Atrial septal aneurysm—a potential cause of systemic embolism

An echocardiographic study

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SUMMARY Atrial septal aneurysm is an uncommon condition. Between 1981 and 1984 10 cases of atrial septal aneurysm were diagnosed by real time cross sectional echocardiography performed in 4840 patients. The aneurysm was associated either with mitral valve prolapse (three patients) or with atrial septal defect (three patients) or occurred in isolation (four patients, two of whom had had a previous embolic event leading to the diagnosis of atrial septal aneurysm by cross sectional echocardiography). During cross sectional echocardiography the aneurysm appeared as a localised bulging of the interatrial septum, which was best seen in the subcostal four chamber view and in the parasternal short axis view at the level of the aortic root. The aneurysm either protruded into only the right atrium (five patients) or moved backwards and forwards between the right and the left atria during the cardiac cycle (five patients). This motion pattern might be related to changes in the interatrial pressure gradient. The two patients who had had a systemic embolism were given anti-coagulant treatment, but none underwent surgery.

It is concluded that the true prevalence of atrial septal aneurysm might have been underestimated before the routine use of cross sectional echocardiography, that cross sectional echocardiography enables definitive diagnosis of this condition by a non-invasive technique, and that an atrial septal aneurysm should be suspected and looked for by cross sectional echocardiography after an unexplained systemic embolism.

Atrial septal aneurysm is a localised deformity of the interatrial septum which protrudes into the right or the left atrium or both.¹ It might result from bulging of septum primum tissue through the fossa ovalis.² This lesion, which is uncommon, is often unrecognised in asymptomatic subjects.^{3,4} Serious complications such as embolic phenomena may, however, occur.⁵⁻⁹ Cross sectional echocardiography is a non-invasive technique that allows these aneurysms to be diagnosed definitively. With the exception of 11 cases reported by Hauser *et al*,¹⁰ cross sectional echocardiography has been undertaken in only single or a few

cases.^{3,8,11-19} In our study, 10 cases of atrial septal aneurysm were diagnosed in three years using cross sectional echocardiography. The aneurysm was detected after a systemic embolism in two of the 10 patients. The aims of this study were (a) to determine the clinical circumstances leading to the detection of atrial septal aneurysm by cross sectional echocardiography, (b) to assess the prevalence of associated cardiac lesions, (c) to analyse in detail the echocardiographic patterns of atrial septal aneurysm, and (d) to emphasise the need for looking for such aneurysms after unexplained systemic embolism.

Patients and methods

Between March 1981 and January 1984, cross sectional echocardiography was performed in 4840

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Table Clinical and echocardiographic features in patients with atrial septal aneurysm (ASA)

Case No	Sex/age (yr)	Reason for cross sectional echocardiography	Associated lesions	Direction of ASA bulging
1	M/61	Systemic embolism	None	RA to LA
2	M/43	Systemic embolism	None	RA
3	M/53	Systolic murmur	MVP	RA to LA
4	M/39	Systolic click	MVP	RA to LA
5	F/55	Systolic click and murmur	MVP and TVP	RA
6	F/18	ASD	ASD	RA
7	F/49	ASD	ASD	RA
8	F/74	Suspected mitral stenosis	ASD	RA
9	F/71	Chronic dyspnoea	None (EV)	RA to LA
10	F/70	Suspected pericarditis	None	RA to LA

ASD, atrial septal defect; MVP, mitral valve prolapse; TVP, tricuspid valve prolapse; EV, eustachian valve; RA, right atrium; LA, left atrium.

patients. Of these, 10 (0.2%) were found to have atrial septal aneurysm. Four patients were men and six women (age 18–74 (mean 53) years). Systemic embolism was the reason for cross sectional echocardiography in two of these 10 patients. In the remaining eight cross sectional echocardiography was undertaken because of (a) an abnormal systolic murmur or click or both (n=3), (b) a known atrial septal defect (n=2), (c) suspected mitral stenosis (n=1) or pericardial effusion (n=1), or (d) chronic dyspnoea (n=1) (Table).

Cross sectional echocardiography was performed using a mechanical (ATL) or a phased array (Varian 3400, Kontron 250) sector scanner with a 2.25 MHz transducer. Examination was carried out with the patient either semisupine or in the left lateral decubitus position using parasternal, apical, and subcostal views. A simultaneous electrocardiographic lead was recorded. Cross sectional echocardiographic images were recorded on a Panasonic videotape for delayed analysis. M mode echocardiograms were recorded at 50 mm/s using a strip chart recorder. Contrast echocardiography was performed in one patient by rapidly injecting normal saline solution (10 ml) into an antecubital vein.

