REVIEW ARTICLE

Vascular Manifestation of Thoracic Outlet Syndrome

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SUMMARY

Vascular involvement in thoracic outlet syndrome is, apart from neurological symptoms, one of the possible manifestations of this disease, which typically affects young, otherwise healthy individuals. The most frequent vascular manifestations of thoracic outlet syndrome include symptoms related to venous hypertension with possible development of deep vein thrombosis. Arterial involvement is mostly revealed by the presence of ischemic symptoms. Several key points for correct diagnosis should be stressed. They include specific events in patient history, especially previous effort or trauma of the affected upper extremity, typically unilateral. Arterial manifestations are characterized by the absence of ischemic symptoms in other localisations. Deep vein thrombosis occurs in the absence of other distinct precipitating factors and/or hypercoagulable state. Some provocative manoeuvres may help in correct diagnosis assessment, especially in connection with duplex ultrasound examination. Endovascular procedures both in venous and arterial thoracic outlet syndrome have become quite a feature of the therapeutical process, while their role in diagnosis is decreasing. The treatment of thoracic outlet syndrome patients is currently evolving into an interdisciplinary approach integrating angiologists, interventional angiologists and vascular surgeons. This complex multidisciplinary approach improves long-term results and the quality of life of these patients.
Key words: thoracic outlet syndrome, deep vein thrombosis, upper extremity ischemia, endovascular, surgical management. *Va.*

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Thoracic outlet syndrome is a complex of neurological and vascular symptoms which are preconditioned by compression of the vasculo-nervous bundle in the area between the clavicle and the 1st rib. It was described in the 2nd century before Christ by Galen and Vesalio in a patient with symptoms of compression of vasculo-nervous elements by cervical rib (1).

Vascular symptoms of thoracic outlet syndrome are expressed much less often than neurological symptoms. 95 % of cases are manifested by affection of brachial plexus, symptoms affecting the vascular system represent approximately 4 % of cases, and we can detect arterial symptomathology in 1 % of patients. Women tend to be affected most often, and the proportion of affected women and men is approximately 3:1. Annual incidence of upper thoracic outlet syndrome ranges between 3 – 80 new cases for 1000 individuals, and it typically affects younger individuals aged between 20 and 50 years (2).

PATOPHYSIOLOGY

The brachial plexus, vena and subclavian arteries pass on their way from the lower part of the neck and chest to the axilla and then to the arm through three narrowed areas. The first of them is the so-called scalene triangle, which borders m. scalenus anterior in the front, m. scalenus medius at the back and the medial area of the first rib below. The causes of strain in this area may lie in some other structures, for example, various anomalous ligamentous strips, neck rib, elongated processus transverse C7 (partial neck rib) or anomalous muscle bundles. Another place of strain is the costoclavicular triangle bordered in front by the medial collar bone and posteromedially by the upper edge of the shoulder blade. The third place of strain is the subcoracoidal area under the processus coracoideus, fare below the m. pectoralis minor (Fig. 1). Manifestations following from strains of vasculo-nervous structures in these areas may occur in some cases in rest; however, problems often occur or are more considerable during specific manoeuvres or positions of the upper extremity. These are typically elevation of the arm and simultaneous rotation of head to the opposite side (Fig. 2 and Fig. 3). Appearance of pain in the affected upper extremity, particularly in the shoulder and paresthesia, particularly in ulnar part of the forearm and in IV and V finger, are typical for upper thoracic outlet syndrome diagnosis. In this position and with simultaneous palpation of the pulse on the forearm we can notice that it subsides. Another typical manoeuvre which exacerbates or worsens the patient's symptoms (pain, paresthesia, subsiding of pulse), is maximum exterior rotation of the upper extremity and coincident abduction up to 90°. Chronic and recurrent microtraumatization of these structures occurs in situations when the vasculo-nervous bundle goes through the space with insufficient space, particularly with the movement or in certain positions of the upper extremity. The result of this recurrent traumatizing process is, at a vascular level, the creation of solid ligamentous septa inside the affected vein and ligamentous strips with the characteristics of rings in the near proximity of the vein. The vein gradually becomes translucent in consequence of intraluminal and extraluminal processes which may be clinically manifested by symptoms of venous hypertension on the affected upper extremity. Limitation of speed of flow in the vascular system is obviously the predisposing factor for the origin of venous thrombosis, which is very often the first and the

