

Arterial thoracic outlet syndrome

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Thoracic outlet syndrome (TOS) is a well described upper extremity disorder comprising neurovascular complications caused by thoracic outlet compression. By far neurogenic thoracic outlet syndrome is the most common manifestation of this disorder. Arterial complications of thoracic outlet compression are much rarer, accounting for less than 5% of all operations performed for TOS. Major arterial degenerative and thromboembolic complications can occur as a result of long-standing, intermittent compression of the subclavian artery caused by abnormalities within the thoracic outlet. Early recognition of this abnormality is key to appropriate treatment, before irreparable damage occurs to the extremity.

Etiology

Arterial complications in the thoracic outlet occur from long-standing and intermittent vascular compression. In 1916 Halsted reported on experimentally-induced poststenotic arterial dilatation and degeneration, and included a discussion of 21 clinical cases involving subclavian artery aneurysms associated with cervical ribs [1]. In 1947 Adson described what he termed the “cervical rib syndrome” in an individual with neurovascular compression by this accessory rib [2]. Sporadic reports over the next several decades also report thromboembolic complications caused by a variety of anatomic abnormalities related to the thoracic outlet and compression of the arterial structures. More often than not arterial thoracic outlet compression went unrecognized until

thromboembolic complications ensued [3]. Eden reported on 48 cases in 1939 [4]. Rob and Standeven reported in 1958 on thromboembolic complications caused by TOS [5]. It is now well recognized that the vast majority of these were caused by the presence of bony abnormalities within the thoracic outlet. Chronic, episodic compression of the subclavian artery induces intimal damage leading to stenosis, mural thrombus, and potential thromboembolic complications.

Cervical ribs are present in the vast majority of patients who have arterial complications caused by TOS. Complete cervical ribs are fused to a tubercle on the upper aspect of the first thoracic rib. This fusion point is usually adjacent to the insertion site of the anterior scalene muscle. As a result, the supraclavicular course of the subclavian artery is displaced. Frequently the artery is palpable well above the clavicle and may be tender. Incomplete cervical ribs, although they do not articulate directly with a thoracic rib, usually have an associated fibrous band that can insert on the first thoracic rib, resulting in potential arterial compression. Less frequently an elongated transverse process from C7 or a malunion following a fracture of the clavicle or first rib also can result in arterial TOS. More rarely abnormalities of the first thoracic rib can lead to arterial compression [6]. In addition, isolated congenital bands and on occasion hypertrophic anterior scalene muscles can lead to arterial entrapment [7].

Many if not most patients with cervical ribs are completely asymptomatic and never develop arterial pathology. The incidence of cervical ribs found on routine chest radiograph has been shown to be as high as 0.7% among the general population [3]. Adson reported vascular symptoms

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in only 5.6% of patients he evaluated with documented cervical ribs [2].

Early on, arterial compression is asymptomatic or minimally symptomatic and not easily recognized. Chronic intermittent compression subsequently can lead to pathologic changes within the arterial wall. The earliest lesion is usually simple stenosis of the vessel lumen. At this point the stenosis can spontaneously reverse following thoracic outlet decompression [8]. Over time, however, the artery becomes fibrotic because of chronic inflammation. Turbulent blood flow, poststenotic dilatation, and even aneurysmal degeneration are well described in the literature [9]. The consequences of more advanced arterial wall changes range from relatively minor microembolic events to major vessel thrombosis resulting in potentially limb-threatening ischemia.

Clinical symptoms

Arterial TOS is rarely diagnosed early in its course. Typically some form of thromboembolic complication occurs with resulting symptoms, frequently relatively minor. Small, punctate lesions of the thumb and fingers appear from platelet aggregates that embolize directly downstream [9]. Symptoms can be mild and infrequent and are overlooked by patient and physician. In more advanced cases or when diagnosis has been delayed, major arterial occlusion and potentially limb-threatening ischemia can occur. Many patients who have been diagnosed with major thromboembolic complications caused by thoracic outlet compression give a prior history of a constellation of symptoms similar to Raynaud syndrome, including episodic pallor, erythema, and cyanosis. Chronic pain, coldness, and paresthesias can be present with long-standing disease from multiple microemboli [2]. The presence of unilateral Raynaud symptoms should raise suspicion about the possibility of microemboli from a more proximal source. Many times, even though TOS has been recognized, the arterial manifestations are overlooked because of the belief that nerve compression is leading to the patient's symptoms. As a result the concomitant arterial complications frequently are overlooked. The average age of patients diagnosed with vascular TOS is 10–15 years older than their counterparts with the more common neurologic TOS. Unfortunately the appropriate diagnosis frequently is not made until obvious ischemic changes of the upper extremity occur [10].

