So-Called Primary Venous Obstruction in the
Upper Extremity
Paget-Schroetter Syndrome

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A case is reported of primary venous obstruction in the upper extremity, the Paget-Schroetter syndrome. The literature of this unusual condition is reviewed and the mechanism of its development and its treatment is discussed.

Venous obstruction in the upper extremity is not commonly encountered clinically, and when it does occur it is usually associated with metastatic carcinoma involving the axilla, or from pressure due to mediastinal or lung tumors. A number of cases have been documented in the medical literature where venous obstruction has occurred in the absence of these contributing factors and, as such, a variety of synonyms, such as "effort," "strain," or "traumatic" thrombosis of the axillary or subclavian vein, or of both, have been used to describe it. In addition, Hughes has suggested the term "Paget-Schroetter's syndrome" after the two physicians who first described this condition as a clinical entity. More recently, Lord and Rosati have employed the term "neurovascular compression syndrome of the upper extremity" to indicate abnormal anatomic compression of the axillary and subclavian vessels and associated nerves, irrespective of whether these structures were involved singly or in combination. However, when venous obstruction occurred, it is the principal clinical picture, we prefer the term "Paget-Schroetter syndrome."

Because this condition has received but little attention in the medical literature, it is the purpose of this paper to present a case report of the Paget-Schroetter syndrome recently observed at our hospital and to review the literature related to its etiology, diagnosis, treatment, and prognosis.

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Case Report

The patient, a 23-year-old white man, was first seen for persistent swelling of his right arm. He stated that 3 months previously he noted a sudden swelling of his right upper extremity, associated with mild pain, heaviness, and discoloration of the entire limb. He was not unduly incapacitated and continued to work as a machine operator. In operating the machine, he was required to abduct the right arm to about 160°, and then forcibly push a switch about every 4 minutes. He had been employed at this type of work for about 3 years prior to the onset of his present illness.

On physical examination, the right upper extremity was grossly enlarged when compared with the left arm; definite dilatation of the superficial veins of the right hand, forearm, and upper arm was noted; and slight, but definite, visible cyanosis of the involved extremity was also observed. Palpation was not remarkable, and pitting edema was absent. The circumference of the right forearm was 2 inches greater than the left; the mid-upper arm was 1½ inches greater than its left counterpart. Arm, shoulder, and hand motions were within the normal range. The remainder of the physical examination was negative.

Routine blood studies, coagulation time, prothrombin-time determination, and urinalysis were normal. Roentgenograms of the chest, shoulder, and cervical vertebrae were reported as negative. The venous pressure in the right arm was 60 cm. higher than the left arm. Figure 1 shows an axillary venogram.

Conservative measures were employed in the treatment of this patient, and in 2 weeks temporary improvement was noted. The patient, however, refused surgical treatment.

Review of the Literature

Clinical Findings. As in our case, the majority of patients described with this condition have been men between the ages of 20 to 25 years. In addition, the right arm has
been involved more frequently than the left and, according to Hughes,1 this phenomenon is not related to the right-handed predominance of the general population.

The signs and symptoms of this syndrome are usually quite characteristic and, in most collected cases, sudden nonedematous swelling, discoloration, pain, and prominent cutaneous veins of the involved arm have been noted. In most patients some unusual form of muscular exercise has preceded the onset of symptoms. In addition, the venous pressure of the involved arm has been reported as elevated, principally following exercise. Infrared ray photography has demonstrated an increased collateral venous circulation of the involved limb, particularly over the upper arm and anterior chest wall. Furthermore, a number of authors have shown by venography nonvisualization of the axillary vein, suggesting this vein to be the principal site of obstruction.3-5 Others, however,6, 7 have described the area of obstruction at the subclavian vein level and, in some instances, normal venograms have been described.8

Etiology. The underlying mechanism for the venous obstruction has been the subject of many reports. Earlier writers9 stressed the importance of syphilitic infection; however, it is now considered of little importance. Most authors have directed their attention to the anatomy of the axillary and subclavian veins, with particular interest given to possible sites of compression by contiguous bony and soft-tissue structures.