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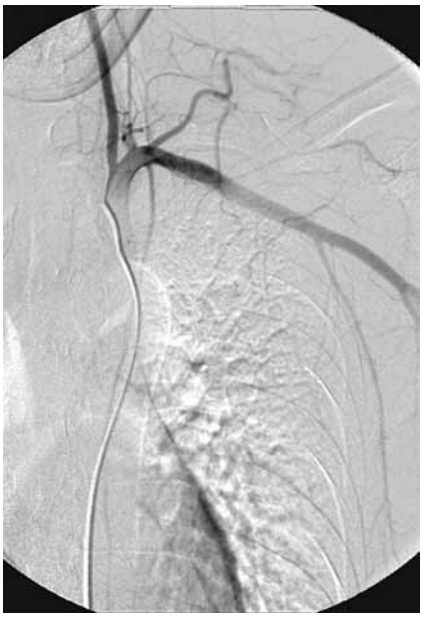

Figure 1. Selective arteriography of left subclavian artery when the upper extremity is situated at an angle of approximately 30° – ordinary medical finding.

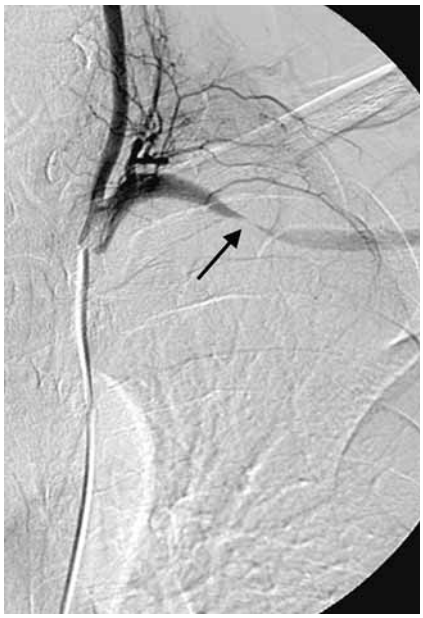

Figure 2. Selective arteriography of left subclavian artery when the upper extremity is raised and the head is turned to the right (tight stenosis of this artery at the level of costoclavicular area (arrow)).

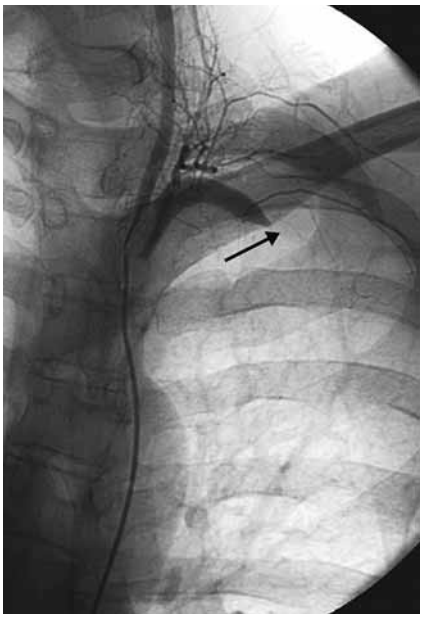

Figure 3. Selective arteriography of left subclavian artery when the upper extremity is raised and the head is turned to the right (tight stenosis of this artery at the level of costoclavicular area (arrow); without using digital subtraction).

most frequent manifestation of venous affection at upper thoracic outlet syndrome.

UPPER THORACIC OUTLET SYNDROME

Thrombosis brought about by this cause was described as early as 1875 by Paget (3), though he considered the painful swelling of the upper extremity to be a consequence of vascular spasm. Von Schroetter in 1884 determined subclavian vein and axilla as the cause of the trombosis (4). Then, in 1949, Hughes used the term Paget-von Schroetter syndrome for primary thrombosis of subclavian vein and axilla (5), which has been used for this disease ever since. It typically affects young, otherwise healthy individuals, sportsmen or manual workers. In the anamnesis of patients there is generally unusual physical effort, carrying heavy strain, etc, several days before the appearance of typical symptoms of venous thrombosis of upper extremity. Men are affected more frequently in the rate 2:1, and in 60 – 80 % it is an affliction of the dominant extremity (2). It appears as swelling, localized first in the palm and forearm, and which gradually spreads to the arm and shoulders. Further, a peripheral cyanosis appears on the fingers, together with a pain which is often characterized by the patient as a pressure and tonus in the affected extremity. It should be remembered that elevation of the extremity often leads to increased tension of the vasculo-nervous bundle, and therefore pain is not eased, as might be expected.