The diagnostic criterion for atrial septal aneurysm was localised bulging of the interatrial septum, protruding

>6 mm into the right or left atrium or both. Bulging of the entire atrial septum or localised bulging protruding <6 mm was not considered to be a true aneurysm. The aneurysmal deformity always affected the middle part of the interatrial septum—that is, the fossa ovalis area—and was seen in the subcostal or apical four chamber views and in the parasternal short axis view at the level of the aortic root. A significant left to right shunt at the atrial level was suspected if the right ventricle was appreciably enlarged (right ventricular end diastolic diameter to left ventricular end diastolic diameter ratio >0.5) with paradoxical motion of the interventricular septum. Mitral or tricuspid valve prolapse or both was diagnosed in the parasternal long axis or apical four chamber view using previously described criteria.²⁰

Results

The 10 patients with atrial septal aneurysm were classified into four groups according to their clinical findings.

SYSTEMIC EMBOLISM (GROUP 1)

An atrial septal aneurysm was detected in two patients (cases 1 and 2) by cross sectional echocardiography

Fig. 1 (a) Cross sectional echocardiogram in the parasternal short axis view at the aortic root level (case 1) showing the interatrial septum (IAS) bulging into the left atrium (arrow). (b) Simultaneous M mode recording showing an abnormal echo in the left atrium during systole (arrows). AO, aorta.

Fig. 2 Cross sectional echocardiograms in the subcostal four chamber view (case 1) showing the atrial septal aneurysm bulging into (a) the left atrium (LA) during systole (arrow) and (b) the right atrium (RA) during diastole (arrow). LV, left ventricle; RV, right ventricle.

after a history suggesting arterial embolism.

Case 1—A 61 year old man, who had been previously well, was referred to our institution because of a sudden and permanent right hemiplegia. Physical examination and an electrocardiogram were normal. A computed tomogram of the brain showed a low density area in the left frontoparietal region suggesting an ischaemic cerebral infarct. Embolism was suspected in view of the suddenness of the stroke and because of the absence of hypertension. Intravenous digital subtraction angiography of the cervical vessels was normal as was the 24 hour electrocardiographic recording. An atrial septal aneurysm was detected by cross sectional echocardiography. A parasternal short axis view showed systolic bulging of the interatrial septum into the left atrium. A simultaneous M mode recording showed an abnormal echo in the left atrium moving posteriorly during early systole and anteriorly during mid systole (Fig. 1). A subcostal four chamber view showed a thin outpouching in the middle part of the interatrial septum, bulging into either the left atrium or the right atrium throughout the cardiac cycle (Fig. 2). The motion of the aneurysm was more clearly seen on a simultaneous M mode recording. The aneurysm moved from the right atrium towards the left atrium during mid diastole, end diastole, and early systole. It then suddenly moved in reverse from the left atrium towards the right atrium during mid systole, end systole, and early diastole (Fig. 3). Contrast echocardiography showed a filling defect in the right atrium corresponding to the protrusion of the aneurysm. There was no evidence of contrast shunting. Right heart catheterisation showed normal right sided and pulmonary capillary wedge pressures without any intracardiac shunt. Cineangiography was performed with an injection of contrast into the right atrium. It confirmed the presence of a small atrial septal aneurysm distorting the middle part of the interatrial septum and protruding into the left atrium

during atrial early diastole. Anticoagulant treatment was started. The patient had no further emboli during six months' follow up.

Case 2—A 43 year old man was referred after an acute arterial occlusion of the right leg which later resolved. Physical examination and an electrocardiogram were normal. Arteriography showed an abrupt occlusion of both superficial femoral arteries. The popliteal arteries were revascularised through the profunda femoris arteries. A bilateral femoral embolism was then suspected. There was no aortic aneurysm, and a 24 hour electrocardiogram showed no arrhythmia. Cross sectional echocardiography showed an

Fig. 3 M mode echocardiogram in the subcostal view of the aneurysmal motion (case 1) showing the interatrial septum (IAS) moving from the right atrium (RA) towards the left atrium (LA) during mid diastole, end diastole, and early systole and in reverse from the LA towards the RA during mid systole, end systole, and early diastole. Vertical lines emphasize the sudden reversal of the aneurysm from one atria to the other.

Fig. 4 Cross sectional echocardiogram in the parasternal short axis view at the aortic root level (case 6) showing the atrial septal aneurysm bulging into the right atrium (arrow). T, tricuspid valve; AO, aorta.

atrial septal aneurysm bulging into only the right atrium in the apical four chamber view. A right femoropopliteal bypass graft was performed because of persistent rest pain. Anticoagulant treatment was started. No embolic event recurred during four months' follow up.

MITRAL VALVE PROLAPSE (GROUP 2)

Atrial septal aneurysm was detected in association with mitral valve prolapse in two patients (cases 3 and 4) and with tricuspid valve prolapse in one (case 5). In all three cases cross sectional echocardiography was undertaken because of an abnormal systolic murmur or click or both. The atrial septal aneurysm either bulged only into the right atrium (case 5) or undulated alternately between the right atrium and left atrium throughout the cardiac cycle, with the same motion pattern as in case 1 (cases 3 and 4).

