The Abnormally Situated Azygos Vein

X-Ray Demonstration of its Distention in Congestive Failure and in Various Positions


The azygos vein may be abnormally situated due to anomalous embryologic development. Its abnormal location is infrequent, but radiologic recognition is much easier than that of the normal vein. This vein, normal or abnormal, becomes distended in conditions with an elevated venous pressure, systemic or portal. Changes in the size of the azygos vein on x-ray are noted to parallel fluctuations of venous pressure. We have demonstrated such changes in a patient with an abnormally situated vein during congestive failure, after recovery, and in various physical positions. X-ray demonstration of such changes can have significant diagnostic application.

The recognition of the normal azygos vein in posteroanterior chest films is not easy, because its shadow merges and is often obscured by the prominent densities of the vertebral column and structures at the root of the lung. In 1918, Crane1 first described the "inverted comma sign," but failed to associate it with the azygos vein. Years later, in 1931, Ottonello2 recognized the inverted comma as the arch of the azygos vein and described it fully. Stauffer, Labree, and Adams3 believed that the normally situated azygos vein can be recognized frequently in the right anterior oblique position and that the image of its arch is almost constant in anteroposterior planigrams of the chest at the level of the bifurcation of the trachea. In general, though, it is more often missed than recognized. However, the abnormally situated azygos vein, associated with an azygos lobe of the right lung, is more readily recognized on chest films because it is situated more laterally and because of the pleural "mesentery" of the vein, which divides the right upper lobe.4 On posteroanterior chest films, this "mesentery," or the fissure of the vein, is seen as a curved hairline shadow, at the right apex, with its convexity outward and running obliquely from above the hilus toward the right shoulder.5 At its upper end a small triangular fuzzy peak may be noted; at its lower end is a small pear-shaped or comma-shaped shadow which is thought to represent the arch of the vein projected "end-on."6 The differentiation between the normally and the abnormally situated vein is not difficult. The aberrant vein always has pulmonary parenchyma on each side and is associated with the curved hairline, while the normal vein lies along the right border of the trachea or the right main bronchus completely within the mediastinum.

That distention of the superior and inferior venae cavae and their tributaries results from the increased venous pressure in congestive heart failure is well known. This distention is easily appreciated by the naked eye in the superficial veins of the neck and of the upper extremities. However, in the case of the azygos vein, which is a deep tributary of the superior vena cava, its distention can be recognized only by roentgenologic identification of the enlarged pear-shaped or circular shadow of the arch of the vein. Various authors have described changes in the size of the shadow of the azygos vein in relation to elevated venous pressure and congestive failure.3,4-8 Except for Joselevich and Mactas,6 they have dealt with normally situated azygos veins. These 2 authors reported instances of distention of both the abnormal azygos and the normally situated azygos veins in cardiac failure. Recently, we have observed an aberrant azygos vein in a patient with cardiac failure. It is the purpose of this paper to present this case of pulmonary heart disease in congestive failure with chest films showing
(1) marked distention of the abnormally situated azygos vein while the patient was in rightsided heart failure, (2) disappearance of the distention after recovery from heart failure, and (3) marked distention of the azygos vein with the well compensated patient in the Trendelenburg position.

Anatomy of the Normal and Aberrant Azygos Vein

The azygos vein\(^4, 5, 9, 10\) originates below the diaphragm opposite the first or second lumbar vertebra, usually as the right ascending lumbar vein; occasionally it arises as a branch from the right renal vein or a branch from the inferior vena cava. It enters the thorax through the aortic hiatus in the diaphragm and proceeds along the right side of the vertebral column. At the level of the fourth thoracic vertebra it arches forward over the root of the lung and empties into the superior vena cava just outside the pericardium. It drains the venous blood principally from the chest wall through the intercostal and the hemiazygos veins. It also receives several esophageal, mediastinal, pericardial, and pleural veins. In obstruction of the inferior vena cava, the azygos vein is among the principal channels through which blood is returned to the heart. Through its connections with esophageal veins it also forms part of the collateral circulation of the portal system. Actually, in many cases of advanced portal hypertension, the azygos vein is distended to a large size.

The abnormally situated azygos vein owes its existence to an anomaly in its embryologic development. It is always associated with an azygos lobe of the right lung. The incidence of the latter is not high. Anson reports it as 0.5 per cent in his series.\(^10\) At our hospital its incidence is about 0.15 per cent. In early fetal life the azygos vein runs over the apex of the right lung, but as the lung grows upwards, the vein is normally pulled down by the heart and it finally arches over the root of the lung. If this gliding movement is arrested, the venous loop of the azygos vein indents the lung and produces a deep cleft in the apex of the right upper lobe. As the lung grows farther upwards the vein becomes deeply imbedded in it, drawing down a double fold of both parietal and visceral pleurae. This is often referred to as the "mesentery of the vein" or the "fissure of the vein," and consists of 4 layers of pleura. It should be emphasized that the normally situated vein and its arch lie entirely outside the lung and within the mediastinum, whereas lung parenchyma lies on each side of the arch of the anomalous vein.

Case Report

The patient, a 60-year-old white male coal miner, was first admitted to this hospital in March 1954 for a minor surgical problem. He was found to have anthracosilicosis, emphysema, and chronic bronchitis. Although the electrocardiogram was considered to be probably abnormal, the evidence of heart disease was not conclusive and the patient was not in cardiac failure. The heart was of normal size on x-ray.

