A Family with Lobus Venae Azygos

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Lobus venae azygos was found in five out of 12 members of one family. There is vertical transmission of the abnormality through three generations. The mode of inheritance is probably autosomal dominant.

In 1778, Wrisberg3 for the first time described an anomaly of lobation of the upper lobe of the right lung, usually called lobus venae azygos. The anatomic base for this anomaly is a persisting lateral position of the azygos vein during embryologic development, dividing the lung when the heart descends. Ultimately, it lies at the base of a parietal pleural fold, the mesoazygos septum.

The incidence of lobus venae azygos has been reported by several authors as between 0.07 and 1 percent.4 One of the most recent and largest series of routine chest roentgenograms reported an incidence of 0.7 percent in healthy blood donors.5 In this report, we describe a family with a high incidence of this malformation.

CASE REPORT

The 55-year-old patient was referred to the outpatient clinic of the Department of Pulmonary Diseases for evaluation of hemoptysis. A chest roentgenogram showed a shadow in the left lower lobe and a lobus venae azygos (Fig. 1). Extensive evaluation revealed a squamous cell carcinoma of the left lower lobe, and lobectomy was performed.

Two years later, the patient was feeling well and was without signs of recurrence of tumor. A few months later, his son, aged 21 years, was seen for persistent dyspnea with cough and wheezing caused by allergic asthma. A chest roentgenogram was normal except for a lobus venae azygos.

This rare coincidence led us to examine chest roentgenograms of other members of the family. Of ten other members of the family, we obtained chest roentgenograms made for different purposes (eg, mass or professional annual screening for tuberculosis). In three male relatives the same abnormality was seen (the patient’s father, who died at the age of 71 years of cardiac failure; his brother, aged 36 years; and a son of the latter, aged six years). A pedigree of the family is shown in Figure 2.

DISCUSSION

The lobus venae azygos is a well-known abnormality with three characteristic signs: a triangle at the apex of the lung, connected by a thin line curving downwards and inwards to a “comma shaped” shadow at the level of the second costal cartilage. The clinical significance is minimal. On some occasions, it may cause some confusion, eg, infiltration of it may give rise to the false diagnosis of a substernal thyroid,6 local emphysema suggests a mediastinal herniation,7 and after mediastinal operations, it may resemble mediastinal widening due to bleeding.8

The reason for the occurrence of this abnormality during development is unknown. In one early report a hereditary base was already suggested,9 whereas the presence of a lobus venae azygos in three out of four children of an affected father strongly supported an autosomal mode of inheritance.10 The occurrence in two siblings of apparently normal parents has

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Figure 1A (upper). Chest roentgenogram of patient. B (lower). Detail of Figure 1A with lobus venae azygos.
The association of annuloaortic ectasia and polycystic kidney in 18 consecutive patients who had intravenous pyelograms was 22 percent (4/18). Due to this high association rate, some type of work-up to study the kidney anatomy should be performed in every case of annuloaortic ectasia. For the same reasons, patients with adult polycystic kidney should have a careful cardiovascular evaluation.

Adult polycystic kidney disease is commonly associated with some other abnormalities such as cysts in the liver, pancreas, and spleen. Other well known associated anomalies are aneurysms of the brain arteries, diverticulosis, endocardial fibroelastosis, muscle dystrophy, neurofibroma, and syndactyly.12

Aortic annuloectasia has been related to Marfan's syndrome, aortic coarctation, and dissecting aneurysms. Association of annuloaortic ectasia and polycystic kidney has rarely been described together.4,5 This association has been found in five cases studied by us. The purpose of this article is to call attention to the frequent association of polycystic kidneys in patients with annuloaortic ectasia.

MATERIALS AND METHODS

From January 1975 through December 1984, 60 patients with annuloaortic ectasia were operated upon at the Cardiac Surgery Service of the University of Madrid. Routine cardiovascular studies including cardiac catheterization and aortography were performed in all patients. In the years 1983 and 1984, 18 cases of annuloaortic ectasia were seen. In these 18 cases, intravenous pyelogram (IVP) was visualized at the end of cardiac catheterization. Four cases of adult polycystic kidney were seen through this two-year period, while in the eight-year period prior to routine IVP, only one case of polycystic kidney was diagnosed out of 42 annuloaortic ectasia cases.

DISCUSSION

Aortic annuloectasia may be part of the Marfan syndrome but most frequently, it is an isolated anomaly, without family stigma of Marfan's. Although the Marfan syndrome is considered an hereditary anomaly transmitted through an autosomal dominant gene,16 the penetrance of such a gene may be variable. The cause of the vascular lesion seen in annuloaortic ectasia is unknown, although a hereditary defect of the collagen tissue has been blamed. Possibly in these patients, the continuous replacement of collagen tissue is faulty in quantity, quality, or both. Therefore, the aortic medial layer lacks a proper collagen support rendering the aortic wall.

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Annuloaortic Ectasia and Adult Polycystic Kidney*
A Frequent Association


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