THORACIC OUTLET SYNDROME

Pages with reference to book, From 303 To 307

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An unusual case of thoracic outlet syndrome with thyrotoxicosis and pulmonary tuberculosis is reported here.

CASE REPORT

A 55 year old Balochi lady (AM) had been losing weight for two years despite possessing a normal appetite. She was summarily investigated in a small village, pulmonary tuberculosis diagnosed and a course of anti -tuberculous chem. otherapy started. Her compliance was extremely poor and she herself stopped the chemotherapy after a month and continued to lose weight. Soon afterwards the developed palpitations, headaches, excessive perspiration, heat -intolerance and tremors and persisted, untreated, with these symptoms, until the time of her presentation. Two months prior to her admission, the had noticed progressive numbness and coldness of her right hand associated with exertional angina brachii and exercise-related cyanosis of hand. A month later the tip of the right middle finger became deeply discoloured and the developed severe rest pains in the whole hand.

On admission, cyanosis of right hand and lower forearm was seen. Gangrene of the tip of the right middle finger was also present. Proximal axillary pulse was present which disappeared on abduction of the arm to 45 degrees. Distal pulses were absent to Doppler flow study. On the contralateral side (left), all the upper limb major pulses were present to palpation but on abduction of the left arm to 90 degrees, the radial pulse was obliterated easily. Bilateral proximal axiliary artery bruit was present. Atrial fibrifiation and hypertension (left arm=l 60/110 mm Hg) were present with clinical and electrocardiographic evidence of left heart failure. A fleshy, congested, globular exophthalmic goitre was present with bruit in the region of both superior thyroid pedicles. This was associated with asthenia (weight= 40Kg). Both hands were dry but both feet were moist and warm. An angiogram via the femoral route showed minor compression of both sub. clavian arteries by cervical fibrous bands. There was no appreciable dilatation in the post stenotic segments. A rat-tail deformity proximal to the obstructing lesion was seen culminating in total obstructive thrombosis of the right proximal brachial artery.

MANAGEMENT

The patient was heparinised (I/V Heparin 5000 IU six hourly). With the clinical diagnosis of acute on chronic thromboembolism of the right brachial artery, under a local anaesthetic, an immediate successful thromboembolectomy was performed. The yield was a large soft thromboembolus. The upper limb ischaemia immediately improved and Doppler revealed restoration of proximal ulnar pulse. The arteriotomy was closed without a patch with 5/0 prolene. She was digita. lised and propranolol and diuretics were administered. I/V Heparin was changed to oral Warfarin. The hand became warm and pink and her ischaemic pains disappeared and although the
ulnar pulse was restored, the radial remained blocked at this stage.

INVESTIGATIONS

Haemoglobin = 10G%	Indices = Normal
ESR = 60mm 1st hour	RBCs = Normal
Platelets = Normal	Glucose = Smmol/ml

Blood Urea = 25mg% Electrolytes = Normal
T4=l5ng/ml T3 = 2.4Sng/ml
Prothrombin time = 13/13 Urinalysis = Normal
Sputum No AFBs, sterile TB culture = Negative
Chest Xray = Patchy apical Pulmonary tuberculosis
Thoracic outlet = Bilateral incomplete Cervical ribs

MEDICAL TREATMENT

1. Carbimazole 20mg/24 hours
2. Propranolol l20mg/24 hours
3. Warfarin l0-lSmg/24 hours (Prothrombin ratio2.5)
4. Digoxin 0.2Smg/24 hours
5. Rifampicin 450mg/24 hours
6. Isoniazid 300mg/24 hours.

SURGICAL TREATMENT

Five weeks later when she had become euthyroid and the general condition had improved Warfarin was stopped. A week later, radical surgery was performed. A synchronous excision of bilateral cervical fibrous bands, with anterior scalenotomies was performed through supraclavicular approaches. The access was then coverted to a low collar incision by division of the central skin bridge.