Lowenstein,10 in 1924, following a detailed anatomic study of the shoulder region found the axillary vein to be compressed by the costocoracoid ligament and subclavius muscle, when the arm assumed a position of hyperabduction and lateral rotation. He suggested that compression of the axillary vein by this maneuver injured the endothelium and thus introduced a site for thrombus formation. Furthermore, he considered the venous stasis that occurred during periods of muscular exertion to be an important contributing factor. It is of interest that Lord and Rosati2 in a recent paper emphasized that this area is a frequent point of obstruction to the axillary vein.

Gould and Patey11 concluded from their anatomic investigation that a sudden contraction of the subclavius muscle compressed the axillary vein, rupturing the delicate underlying intraluminal subclavia-axillary valve and thereby providing a nidus for the development of a thrombus. These authors also stressed the importance of venous stasis as a predisposing factor.

Another possible explanation has been advanced by Veal and McPetridge,12 who showed by venographic examination that compression of the axillary vein by the subscapularis muscle and head of the humerus occurred when the arm assumed the position of hyperabduction. Wright,13 however, demonstrated by anatomic dissection that the arm in hyperabduction stretched and compressed the axillary and subclavian vein at two sites: the interspace between the first rib and clavicle and at the origin of the pectoralis minor muscle and coracoid process.
Falconer and Weddel,\textsuperscript{14} have indicated that hyperabduction of the arm is not the only motion of the shoulder girdle that leads to compression of the vessels and nerves that pass between the first rib and clavicle. They have shown that backward and downward bracing of the shoulders, and hyperextension of the neck, narrows this space sufficiently to compress the underlying artery, vein, and accompanying nerves.

Sampson, Sanders, and Capp,\textsuperscript{15} from a study of patients with prominent arm and chest veins, concluded that this condition was observed principally in deep, broad-chested individuals with soldier-like postures. Furthermore, their patients presented radiographic evidence of prominent first ribs that projected laterally and anteriorly, in conjunction with upward- and backward-directed clavicles. Under these conditions, the space between the first rib, clavicle, and subclavian muscle is sufficiently narrowed to cause compression of the subclavian vein. Sampson\textsuperscript{16} has described cases of the Paget-Schroetter syndrome that illustrated this factor.

More recently, Mc Cleery and his co-workers\textsuperscript{17} have described obstruction of the subclavian vein principally in the area bounded by the scalenus anticus muscle posteriorly, the subclavius muscle and clavicle anteriorly, and the first rib inferiorly. They refer aptly to this area as the "bottleneck" and have described in detail the technic for exploring this region.

A unique hypothesis has been presented by Hughes,\textsuperscript{18} who has suggested that an abnormally placed phrenic nerve, i.e., one that passes posteriorly to the subclavian vein, may be the underlying mechanism. He has postulated that increased diaphragmatic excursions that accompany muscular exertion tense the phrenic nerve which, in turn, compresses the subclavian vein against the adjacent medial border of the scalenus anticus muscle. He has indicated that the taut, bow-string-like action of the phrenic nerve may act as a ligature. Such a theory, he believes, could explain the absence of emboli and reported negative exploratory findings in some of the published case reports.

Less common etiologic factors have been implicated as possible sites of vein compression, namely, bony lesions of the clavicle and complications following fracture of this bone. However, these conditions appear to involve the entire neurovascular bundle rather than the vein per se.\textsuperscript{2} The theory of venous spasm has been emphasized by Ochsner, De Bakey, and others.\textsuperscript{19,20} However, this mechanism would appear to be a complication of the venous obstruction rather than an etiologic factor.

Although considerable emphasis has been directed to the scalenus anticus muscle, with an associated cervical rib, as the underlying mechanism for neuroarterial complication arising from this area, Hughes collected only a few reports where a cervical rib has been found in association with the Paget-Schroetter syndrome.

\textit{Prognosis.} The prognosis for patients with this condition is difficult to evaluate. Hughes\textsuperscript{3} has reported that the majority of patients recover in time; however, slight edema, cyanosis, and prominent cutaneous veins may persist, although they seldom cause any great degree of discomfort. He states that pulmonary emboli are rarely, if ever, encountered, and no death has occurred from this entity. Nevertheless, a number of documented cases have been published where symptoms have been severe and disabling. In this regard, Kleinsasser\textsuperscript{21} has reported that 75 per cent of patients with the Paget-Schroetter syndrome have some residual effects.