When the vena subclavian and axilla is blocked by a blood clot a subcutaneous collateral venous drawing appears in typical localizations – the lower part of neck and front side of chest. It tends to be relatively noticeable and clearly visible, because it forms gradually from the phase of creation of ligamentous septa and rings, which gradually decreases the translucence of the vein. Venous thrombosis affecting upper extremity is in 7 – 20 % of cases in the acute phase, complicated by pulmonary embolism, and in 25 – 40 % a post-thrombotic syndrome appears after several weeks or months (6, 7). The diagnosis is based on the clinical picture, in which a painful swelling and cyanosis of the affected extremity is dominant. Sudden emergence of pain preceded by noticeable or unusual effort or trauma, which leads to hyper-abduction of the extremity, is typical. In case of rich collateral circulation the symptoms may be weaker; however the subcutaneous collateral venous drawing tends to be more evident in the lower part of the neck and on the front breast wall in the area of pectoralis. Only D-dimer determination is the most significant of all laboratory examinations. Duplex sonography currently plays an important role in venous thrombosis in the upper extremity, and in most cases it definitely determines the final diagnosis. Although classic phlebography of the upper extremity is still considered "the gold standard", it is meaningful to carry it out as a part of intervention approach to the treatment of venous thrombosis of upper extremities (Fig. 4 and Fig. 5).

Treatment of venous thrombosis of upper extremity historically consists of the application of bandage and anticoagulant treatment, first parenterally given heparin, and followed by classic peroral anticoagulant treatment. However, this type of "conservative" treatment involves a high percentage of long-term post-thrombotic complications (8, 9), which may be regarded as unsatisfactory considering the fact that the patients are mostly young and in other respects healthy and active individuals, whose quality of life may be negatively influenced by this disease. Therefore the multidisciplinary concept of treatment of these patients shared by intervention angiologists and venous surgeons is currently accepted. It is based on application of local thrombolytic, catheterization treatment and simultaneous surgical intervention, by means of which the cause of tension syndrome of upper thoracic outlet syndrome is removed. Another catheterization treatment which resolves the occurrence of intraluminal infliction of subclavian vena may in certain cases follow after the surgical intervention.

Local thrombolysis

The most often used thrombolyticum is rt-PA in small doses 0.5–1 mg/hour. In most cases the initial flebography is carried by means of forearm veins, and further it penetrates from this distal

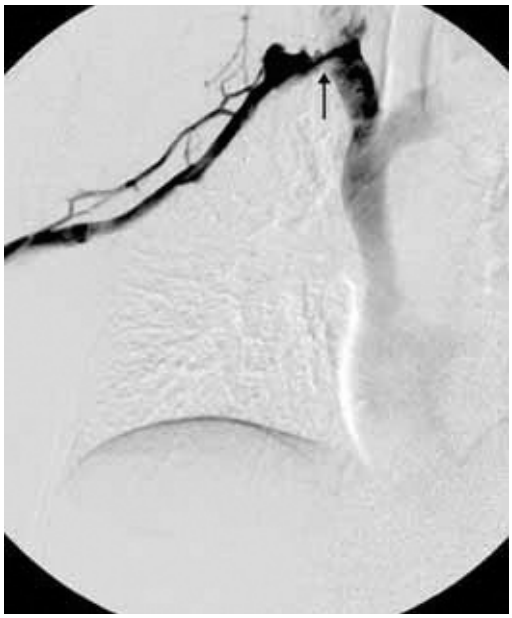

Figure 4. Flebography of right upper extremity when the upper extremity is situated at an angle of approximately 30° (significant stenosis of right subclavian artery in proximal section (arrow); contrast medium penetrates to brachiocefalic trunk through stenosis).