The second admission was from January 27 to February 10, 1956, because of right-sided cardiac failure with marked dyspnea, cardiac enlargement, peripheral edema, abnormal electrocardiogram, and fluid at the lung bases. Figure 1 shows the chest film taken the same day. For the first time the azygos lobe was recognized, and the pear-shaped density in the right apex was noted and eventually identified as the arch of an abnormally situated azygos vein. Also seen clearly is the distended terminal portion of the vein. It is interesting to note the pleural fluid

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**FIG. 1.** January 27, 1956, 6-foot erect posteroanterior chest film. The distended azygos vein and the pear-shaped arch of the vein are seen in the first right anterior interspace; the curved hairline shadow in the right apex represents the fissure of the anomalous azygos vein. Pulmonary congestion, fluid at both bases, and enlargement of the heart suggest congestive failure.
along with the distention of the vein, since the parietal pleural veins drain into the azygos through the intercostal veins. By February 7, the patient was much better; he had lost 20 pounds of weight and was practically free of congestive failure. The venous pressure was 7 cm. of water and Decholin circulation time was 16 sec. The chest film at this time (fig. 2) is similar to the one of March 1954.

FIG. 2. February 6, 6-foot erect posteroanterior chest film. The distention of the azygos vein and its arch has disappeared; the heart is much smaller and the fluid at the bases is clearing. At this stage patient was practically free of congestive failure.

FIG. 3. March 7, 6-foot erect posteroanterior chest film. The slight increase in the size of the azygos vein and fluid in the right costophrenic angle suggest mild cardiac failure. A pneumonic process is noted at the left base.

Along with the disappearance of the distention of the azygos vein, the heart became normal in size, and the fluid at the bases practically disappeared.

The third admission was on March 7, 1956, for pneumonia. Physical examination showed signs of a
THE ABNORMALLY SITUATED AZYGOS VEIN

Discussion

Although described in the literature as visible in a large percentage of chest films, in our experience the normally situated azygos vein is not easily recognized. Indeed, many current textbooks on cardiology and x-ray diagnosis do not refer to the normal azygos vein among the structures forming the right cardiac border. Even when this density is recognized as a vascular shadow, its differentiation from an enlarged lymph node is difficult. When distended in heart failure, it may even be more difficult to identify because of concomitant distention of the superior vena cava and pulmonary veins. On the other hand, the aberrant azygos vein is easily seen on posteroanterior chest films. The presence of the azygos lobe and the fissure of the vein make its recognition easier. The curved hairline shadow of the fissure of the vein is easily noted, as well as the comma or the pear-shaped density at its lower end. Both these structures are placed considerably lateral to the right border of the heart and the great vessels, and therefore are clearly visible. The shadow should not be confused with a lymph node or an infiltrative lesion of tuberculosis. It should be kept in mind that the azygos vein becomes distended in situations with an elevated systemic venous pressure such as right-sided heart failure, obstruction of the superior vena cava, or obstruction of the inferior vena cava, as well as in portal venous hypertension. The distention of the azygos vein in obstruction of the inferior vena cava or portal vein is due to the presence of venous anastomoses between the azygos vein and the collateral tributaries of the inferior vena cava and the portal vein.

In the case reported in this paper, an abnormally situated azygos vein became distended during right-sided heart failure and disappeared after recovery. These changes parallel fluctuations of the venous pressure. Distention and its relief were also demonstrated in various positions, again paralleling fluctuations of venous pressure. Distention of an abnormally situated azygos vein, which is readily recognized radiologically, should be considered a sign of increased venous pressure in the superior and inferior venae cavae and the portal venous system.

Summary

The aberrant azygos vein is easily recognizable on posteroanterior chest films, while the normally situated vein is difficult to identify.

A case is reported in which distention of an aberrant azygos vein was demonstrated radiologically during periods of right-sided heart failure. The distention disappeared when the patient recovered from heart failure.

Distention of the vein was demonstrated radiologically in the supine and Trendelenburg positions when the patient was free of heart failure. Distention was more marked in the Trendelenburg position.

Early right-sided failure may be identified by the demonstration of distention of an aberrant azygos vein.

The aberrant azygos vein provides the clinician with an unusual sign that can focus attention upon increased venous pressure in the superior and inferior venae cavae and the portal vein.

Summario in Interlingua

Le aberrante vena azygos es facilmente recognoscibile in pelliculas thoracic posteroanterior. In su sito normal illo es difficile a identificar.
Es reportate un caso in que distension de un aberrante vena azygos eseva demonstrate radiologicamente durante periodos de disfallimento dextero-cardiac. Le distension dispereva quando le patiente se restabliva ab le disfallimento cardiac.

Le distension del vena eseva demonstrate radiologicamente in le position supin e in le position de Trendelenburg quando le patiente eseva libere de disfallimento cardiac. Le distension eseva plus marcate in le position de Trendelenburg.

Precoce disfallimento al later e dextere pote esser identificate per le demonstratione de distension de un aberrante vena azygos.

Le aberrante vena azygos provide al clinico signos inusual que pote traider su attention al presentia de augmentate pressiones venose in le venas cave superior e inferior e in le vena portal.

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Complications appeared in 6 of 650 angiographies. In 1 case extensive cutaneous necroses appeared after injection of oxygen and dye, in 1 case transient paralytic ileus resulted from aortography, and in 2 cases thrombosis and embolism appeared. In 2 cases reversible cerebral damage was seen after aortography or angiography of the subclavian artery.

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