Imaging Features and Outcomes in 10 Cases of Idiopathic Azygos Vein Aneurysm

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Background. **Idiopathic azygos vein aneurysm (AVA) is rare. This retrospective study evaluated the imaging features and outcomes in 10 cases of idiopathic AVA.**

Methods. **We retrospectively evaluated 10 patients with surgically proven or typical imaging features of idiopathic AVA encountered in our institution between 1990 and 2012. Chest roentgenography and computed tomography (CT) were performed in all 10 patients, and magnetic resonance imaging (MRI) was performed in 4 of these patients. The clinical features, AVA morphologic characteristics, and outcomes were analyzed.**

Results. **Chest roentgenograms showed a right paratracheal nodule or mediastinal mass in 7 cases. CT and MRI disclosed 4 thrombosed saccular AVAs (short axis, 3–6 cm; mean, 4.7 cm) and 6 fusiform AVAs (short axis, 2.2–3 cm; mean 2.7 cm). Two large saccular AVAs that presented with chest tightness were resected shortly after diagnosis. One saccular AVA manifested as a pulmonary embolism, whereas the remaining AVA was asymptomatic; they showed 25% to 40% short-axis growth in a 3- to 5-year interval before subsequent AVA resection. Conversely, all 6 fusiform AVAs were asymptomatic and found incidentally, remaining rather stable with less than 8% short-axis growth during 3 to 8 years of follow-up. Compared with fusiform AVAs, saccular AVAs were larger and had a greater frequency of AVA-related symptoms, intralesional thromboses, and greater than 20% short-axis growth during the follow-up period.**

Conclusions. **Saccular AVAs are larger than fusiform aneurysms, presenting with greater frequency of chest symptoms, intralesional thrombosis, considerable lesion growth, and need for surgical intervention. In contrast, fusiform AVAs are asymptomatic and rather stable in long-term follow-up.**

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Dilatation of the azygos vein may be attributed to cardiac decompensation, portal hypertension, or pregnancy, and may also be ascribed to a vena cava congenital anomaly or acquired obstruction secondary to tumor or thrombus. In such circumstances, high blood flow or additional pressure from using the azygos system as a collateral vein may lead to long tubular dilatation of the azygos vein [1–3]. Idiopathic azygos vein aneurysm (AVA) is rare, and fewer than 50 cases have been described [1–14]. The natural history of AVAs has not yet been clarified and their management is still controversial. Some reports have advocated surgical intervention, whereas others have recommended conservative observation [4–14]. This retrospective study evaluated the imaging features, surgical findings, and outcomes of long-term follow-up in 10 cases of idiopathic AVA.

Patients and Methods

Between January 1990 and December 2012, a total of 10 patients with typical imaging features or surgically proven AVA were identified. Their imaging data and medical histories were collected from 3 medical facilities of our institution (1 in southern Taiwan and the other 2 in the north, with a total of 6,000 beds and 15,000 outpatients per day). Our institutional review board approved this study and waived the need for written informed consent from the patients because of the retrospective and anonymous nature of the analysis. The subjects' medical records were reviewed for clinical manifestations, past known diseases, and final outcomes. None of the patients included had any evidence of superior or inferior vena cava malformations or obstruction, azygos or hemiazygos veins anomalies, history of chest trauma, mediastinal infection, portal hypertension, heart failure, recent pregnancy, or constrictive pericarditis.

All 10 patients had undergone chest roentgenographic and computed tomographic studies, and 4 of these patients had also undergone magnetic resonance imaging (MRI). AVA was defined as a focal dilatation of the azygos vein with a short-axis length of at least 2.5 times the diameter of the normal azygos vein at the midthoracic level by CT or MRI. Through modification using the morphologic descriptions of aortic aneurysms [15], AVAs were categorized as saccular type or fusiform type based on CT or MRI findings. A saccular AVA was defined as an eccentric focal dilatation bulging out from a part of the

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azygos vein. In contrast, a fusiform AVA was defined as a circumferential short-segment spindle-shaped dilatation of the azygos vein.

Two of the 10 patients underwent AVA resection on diagnosis. The remainder (8 of 10) were followed with annual or biannual chest roentgenograms. Six of 8 patients received CT or MRI at least once during the follow-up observational period ranging from 3 to 8 years after the original imaging study. Subsequently, 2 more patients underwent AVA resection, whereas another 2 patients were checked with thoracoscopy during operations for suspicious metastatic lung disease and bronchopulmonary sequestration, respectively.