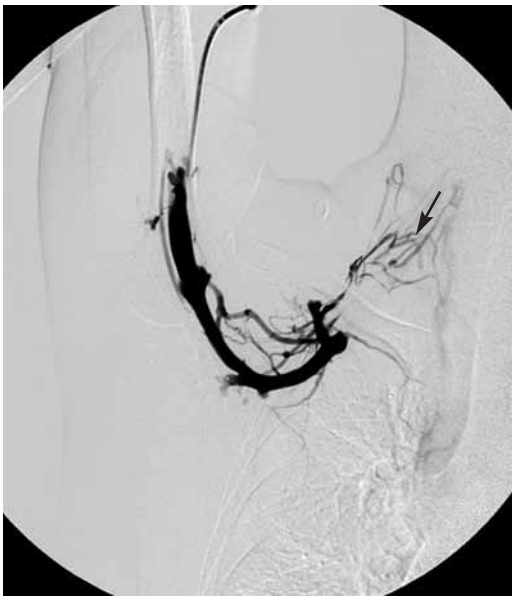

Figure 5. Flebography of right upper extremity when the upper extremity is raised and the head is turned left to the maximum (in the level of the costoclavicular area the right subclavian artery is closed (arrow); contrast medium penetrates through collaterals to cervical arteries).

section of brachial vein up to the area blocked by a blood clot; an infusion catheter is inserted in this place for local application of thrombolyticum in the mentioned dose. Concurrently full anticoagulation treatment is given to the patient, most frequently intravenously infractioned heparin at regular aPTT checks. The process of thrombolysis is clarified at regular flebographic checks, and when the flebographic picture does not change anymore the treatment is completed. If total recanalisation of venous system occurs, provocation manoeuvres on the upper thoracic outlet syndrome are carried out at the final flebography. If this is established, the patient should be indicated for surgical intervention. Patients with recanalisation of axilar vein and part of subclavian vein with continuing impact in proximal section of subclavian vein may be treated first by percutan intervention – balloon angioplastics.

One thing is very important. A metal stent removing the exterior pressure should not be implanted in this area before surgical intervention. The reason is that mechanical damage and other traumatizations of venous walls could be caused. On the contrary, should troubles typical for venous hypertension in upper extremity still continue, there is no other course of action but phlebography. When the affected proximal section of subclavian vein, which corresponds to the place of traumatizations of a vein at the tense syndrome, is revealed, the implantation of endovascular stent for achievement of optimal flebographic result is appropriate. Anticoagulation treatment lasting 6 months follows, provided there are no other reasons for its extension. This multidisciplinary therapy in comparison with "classic" anticoagulation treatment brings the patient two important advantages. Local thrombolytic treatment may totally remove a fresh blood clot in a short period. Thus conditions are given, particularly for restoration of correct function of venous valves, which create the most important patophysiological element for the development of post-thrombotic changes beside the continuing obstruction of venous system. The second factor positively affecting particularly the long-lasting destiny of patients is removal of anatomic causes of tension of upper thoracic outlet syndrome. This way the cause of the venous hypertension and the risk of relapse of venous thrombosis are removed. This combined approach to the patient with Paget-von Schroetter syndrome, which allows the patient a full return to normal life, should be the first choice of treatment, particularly in case of young active patients.

ARTERIAL SYNDROME OF UPPER THORACIC OUTLET

Arterial syndrome is the least common manifestation of upper thoracic outlet syndrome, and it occurs approximately at 1 % of cases (10). Chronic microtraumatization of arterial wall leads to its degenerative changes. In most cases their histological picture corresponds to advanced atherosclerotic changes, which may lead to reduction of translucence or total closure of subclavian artery or – in contrast - an aneurismatic dilatation of artery in the affected place with option for creation of wall thromboses, which may create a source of distal embolisation. Arterial involvement may appear with pains in the area of forearm, arm or shoulder muscles when working with the affected extremity, particularly in case of stretching. Other symptoms may be quick tiredness of the affected extremity during physical work, lower tolerance to cold, changes of colour – pale or peripheral cyanosis in fingers. If an aneurism of subclavian artery with wall thrombosis is caused in consequence of upper thoracic outlet syndrome and repeated microembolisation to digital arteries occurs, the only manifestation of this state may be Raynaud's phenomenon. Unlike venous manifestation, the arterial affliction is most frequently conditioned by damage of arterial wall by chronic traumatization caused by some bone abnormalities, which we find in 88 % of patients (10). In spite of the fact that the subclavian artery may be chronically traumatized in scalene triangle by musculus pectoralis minor or humeral head, the most frequent place where the artery is compressed is the costoclavicular space. Here the entire neck rib, which is jointed with upper area of the first rib laterally from the point of joint of musculus scalenus anterior, is very often influencing. Another possible bone abnormality may be an elongated crosswise projection C7, sometimes also called incomplete neck rib, abnormalities of first rib or col-