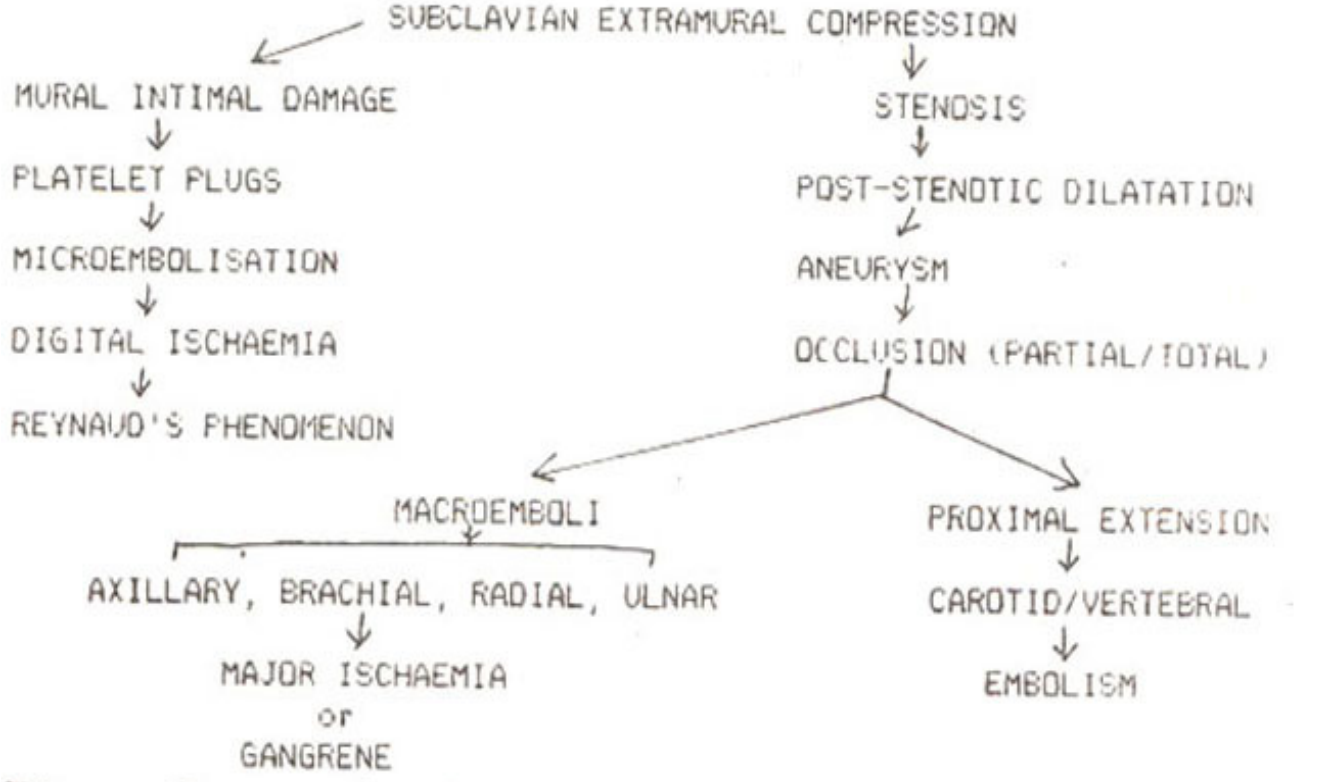

Figure. Progressive features of Thoracic Outlet Syndrome.

A standard 7/8 partial thyroidectomy was performed. Her post-operative recovery was good and fast. She was discharged on the seventh post-operative day on her antituberculous regime. Three weeks later, she was clinically and biochemically euthyroid, both wrist pulses were present in the right upper limb and the dry gangrene of the terminal phalanx had become autoamputated.

DISCUSSION

Thoracic Outlet Syndrome (TOS) refers to a symptom complex of upper extremity. This term was suggested by Rob and Stavenden[1] in 1958 and replaces the terminologies known as the Costoclavicular Syndrome, the Scalene Syndrome, Hyperabduction Syndrome, Paget-von-Schroetter Syndrome and Shoulder Girdle Compression Syndrome. Although the condition was described over a century ago, the predominant emphasis was put on to the neurological aspects of the disorder[2] and this stemmed from the relative infrequency of vascular complications, Roos[3], having reported 370 cases consecutively without serious vascular impairment. Gradually as more dramatic vascular complications became recognised, a re-emphasis was given to the composite problem -

In 1853,. Hilton at Guy's Hospital[4], treated a patient with exostosis of the first rib associated with absent radial pulse and gangrene of the ring and the little fingers. He recognised the compression of the subclavian artery by the exostosis but described the gangrene to be a result of pressure on the ulnar nerve. The patient was not surgically treated and suffered an auto-amputation of the fingers. But eventually all the symptoms subsided. He then treated and reported on a similar experience. In 1861, Coote[5,6] surgically removed a cervical rib from a patient who had an absent radial pulse and the pulse returned after resection of the rib and it is at this point that such a vascular complication of the lOS attracted the interests of the surgeons. In 1869, Poland[7], reported on a case with Reynaud's Phenomen\on, absent pulses and an aneurysm of the subclavian artery associated with a cervical rib. This was treated by continuous digital compression on the cardiac side of the artery for 96 hours. In 1909, Holsted then reported on aneurysms suddenly developing in patients in whom he had resected the cervical rib, In 1916[8], he was able to produce post-stenotic dilatation in a dog's aorta and theorised that this was due to a whirlpool effect beyond the stenotic area.

