Venous Obstruction of the Upper Extremity Caused by a Malformed Valve of the Subclavian Vein

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We are reporting a case of venous obstruction of the upper extremity, because we believe its etiology to be unusual. An extensive search of the literature failed to disclose any reference to an anomalous venous valve as a cause of obstruction. A number of cases with a clinical picture similar to our patient's have been reported over the years under a variety of headings such as "effort" or "traumatic thrombosis" of the axillary or subclavian veins, and also under the eponym "Paget-Schroetter syndrome."

This syndrome, described at length by Paggi and Roelsen, is limited to the venous system and is thus easily differentiated from the composite arteriovenous and neurologic picture of the "shoulder-girdle syndrome."

Case Report

(J.S., no. 33383). A 39-year-old Puerto Rican man was admitted to the hospital because of progressive enlargement and then paresthesia of his left forearm and arm over an approximately 3-year period.

He denied direct trauma to the left upper extremity or hemithorax, although his work, as a stockroom clerk, entailed frequent lifting of heavy weights. Furthermore, he was right-handed. A diagnosis of "traumatic thrombosis" of the subclavian vein had been made at another hospital 2 years earlier on the basis of a phlebogram. No treatment had been given, but the patient was told that with adequate rest the swelling would subside. However, as the swelling progressed the patient was referred to our hospital.

On physical examination the entire left upper extremity appeared slightly bluish and considerably enlarged. The circumference at various levels measured 1½ inches more on the left arm than on the right (fig. 1). There was no pitting on digital pressure. The superficial veins were distended to the level of the pectoral region. The left external jugular vein was prominent. The column of blood in this vessel bounced after release of pressure over it. Moderate exercise produced pain, and the patient was unable to maintain abduction because of the sensation of heaviness. The venous pressure in the left basilic vein was 7.8 mm Hg, twice as high as that of the right basilic vein (fig. 2). The rest of the physical examination was negative, and the pertinent laboratory investigations were normal.

The patient underwent selective bidirectional phlebographic studies. Under fluoroscopic control a no.7 Goodale-Lubin catheter was passed through the left saphenous vein, the femoral, the inferior vena cava, the right atrium, the superior vena cava, and finally into the left innominate vein. A second catheter was then passed through the left basilic vein into the first portion of the left subclavian vein. Dye was injected through each of the catheters alternately and then simultaneously, while films were taken. These showed an abrupt, well defined, point of obstruction (fig. 3). Both catheters were then advanced gently toward one another. A slight resistance to the progression of the catheters was felt at the point of obstruction previously demonstrated, but this was overcome by simply twisting the catheters, and they went into apposition.

This study was interpreted as evidence of partial obstruction of the left subclavian vein due either to a filiform band compressing the vessel, to a venous "coarctation," or to a "diaphragm" within the lumen of the vessel.

The patient was then explored through a neck incision with removal of the medial half of the clavicle. During this procedure a no.7 Goodale-Lubin catheter was again inserted through the left basilic vein and its tip was positioned at the level of the left subclavian vein proximal to the obstruction. Multiple phleagrams were taken while the vein was freed from each of the possible external compressing structures.

As the obstruction persisted (fig. 4) the subclavian vein was incised and explored. A diaphragm-like structure with a central orifice of about 2 mm. in diameter was found within the lumen of the subclavian and was totally excised. A phlebogram, performed after the subclavian was closed, demonstrated a free flow of dye into the innominate vein (fig. 5). The postoperative course was uneventful.

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The patient was last seen 2 months after discharge; by then he had resumed work and offered no complaints. The left upper extremity was equal in size and color to the right, and exercise caused no discomfort.

Discussion

Since Paget's description in 1875 of an apparently idiopathic thrombosis of the axillary vein, many theories have been advanced to explain insidious venous obstruction of the upper extremity. Paget himself reported his case as an example of "gouty phlebitis," and it was Schroetter who, 9 years later, introduced the concept of trauma to the vein. Later this concept was elaborated to include repeated "microtraumas" to the vein by neighboring anatomic structures, produced by motion of the arm. The repeated microtraumas were believed to cause thrombosis, and this is still the prevalent theory. Difference of opinion centers on the particular anatomic structure responsible for traumatizing the vein. Most muscles and ligaments of the shoulder and of the anterior chest wall, the clavicle, the coracoid process, and the phrenic nerve have been ineruminated by different authors.

Thrombosis and obstruction of the subclavian or axillary vein might well be caused by trauma in several instances, but this need not always be the case. Indeed, surgical exploration has frequently failed to reveal a thrombus. Venous obstruction of the upper extremity may be caused by a multitude of mechanisms, such as ununited clavicular fractures, kinked innominate veins, and abnormal venous valves, as our case demonstrates.

It follows that in any case in which the symptomatology is long lasting, recurrent, and disabling a thorough search for the cause of the obstruction should be made without any
preconceived notions about "microtraumas" or thrombi.

In reviewing the phlebogram obtained elsewhere in our case, we were impressed by the inconclusive nature of this approach. It demonstrated what was already apparent on clinical inspection, namely, the presence of a venous obstruction. We chose to perform selective bidirectional phlebography because this technic yields important information with respect, not only to the exact location of the obstruction, but also to its extent and shape. Selective phlebography was repeated during the surgical intervention as a guide to the procedure.

We were unable to find any reference in the literature to a similar valvular anomaly, and we thus believe ours to be the first reported case of a venous obstruction caused by a malformed valve.

We wonder, however, how many cases in which surgical exploration failed to reveal any pathology might have shown a similar etiology if selective phlebography had been used before and during the operation.

Our patient's valvular pathology was probably congenital, although he first complained of symptoms in adult life. We explain this anachronism by postulating that the central opening of the diaphragm had permitted an undisturbed venous flow until the patient began to engage in heavy physical activity involving the upper extremities.

We would like to suggest that the term "primary venous obstruction" of the upper extremity be used in cases in which the etiology has not been ascertained, rather than labeling them as "traumatic" or "effort thrombosis." These terms are misleading as they imply knowledge of the etiopathogenetic mechanism, encourage diagnostic standstill, and might thus be responsible for inadequate treatment.

Summary

A 39-year-old laborer with a venous obstruction of the left upper extremity was
studied by selective phlebography. Surgery revealed that the obstruction was caused by an anomalous venous valve, diaphragm-like, at the middle third of the subclavian vein. The removal of the anomalous valve resulted in complete relief of the obstruction.

This is the only known case of venous obstruction caused by a malformed valve.

The authors suggest that the term "primary venous obstruction" of the upper extremity be used instead of "traumatic thrombosis" or "Paget-Schroetter" syndrome, in cases in which the etiology has not been ascertained.

References

An Old Gibe

By means of an instrument called the stethoscope, applied to the breast bone, some modern medical writers profess to discover what is going on within. This new art is called auscultation, and the internal sounds pectoriloquism. By means of this new discovery a learned physician professes to have heard a "metallic tinkling" at the heart or in the lungs of a patient. A bystander, however, thinks the "metallic tinkling" was only audible from two pieces of coin which found their way into the palm of the doctor.—Observer, April 1830 (11 years after the appearance of Laënnec's classical work). The Quiet Art: A Doctor's Anthology. Compiled by Dr. Robert Coope. Edinburgh & London, E. & S. Livingstone Ltd., 1952, p. 119.
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