lar bone. In most cases it is local bulge, thickening or extrusions, which cause reduction of costoclavicular space. The moment of regularly repeating activities carried out by the afflicted extremity probably plays an important role in the development of arterial upper thoracic outlet syndrome. Some studies document explicitly higher occurrence of arterial infliction on dominant extremity (11 – 13), which may relate simply to the repeating strain of the dominant extremity in common life. Symptoms of arterial upper thoracic outlet syndrome may be ignored for long time or misunderstood. The precondition for correct diagnosis is detailed sampling of anamestic data; when repeating straining of the dominant extremity it is typical for painters, certain instrumentalists (guitarists), athletes (throwers), mechanics, porters, etc. Increased strain very often precedes occurrence of typical symptoms such us problems in the cold, pains particularly when elevating extremity, tiredness, pale peripheral cyanosis and Raynaud's phenomenon. Unilateral occurrence of problems is also typical. Uninvasive examination plays an important role in diagnosis: measuring of pressure in radial artery and ulnaris on both upper extremities, identification of occurrence of murmurs above the collar bone, rarely pulsing resistance behind the collar bone corresponding to aneurism of subclavian artery may be palpable. Duplex sonographic examination may determine exact changes in the flow in arteries of upper extremities at provocation manoeuvres and is a very good method for objective postoperative monitoring of these patients. Displaying X-ray examination as a simple radiogram of chest focused on bone structure. CT examination with 3D reconstruction may disclose present bone abnormalities. CT angio examination may then bring important information on the state of arterial bloodstream in this area, on presence of aneurysm or stenosing infliction of subclavian artery and on its relation to bone abnormalities. Classic arteriography displaying the aorta arc and selectively arteries of upper extremities already steps aside in the time of arterial upper thoracic outlet syndrome diagnostics. This is suggested if the operating surgeon requires a detailed display of effusion at the planned revascularization of upper extremity or as a part of endovascular treatment – local thrombolysis at the distal embolisation to forearm and arm arteries.

Treatment of arterial upper thoracic outlet syndrome generally comprises three parts: A) decompression of costoclavicular space and removal particularly of bone structures compressing subclavian artery; B) removal of the affected artery, which may be the source of distal embolisation; C) restoration of perfusion in distal areas of upper extremity. In some cases it is sufficient only to remove the causes of compression, in other cases surgical intervention in subclavian artery is required, which may be resection of aneurismatically affected part of this artery and its replacement by graft. In case of embolisation of closing of distal parts of arterial system in upper extremity it is possible to carry out surgical thromboembolectomy or catheterization treatment (percutan aspiration of thromboembolic matters or local thrombolysis). We can treat by means of catheterization chronic lesions in subclavian artery caused by traumatization in costoclavicular space after surgical removal of outer compression: rigid stenosis requiring treatment by implantation of rigid metal stents to achieve a good result. If the revascularization performance is not feasible for infliction of small arteries in forearm or palm, an infusion treatment of prostaglandin or sympatectomy are indicated. The outlook for the extremity at the ischemia occurred by other causes than infliction of the binding and vasculitide is good; only 22 % of them grow during fifteen years of monitoring to the stage of ulcerations and only in 6 % of them is amputation of the affected extremity necessary (14).

