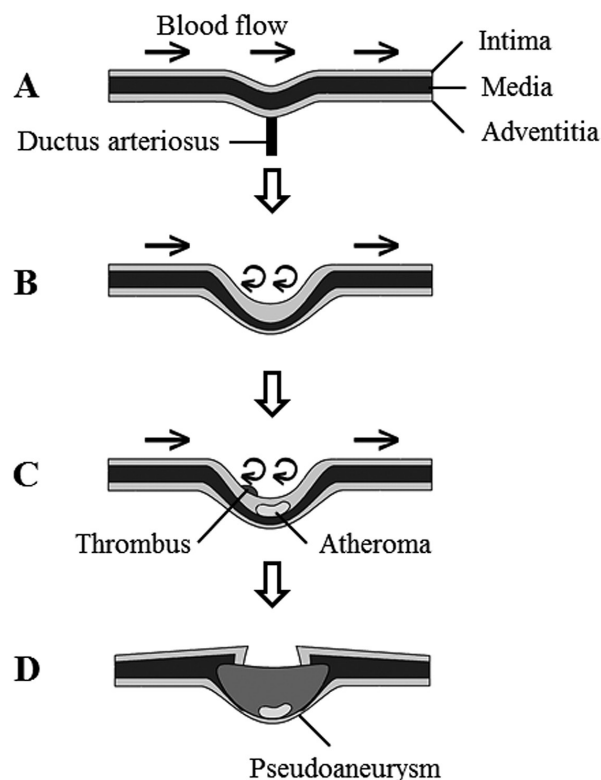


Fig. 3 Proposed mechanisms for the pathogenesis of penetrating atherosclerotic ulcer originating from a saccular ductus arteriosus aneurysm in the present case.

Acknowledgments

This work was supported in part by Grants-in-Aid for Scientific Research from the Ministry of Education, Science, Sports and Cultures of Japan (No. 26420018). The authors thank Ms. Hiroyo Fukagawa for the technical assistance.

Disclosure statement

None declared.

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[The authors declare no conflict of interest.]