ATRIAL SEPTAL DEFECT (GROUP 3)

Atrial septal aneurysm and atrial septal defect were detected in three patients (cases 6, 7, and 8). The aneurysm was associated with a previously known atrial septal defect in two of them (cases 6 and 7). One patient was referred because of suspected mitral stenosis. Cross sectional echocardiography did not confirm the diagnosis but detected atrial septal aneurysm with an associated atrial septal defect. The mean ratio of right to left end diastolic ventricular diameters was 0.86. Interventricular septal motion was always paradoxical. In all three cases the aneurysm bulged only into the right atrium (Fig. 4).

NO SYMPTOMS (GROUP 4)

An atrial septal aneurysm was an unexpected finding in two patients (cases 9 and 10). One patient was referred because of chronic dyspnoea (secondary to carcinomatous lymphangitis) and the second because

of suspected pericardial effusion (which was not confirmed by cross sectional echocardiography). No other cardiac disease was found in these patients. A eustachian valve was, however, present in one patient (case 9). The aneurysm undulated between both atria throughout the cardiac cycle in both patients.

Discussion

An atrial septal aneurysm usually involves the region of the fossa ovalis.^{1,2} It is a localised bulging of the interatrial septum which protrudes into the right atrium or left atrium or both. Its pathogenesis might be explained by an abnormal structure of the interatrial septum or by a change in the normal interatrial pressure gradient or both.⁴ An atrial septal aneurysm is usually considered to be extremely rare. Nevertheless it was detected in 0.2% of 4840 patients in this study and was found at necropsy in 1% of adults by Silver and Dorsey.² The true prevalence of atrial septal aneurysm might therefore have been underestimated in the past for two reasons. Firstly, diagnosis may be overlooked because an atrial septal aneurysm often produces no symptoms. Secondly, no non-invasive diagnostic techniques were available before the advent of cross sectional echocardiography. Sixty three cases of atrial septal aneurysm have been found, of which 36 were reported between 1934 and 1979. A diagnosis was made before death in only seven cases using angiocardiology,^{4-6,9,21,22} whereas the aneurysm was found at necropsy in the remaining cases.^{2,23-29} In contrast, 27 cases were reported between 1978 and 1984 and all were diagnosed before death either by cross sectional echocardiography^{1,3,8,10-19} or more recently by intravenous digital subtraction angiography.⁷ The frequency of diagnosing atrial septal aneurysm during life seems likely to increase in the next few years with the development of cross sectional echocardiography. This non-invasive technique might become the standard diagnostic procedure, though its sensitivity and specificity have yet to be established. The other diagnostic techniques which have been proposed are M mode echocardiography and angiocardiology. M mode echocardiography can detect abnormal echoes within the right or left atrium, but these findings are neither constant nor specific.^{1,15} Angiocardiology is an invasive technique, which does not allow direct visualisation of the interatrial septum. Furthermore, an atrial septal aneurysm may be mistaken for an intra-atrial tumour or thrombus with angiocardiology.^{4,9,22} Intravenous digital subtraction angiocardiology, which is not completely invasive, could, however, improve the detection rate.⁷

Atrial septal aneurysm can be diagnosed definitively by cross sectional echocardiography.¹ The

localised thin and mobile outpouching of the interatrial septum is best visualised in the subcostal four chamber view and in the parasternal short axis view at the level of the aortic root. A minimum radius of 6 mm for the aneurysm was a diagnostic criterion in this study since a small pocket of 3–6 mm long, extending anteriorly and to the left of the limbus fossae ovalis, has been described anatomically in normal subjects.³⁰ The mean value for the radius of the atrial septal aneurysm was 10 mm (range 7–15 mm) in this study. In all cases, the atrial septal aneurysm affected the middle part of the interatrial septum—that is, the fossa ovalis area. In some cases the aneurysm seemed to be wider but it never affected the entire atrial septum. Generalised bulging of the interatrial septum was not considered to be a true aneurysm since such bulging might be seen despite the absence of a true aneurysm.¹ Furthermore, an aneurysm affecting the entire atrial septum has never been documented pathologically.¹ Since no patient in this study underwent surgical or postmortem examination, we cannot confirm that the atrial septal aneurysm was strictly confined to the fossa ovalis area or that it was wider.