Todd in 1913 suggested that the lesion was secondary to sympathetic stimulation caused by intermittent compression of the first thoracic nerve[9]. This was reinforced by Telford and Stafford in 1937[10]. Thus the real pathogenesis was pushed into the background despite having been suggested by Halsted in 1916[8] and by Symonds in 1927[11] , that the vascular turbulence was aetiologically responsible for the symptoms. The modern concept was proposed initially by Lewis and Pickering in 1934[12] and was based on a purely mechanical theory. In 1936 Lewis[12] published "All the vascular symptoms displayed could not arise however directly out of compression or out of permanent obstruction of the subclavian artery but they could arise out of thrombosis with embolism". This is now the accepted modern vascular concept of TOS.

As a result of mechanical compression, turbulence and haemodynamic vibratory forces produce muscle fatigue and stretch leading to the well known post-stenotic dilatation. Intimal ulceration, platelet thrombosis and local thromboembolism produce local ischaemia, spasm and Reynaud's Phenomenon, while direct microembolisations produce palmar and digital ischaemia and gangrene.

Rarely, a proximal extension of the thrombus may embolise via the right vertebral or the right common carotid arteries[13,14]

Heymann et al[15] reported the incidence of TOS as 0.5% to 1% on incidental findings at routine chest x-rays. In 50% of the cases the TOS was bilateral and only 10% of these became symptomatic. In the symptomatic group, only 20% presented with vascular complications as opposed to 80% presenting with neurological disorders. Working from the statistics of his big series, Heymann surmised that the overall incidence of vascular complications secondary to TOS are only about 1 in 5000 population.

In most series, women are affected more often than men and the age ranges from 9 to 84. Amongst men, sportsmen, painters and decorators, by reason of exaggerated arm movements, are affected more than others.

Clinical manifestations associated with most vascular complications are slow to take place until the .onset of a major event such as an embolic occlusion. This is indeed the commonest presentation although on leading questions, most patients admit to experiencing episodic embolic symptoms. There are many symptoms attributed to TOS. Pain in the cervicoscapular regions, paraesthesia of the fingers and fatigue of the whole upper limb are common features. With progression, arm claudication leads to rest pains and finger tip gangrene. Brachial plexus comspression results in loss of manual dexterity and weakness of intrinsic musculature of the hand. Major venous compression may cause oedema and cyanosis. The most common complication of TOS is an aneurysm formation of the subclavian artery which can cause peripheral embolisation, acute thrombosis, rupture and concomitant compressive venous thrombosis. [16,17]

In a large series, Gruss,[18-20] noticed that in 95% of his patients, Adson's test was positive[21,22] The diagnosis was then confirmed by doppler and acro-oscillography.

A good protocol for diagnosis is to perform a three minute exercise test of the symptomatic hand (rapid opening and closing of the hand while the arm is in the 90 degrees abducted position with the palm facing the examiner). Weakness, prickling, numbness, pain and pallor are features suggestive of TOS. Plain X-rays of the cervical region in four directions will reveal disorders of cervical ribs, large transverse vertebral apophyses, clavicular callus, bone metastasis and lung apex disorders-factors contributing to the TOS. In the causation of the TOS, by far the most common is bilateral cervical rib compression. In incomplete cervical ribs, neurological symptoms and signs far exceed those relating to the vascular complications. The arterial distension and aneurysm generally produces a supraclavicular bruit. In patients with associated cardiac disorder such as fibrillation, the incidence of digital embolisation far exceeds the incidence of larger arterial lesions. Arteriography characteristically shows the post-stenotic dilatation or aneurysm of the third part of the subclavian artery with usually multiple lesions of the distal branches. Nerve conduction tests should be done to assess the neurological component of the TOS and frequently this differentiates the condition from carpal tunnel syndrome and disc lesions. Doppler pressure, pulse study, nerve conductions, observation of ulceration, capillary filling, x-rays,thermography, duplex scanning and doppler flow studies are all simple non-invasive methods of diagnosing and evaluating the TOS. DSA and formal arteriography should be reserved for selected cases and the value of such an invasive investigation is further enhanced with some kind of provocative manoeuvre, such as Adson's test, shoulder bracing etc.