\textit{Treatment.} The treatment of this condition is far from standardized. Almost all the authors stress the importance of minimizing the formation of edema by elevation and rest of the arm. Hughes has suggested anticoagulant therapy to limit the propagation of the thrombus and reduce the degree of venous obstruction, and Jones\textsuperscript{22} believes that caution should be exercised in permitting the patients to return to their usual form of occupation, especially if the mode of employment appears responsible.

Mc Cleery and others\textsuperscript{2,17} have suggested surgical exploration of the axillary and subclavian vein. In their patients obstruction of
the axillary or subclavian veins was extravascular in nature, and relief of symptoms was prompt following its relief. According to Hughes, surgical removal of the thrombus has been described and, in some instances, favorable results have been obtained. Additional surgical measures have been reported, namely, cervical sympathectomy and excision of the obstructed vein segment. In each instance, alleviation of symptoms has been reported; however, the cases that have been treated in this manner have been few in number.

DISCUSSION

From this account it will be seen that the current tendency is to attribute the underlying mechanism of the Paget-Schroetter syndrome to direct interference with the venous flow of the arm (with or without thrombus formation) by extravascular compression of the axillary or subclavian vein. It is also clear that the anatomic site responsible for the venous compression may be variable, a point that requires emphasis, particularly if surgical intervention is decided upon.

The treatment of this condition has been primarily conservative in nature and not unlike that employed for deep venous obstruction of the lower extremity. If our concept regarding this type of venous obstruction is correct, it would seem reasonable to advocate early surgical intervention to remove the extravascular venous obstruction and the axillary or subclavian venous thrombus, if present. Anticoagulant therapy would also appear to be an additional useful therapeutic measure.

However, as the majority of cases are observed days after the onset of symptoms, the fact remains that a venous thrombus, if present, would be so organized as to preclude its surgical removal. Under such conditions, a reasonable course of conservative therapy would seem advisable, with attention directed to methods of controlling edema and avoiding motions of the arm that initiated the process. If symptoms persist, surgical exploration of the infraclavicular and supraclavicular spaces should be performed with the idea of removing the source of the venous compression.

However, it should be done with the full understanding that the venous compression point may be variable and, as such, a thorough exploration of the axillary and subclavian veins is necessary. Even though the most consistent site of venous obstruction appears to be the region where the axillary vein passes between the costocoracoid ligament, the subclavian muscle and the first rib, a diligent search must be made for additional compression points that have been described.

SUMMARY

A case report of the Paget-Schroetter syndrome and a review of the literature pertaining to this subject have been presented. This condition represents extravascular obstruction to the axillary or subclavian vein, or both, with or without thrombosis, by adjacent bone and soft-tissue structures. The treatment of this syndrome has been primarily conservative in nature; however, it would appear that early operative intervention with the purpose of removing the extravascular obstruction and re-establishing venous flow by a surgical phlebotomy, when necessary, is the procedure of choice.

SUMMARIO IN INTERLINGUA

Es presentate le reporto de un caso de syndrome de Paget-Schroetter e un revista del litteratura concernite con iste thema. Il se tracta de un condition in que obstruction extravascular del vena axillar o del vena subclavian o de ambes (con o sin thrombosis) es effectuate per adjacente structuras de osso e de histos molle. Usque nunc le tractamento de iste syndrome ha essite primarimente de natura conservative, sed il pare que le manovra de election debereia esser plus tosto un prompte intervention chirurgic serviente le objectivo de eliminar le obstruction extravascular e de restablir le fluxo venose, si necessari, per phlebotomia.

REFERENCES

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While increased serum transaminase levels within 6 hours to 4 days after the onset of the infarct were found most commonly in conjunction with acute myocardial infarction, elevated levels even to the extent of 500 units were found often enough in necrosis and infarction of other organs to suggest caution in interpretation. Curves of the same configuration as that obtained with myocardial infarction were obtained in other situations as was illustrated in patients with multiple infarctions, particularly pulmonary infarctions.

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