The first few episodes of embolization usually are well tolerated and carry favorable outcome even if no surgical intervention is performed. The resulting ischemia of chronic, episodic, embolic episodes soon leads to progression of tissue damage; surgical management then becomes much more difficult, particularly if the emboli are in the distal circulation. In advanced cases the surgical outcome is poor and major amputations can become necessary.

The more proximal the thromboembolic complication, usually the better the ischemic insult is tolerated, because of collateralization around the occluded segments. Even complete subclavian artery occlusion can occur with symptoms many times quite minimal. It has been shown that a proximal arterial source accounts for up to 50% of all upper extremity emboli and thoracic outlet compression is responsible for most [10].

Factors that favor the diagnosis of artery to artery emboli include: unilaterality of the complications, a predominant distribution in the distal circulation of the hands and digits, and the absence of any underlying vascular disorders such as collagen vascular diseases, Takayasu disease, and Buerger disease. Emboli from a cardiac source tend to be larger in size and therefore occlude the large and medium sized vessels of the extremity.

Diagnosis

In the early stages of arterial TOS, symptoms are mild and frequently intermittent. Differentiating arterial TOS from manifestations of neurologic compression sometimes can be difficult. Carpal tunnel syndrome, neurogenic TOS, cervical nerve root compression, and cervical disc can all give confusing symptoms and should be considered. Collagen vascular diseases and autoimmune diseases also can be entertained as possible etiologies, especially if tissue changes have occurred. Usually these disease entities are more diffuse and bilateral, which would not favor the thoracic outlet as the source of the problem.

The diagnosis may be established initially by physical examination with a pulsatile mass in the supraclavicular area as a result of deviation of the subclavian artery. If aneurysmal degeneration has occurred the pulsation may be prominent. Auscultation also may reveal a subclavian artery bruit. Changes in the distal circulation leading to atrophy, skin changes, or even frank distal gangrene may become apparent. The history of

major upper thoracic trauma including fractured clavicle or ribs also may point the clinician to the diagnosis of thoracic outlet compression. Atrophy in the intrinsic muscles of the hand, ulcerations of the tips of the fingers, and decreased blood pressure in the affected extremity can be seen in more advanced cases.

Classic maneuvers for detecting vascular or neurologic manifestations of TOS are discussed frequently but are of questionable significance. The Wright hyperabduction [9] and Adson [2] maneuvers are commonly used clinical tests that can lead to a decrease in arterial pressure or even a loss of the radial pulse, confirming the existence of compression of the vascular system. Although these simple examinations can be helpful, many normal individuals manifest some degree of arterial compression with these maneuvers, making the usefulness of these tests questionable.

Noninvasive testing can consist of pulse volume recordings to include digital waveforms. Depending on the chronicity of the symptoms, these tests may be normal or only mildly depressed. The more advanced the disease and the more chronic the embolic process, the more likely it is for this test to be abnormal. This is also useful in evaluating the overall prognosis when patients reach a more severely ischemic phase of the disease.

Probably the most useful tests that should be obtained initially are routine chest radiographs with oblique cervical spine films to assess bony abnormalities including cervical ribs. The vast majority of patients with arterial TOS demonstrate some abnormality on these films.

An additional noninvasive test that can be useful is Doppler interrogation to include B mode imaging of the vascular system at the subclavian artery. Occlusion, poststenotic dilatation, mural thrombus, and aneurysms can be seen readily during this evaluation. Electrodiagnostic studies such as EMG have limited value in arterial TOS unless there is believed to be concomitant nerve compression.

Transfemoral arteriography of the entire upper extremity vascular system is usually the most reliable test and should be used liberally in anyone suspected of having arterial thoracic outlet compression. Dynamic views may help define the compression point in the early stages of the disorder. Early on, the arteriographic patterns can be subtle; digital subtraction angiography with multiple view planes is recommended.

Patterns of disease on the arteriogram can include:

1. Subclavian artery stenosis at the thoracic outlet
2. Poststenotic dilatation or frank aneurysm
3. Mural thrombus
4. Embolic complications to the axillary, brachial, radial, and ulnar arteries
5. Microembolic complications to the digital, palmar, or interosseous vessels
6. Complete subclavian artery thrombosis

Treatment

Vascular complications associated with TOS usually pose a greater sense of urgency than those caused by purely neurologic compression. The presence of any ischemic changes of the upper extremity requires prompt diagnosis and surgical treatment when needed. Surgical treatment of vascular TOS consists of (1) decompression and repair of the arterial lesion, and (2) management of the associated ischemic problems of the extremity if necessary. Ideally the best approach would allow simultaneous treatment.