TREATMENT OF THORACIC OUTLET SYNDROME

Cases which are not too severe are first of all treated conservatively, with local application of heat, massage and isometric exercises to strengthen the shoulder girdle muscles.

When severe symptoms exist, appropriate exploration is necessary. Sufficient evidence, exists to show that when a complete cervical rib is present, the risk of major vascular complication is sufficiently high to warrant excision of the cervical rib regardless of whether major symptoms are present or not.

In severe cases excision of the constricting agent must be undertaken and this may involve excision of the cervical rib, excision of cervical fibrous band, exostosis and if indicated, the first rib as well. A thorough removal of all anamolous insertions and attachments between the spine, cervical rib and the first rib must be undertaken. These procedures can be easily performed either through the axillary or through the supraclavicular route.

Haimovici[23] has reported that better results in respect of revascularisation are obtained when such decompressive procedures are combined with cervicothoracic sympathectomy especially when obstructive lesions are present in the digital arteries. Sympathectomy as an isolated procedure is insufficient.

In conditions where subclavian arterial aneurysm is present, an aneurysmectomy or a bypass procedure with ligation of the aneurysm becomes necessary. Short[24] has postulated that the aneurysm develops as a complication of poststenotic dilatation due to repeated trauma by the scissor like effect between the

clavicle and the first rib during respiration or in shoulder movements. If the aneurysm is left in situ, the damaged intima precipitates episodes of thrombosis and embolism.

Uncomplicated minor post-stenotic dilatation of the subclavian artery usually decreases when stenosis is relieved and this has been aptly described by Bertelsen et al[25] in 1968 in a fairly large and well controlled series. In such cases simple decompression suffices.

In extreme cases where the aneurysm becomes thrombosed, mechanical pressure on the subclavian vein precipitates venous occlusion.

Reconstruction of both vessels then becomes mandatory and this can be performed via the transaxillary approach as popularised by Roos[26]. In the series reported by Gruss[18] out of a total of 128 rib resections, 21 were due to acute venous obstruction. Venous occlusion is often not recognised early and this leads to greater morbidity and mortality.

An aggressive approach, although justified, carries with it its own hazards of pneumothorax, haemothorax, winged scapula and Homer's syndrome, etc. The judicious use of dextran and heparin in the post.operative phase has improved management considerably. Recent experience with Dextran 40 has shown a marked increase in the flow in major peripheral vascular repairs. In addition, thtantithrombotic effect of both agents is well known and with judicious use, the incidence of haematoma is negligible. Fibrinolytic enzymes in real experience are not superior to dextran and heparin.

If the results of the treatment are to improve even further then the diagnosis of vascular complications in the TOS must be made early and treatment offered aggressively. Arteriography or DSA should be performed early. Where feasible, direct arterial repair or bypass should be performed to restore flow in the upper limb. When this proves impossible, further embolic episodes should be minimised by appropriate ligatures.

SUMMARY OF OPERATIVE PROCEDURES FOR VASCULAR LESIONS ASSOCIATED WITH THORACIC OUTLET SYNDROME

There are two basic principles involved:-

(A) Decompression of vessels.

(B) Reconstitution of vessels with sympathectomy.

(A) DECOMPRESSION OF VESSELS

1. Excision of cervical rib and exostosis of first thoracic rib.
2. Routine removal of first thoracic rib and all attached structures causing compression of vessels.
3. Scalenotomy.
4. Claviculectomy (partial or complete) may occasionally be necessary.
5. Pectoralis Minor tenotomy, when axilary vessels are compressed in continuity.

(B) VASCULAR REPAIR WITH SYMPATHECTOMY

1. In order to achieve cbmplete revascularisation, endarteriectomy or a bypass graft of the subclavian artery may be necessary.
2. Thromboendarteriectomy of proximal brachial artery may provide adequate flow through profunda brachii to the forearm and should be considered, followed by a vein patch.
3. Cervicothoracic sympathectomy is now rarely used in isolation. Concomitantly performed with proximal arterial repair it may accelerate collateral development to the forearm and to the hand. Most surgeons now favour sympathectomy as a concomitant necessary procedure[23].

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