The age, sex, clinical presentation, comorbidities, AVA morphologic features and sizes, surgical findings, AVA short-axis changes measured in follow-up studies (CT or MRI in 6 patients and chest roentgenography in 2 patients), and number of AVAs with short-axis growth greater than 20% during the follow-up period were recorded.

Results

A summary of the 10 cases of AVA is shown in Table 1.

Clinical Features

There were 6 women and 4 men, ranging in age from 33 to 77 years (mean, 54.3 years). Of these 10 patients, patient 1 and patient 2 had chest tightness that could be ascribed to the mass effect of large thrombosed AVAs. Patient 3 had experienced 2 previous episodes of sudden-onset dyspnea and computed tomographic documentation of pulmonary thromboembolism. Because no other deep vein thrombosis could be sought and ultrasonography with Doppler evaluation of the pelvis and lower extremities yielded normal results, the pulmonary emboli were believed to be instigated from the thrombosed AVA. In contrast, the other 7 AVAs were found incidentally on chest roentgenograms or thoracic CT done for other reasons. Of these 7 patients, 1 had had an operation for rectal cancer and a metastatic lung nodule in the left upper lobe, and another patient presented with hemoptysis and suspected bronchopulmonary sequestration in the right lower lobe and also underwent thoracoscopic evaluation for AVA during resection of the lung nodule and sequestrated lung. The laboratory findings were unremarkable in all but 3 patients, including patient 3 who had an elevated D-dimer level, patient 4 who had leukocystosis caused by brachiobasilic arteriovenous fistula infection, and patient 6 who had mild anemia resulting from abdominal blunt trauma and femoral fracture.

Imaging Features

Chest roentgenograms showed right mediastinal masses in 5 patients and right lower paratracheal nodules in 2 patients (Fig 1A). Thoracic CT revealed 4 saccular and 6 fusiform AVAs all confined to the azygos arch, whereas the azygos veins at the mid- to lower thoracic level appeared normal. All 4 saccular AVAs were thrombosed, and 2 manifested as large poorly enhanced right posterior to middle mediastinal masses (patients 1

Table 1. Summary of 10 Cases of AVA

Patient No.	Age (y)	Sex	Clinical Findings	Associated Diseases	AVA Type, Size	Imaging	Operation	Follow-Up Imaging/Duration/ AVA Short-Axis Change
1	33	M	Chest tightness	None	Saccular, 6.5 × 6 cm	CXR, CT, MRI	AVA resection	NA/ NA/6 cm
2	72	F	Chest tightness	None	Saccular, 6 × 5 cm	CXR, CT, MRI	AVA resection	NA/ NA/5 cm
3	55	M	Arteriovenous fistula infection	End-stage renal disease	Saccular, 3.7 × 3 cm	CXR, CT	AVA resection	CXR, CT/5 y/3 to 4.2 cm
4	66	F	Dyspnea, pulmonary thromboembolism	After breast cancer operation	Saccular, 2.8 × 2.4 cm	CXR, CT	AVA resection	CT/3 y/2.4 to 3 cm
5	77	F	Head injury	Diabetes mellitus	Fusiform, 5.5 × 3 cm	CXR, CT	None	CXR/3 y/3 cm, stable
6	56	M	Blunt abdominal trauma, femoral fracture	None	Fusiform, 3.2 × 2.7 cm	CXR, CT	None	CXR/8 y/2.7 cm, stable
7	46	F	Hemoptysis, left upper lung metastasis	After rectal cancer operation	Fusiform, 2.7 × 2.2 cm	CXR, CT	Thoracoscopy, left upper lobectomy	CT/5 y/2.2 cm, stable
8	67	F	Stroke	None	Fusiform, 3 × 2.7 cm	CXR, CT	None	CT/6 y/2.6 to 2.7 cm
9	37	F	Cough, hemoptysis	Bronchopulmonary sequestration	Fusiform, 3.5 × 3 cm	CXR, CT, MRI	Thoracoscopy, right lower lobectomy	CXR, CT/8 y/3 to 3.2 cm
10	34	M	Fever, cough	Drug addict, mediastinal hemangiomatosis	Fusiform 4.5 × 2.5 cm	CXR, CT, MRI	None	CXR, MRI/5 y/2.5 cm, stable

AVA = azygos vein aneurysm; CXR = chest roentgenogram; CT = computed tomography; MRI = magnetic resonance imaging; NA = not applicable.