Surgical approach

The posterior approach to the thoracic outlet, well described by Pat Clagett in 1962 [11], and the transaxillary approach advocated by Roos and Owens [12] have been described for dealing with neurogenic TOS. The inability to expose adequately the subclavian artery and its compression usually makes these approaches less than optimal, especially when concomitant arterial reconstruction is necessary.

The supraclavicular approach usually offers complete exposure of the subclavian artery, cervical ribs, and muscular or bony abnormalities associated with arterial thoracic compression. First described in 1910 by Murphy [13], it is now the favored approach by most vascular surgeons. In addition, exposure and removal of the normal first rib, when desired, is often straightforward and safe with this exposure also. If the arterial lesion produced degenerative or aneurysmal deterioration of the subclavian artery, additional exposure can be obtained in the infraclavicular or deltopectoral region [14].

First and foremost in the treatment of vascular TOS is thoracic outlet decompression. It is the mainstay of therapy. Complete resection of the cervical ribs or the bony abnormalities is necessary, together with any fibrous bands. Frequently, further decompression by resection of the first rib

and complete anterior scalenectomy is also required. If prior trauma has caused malunion or hypertrophy of the clavicle, then partial or total clavicular resection can be included. Claviclectomy has been shown to offer excellent exposure to the supra- and infraclavicular areas but is not routinely necessary [15].

Once the arterial system has been decompressed, arterial reconstruction can be entertained. On occasion, only mild stenosis with mild poststenotic dilatation is present. Treatment is somewhat controversial, with some investigators recommending no formal arterial repair in the absence of intimal disease or mural thrombus. Spontaneous regression of the stenosis has been observed [16]. Others favor a more aggressive treatment with at least local exploration of the vessel, noting that intimal disease is hard to exclude based solely on preoperative tests [7]. In support of this, Kieffer reported late arterial complications when observation alone is used [17].

Once decompression is achieved and adequate exposure of the subclavian artery is performed, there is usually excessive arterial length and frequently this stenotic region of subclavian artery can be resected and a primary anastomosis performed. In cases in which arterial degeneration, dilatation, or aneurysm has formed, a short segment of graft interposed in the subclavian artery region is necessary. Simultaneous infraclavicular exposure facilitates a longer segment of reconstruction. Material available for reconstruction includes autogenous saphenous vein, arterial autograft, or synthetic prosthesis.

If mild to moderate stenosis of the subclavian artery is present after decompression and no substantial mural thrombus or degeneration has occurred, some investigators favor the use of endovascular stent as the primary repair vehicle [18]. With the advent of covered stents, this application may be expanded in the years to come. Infrequently, isolated intimal fibrous plaques without associated significant arterial dilatation can be treated by arteriotomy and patch angioplasty.

Management of distal embolic disease

Distal embolic occlusions when present are some of the most difficult problems in the definitive surgical management of these patients. They are frequently multiple and diffuse. Chronic emboli are usually firmly adherent to the arterial

wall and not simply extractable with embolectomy catheters. If the distal embolic occlusions involve large vessels and are recent, a thromboembolectomy can be attempted with a balloon embolectomy catheter or similar device. When emboli are chronic, passage of a catheter is often impossible. Intraoperative arteriography can be helpful in these circumstances.

When embolectomy is ineffective or incomplete and the patient is having continuing ischemic symptoms, then formal arterial reconstruction or bypass usually is indicated. Proximal segments may be amenable to prosthetic bypass material; however, autogenous vein is usually preferred. The more distal the embolic occlusions, the harder it is to re-establish adequate perfusion, and major digital and extremity amputations are sometimes unavoidable [19]. Depending on the extent and chronicity of the emboli and presence or absence of collateral flow, symptoms of continuing ischemia and pain can occur. In contrast, when distal embolization is not present or is minimal in nature, the outcome can be satisfactory, provided the initial surgical management is appropriate.

Sympathectomy may be of value as an adjunct in some patients with small areas of distal gangrene of minor trophic changes. Although frequently a transient phenomenon, increased cutaneous perfusion and improved pain threshold can be observed [8].

Arterial TOS, although rare in comparison with the more common neurologic form, can be devastating when the diagnosis is overlooked or delayed. Chronic pain and limb-threatening ischemic changes to the upper extremity can occur. It is generally recommended that thoracic outlet decompression with arterial reconstruction when necessary be performed as soon as any arterial pathology is uncovered. Uniformly excellent outcomes can be expected with timely diagnosis and treatment.

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