Whether or not the aneurysm bulges into the right atrium or the left atrium or both depends on the interatrial pressure gradient.^{4 11} If the pressure gradient is normal, the aneurysm usually protrudes into the left atrium during early systole and into the right atrium during early diastole as shown in Fig. 3.^{11 15} This motion vaguely resembles that of the normal interatrial septum, as described by Tei *et al*.³¹ Five of the 10 patients in the present study had such a motion pattern (three patients with isolated atrial septal aneurysm and two with associated mitral valve prolapse). The aneurysm may bulge into the left atrium in early systole solely during inspiration.¹¹ If the interatrial pressure gradient is reversed because of raised pressure in the right atrium, as seen in tricuspid atresia or hypoplastic right heart syndrome, the aneurysm protrudes into the left atrium.^{12 13 21} Conversely, if the interatrial pressure gradient is increased because of raised pressure in the left atrium, as seen in mitral stenosis, the aneurysm bulges into only the right atrium.^{1 4 22 28} The aneurysm protruded into only the right atrium in five patients in this study, including the three with an associated atrial septal defect, which is in agreement with other reports.^{1 3 5 7} In the two other cases (one isolated aneurysm and one aneurysm associated with mitral valve prolapse) the lack of bulging into the left atrium might be explained either by an unknown increase in the left atrial pressure or by an end expiratory cross sectional echocardiogram.¹¹ Nevertheless, we can only speculate on the relation between the motion pattern of the aneurysm and the interatrial pressure gradient since no patient in this study underwent left heart catheter-

isation. In addition, cross sectional echocardiography allows detection of a shunt at the atrial level using contrast.³²

An interesting finding in this study was the detection of atrial septal aneurysm after an unexplained systemic embolic episode in two of the 10 cases. Only six cases of atrial septal aneurysm associated with embolism have been reported to date. Four cases of cerebral embolism with an angiographically documented atrial septal aneurysm have been reported.^{5 7 8} Other territories have been reported as the site of an embolism resulting from atrial septal aneurysm including the coronary⁶ or pulmonary⁹ arteries. We can only speculate on the relation between atrial septal aneurysm and peripheral embolism in this study. Such a relation should be considered likely only if a previously visualised thrombus within the atrial septal aneurysm after a peripheral embolism has disappeared. Hitherto, cross sectional echocardiography has not visualised such a thrombus in any case of atrial septal aneurysm. The embolic potential of atrial septal aneurysm is, however, supported by previous findings: the presence of a thrombus at the base of the aneurysm at necropsy² or histological evidence of a partly organised thrombus in a resected atrial septal aneurysm.⁵ Another cause of systemic embolism might be paroxysmal arrhythmias, since the triggering role of atrial septal aneurysm in such arrhythmias has recently been suggested.¹⁰ This embolic risk could make what would otherwise have been a simple anatomical anomaly a potentially severe disease. This has led some authors to propose surgical repair of atrial septal aneurysm associated with peripheral embolism in order to prevent the risk of embolic recurrence and to avoid the need for anti-coagulant treatment.^{5 8} Other complications of atrial septal aneurysm have been reported in association only with large aneurysms. They include pulmonary venous obstruction by an atrial septal aneurysm or prolapse of an atrial septal aneurysm through an atrioventricular orifice.^{2 12 29}

In group 2 the atrial septal aneurysm was associated with a mitral valve prolapse. Such an association was found in 30% (3/10) of our patients, whereas it has previously been reported in only two other cases.^{3 16} An abnormal systolic murmur or click or both was heard in all the patients in group 2. Alexander *et al* have suggested that a systolic click might be produced by an atrial septal aneurysm. The sudden reversal of the motion of the aneurysmal bulging from the left atrium into the right atrium during mid systole could explain the click.¹⁵ This hypothesis has, however, been questioned by others, who consider that an associated mitral valve prolapse might cause the click.³³ The association of atrial septal aneurysm and mitral valve prolapse suggests that a myxomatous

degeneration might be responsible for both abnormalities.¹⁶ A slightly redundant interatrial septum might, with age and even without change in the intra-atrial pressures, become aneurysmal.²

In group 3, the atrial septal aneurysm was associated with an atrial septal defect. It is known that an atrial septal aneurysm can contain multiple perforations or fenestrations, resulting in a significant left to right shunting of blood.^{1,3,5} Conversely, the spontaneous closure of an atrial septal defect might result in the formation of an atrial septal aneurysm.¹⁴

In group 4, the asymptomatic atrial septal aneurysm occurred in isolation and was unexpectedly detected by cross sectional echocardiography. A eustachian valve was noted in one patient (case 9). A partial obstruction of the inferior vena cava by such a eustachian valve might result in the blood flow hitting the fossa ovalis area, which might favour the formation of an atrial septal aneurysm.⁴ Isolated atrial septal aneurysms have been previously reported.¹⁵ It must be pointed out that the group 1 patients, who had had an embolic episode, also had isolated atrial septal aneurysm. The management of such asymptomatic and fortuitously detected atrial septal aneurysms is therefore debatable. We believe that further studies are necessary to answer this question.

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