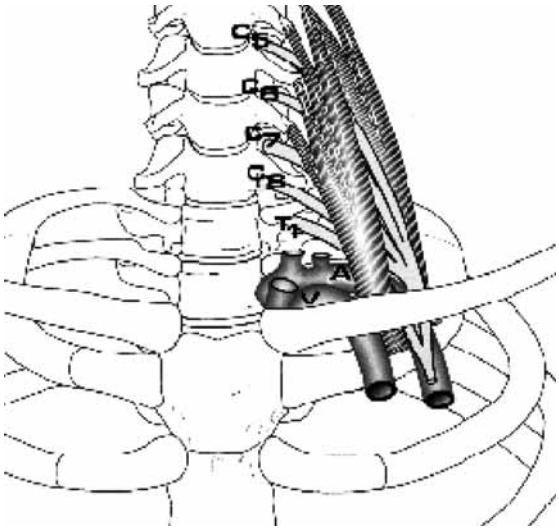

Figure 6. Anatomy of upper thoracic outlet

CONCLUSION

Arterial infliction at the upper thoracic outlet syndrome appears in approximately 5 % of patients with this diagnosis and is typical for young, otherwise healthy and active individuals. Venous thoracic outlet syndrome manifests most often with arterial thrombosis, arterial upper thoracic aperture manifests with ischemic symptoms. Diagnosis of vascular infliction does not generally cause problems; however, it is necessary to focus on the upper thoracic aperture area to exclude compression of vasculonervous bundle in this area in younger patients with ischemic symptoms of upper extremity or in patients with venous thrombosis without evident cause. Correct diagnosis leads to better long-term results for these patients and to improvement of the quality of life, which is very important for otherwise healthy individuals.

Abbreviations

aPTT – Activated Partial Tromboplastin Time
rt-PA – recombinant tissue-type plasminogen activator

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REVIEW ARTICLE

Superior Vena Cava Syndrome – Possibilities of Intervention Therapy

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SUMMARY

Superior vena cava syndrome is a relatively frequent complication in patients with lung or mediastinal malignant disease. Standard treatment is usually based on radiotherapy or chemotherapy. During the last 20 years endovascular methods such as stent implantation and local thrombolysis have been increasingly employed, being more effective in the treatment of superior vena cava obstruction or stenosis.
Key words: superior vena cava syndrome, stent, thrombolysis. *Be*

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Superior vena cava syndrome is a complex of symptoms caused by deterioration of blood flow through the superior vena cava (SVC) to the right heart atrium, resulting in congestion of blood in the upper part of the body. The syndrome was first described by William Hunter in a patient with an aortic aneurysm in 1757 (1).

ANATOMY OF THE SUPERIOR VENA CAVA

SVC is a thin – walled vessel about 6-8 cm long, situated in the upper mediastinum. It is formed by the joining of two brachiocephalic veins and flows to the right atrium. It is in close contact with the trachea, right bronchus, aorta, pulmonary artery and thymus. It is also surrounded by lymph nodes. Dorsally, the azygous vein flows into the SVC and may represent an important collateral circulation if the SVC is obstructed (2, 3).

ETIOLOGY

The causes of superior vena cava obstruction have changed during the years. While until the first half of the last century the syndrome was mainly caused by the pressure of tuberculous or syphilitic aortic aneurysms (4), nowadays more than 80 % of SVC syndromes are triggered by an advanced malignant disease, most frequently by bronchogenic carcinoma and particularly its small–cell type, by non-Hodgkin lymphoma (NHL) and by metastatic mediastinal tumours (5, 6). Thyroid carcinomas and thymomes are more rare causes. SVC syndrome may be caused either by an external SVC compression, or possibly by a stenosis-related thrombosis or a direct infiltration of a tumour into the vessel. Approximately 5-15 % of patients with bronchogenic carcinoma and 3-8 % of patients with NHL develop SVC syndrome (7). Benign etiology is a more rare cause of SVC syndrome, occurring in approximately 15-20 % of all cases (6, 7), but considering the increasing frequency of central venous catheters and stimulating electrodes implementing, SVC thrombosis related to venous catheters and cardiostimulating electrodes is not infrequent. On the other hand, mediastinal fibrosis is a less common cause, resulting most often from previous lung and mediastinal radiotherapy (8).

CLINICAL PICTURE

SVC syndrome symptoms follow from the venous congestion in the drained areas. The seriousness of the syndrome depends on its onset rapidity and on the duration of the SVC obstruction, as well as on the related possibility of the dilatation of collateral circulation. Moreover, it depends on the location of obstruction. If there is a stenosis or obstruction above the azygous vein ostium, the collateral flow is ensured via this vein contrary to the obstruction below the azygous vein ostium (9).

Venous hypertension can produce headaches, a feeling of pressure in the neck and head, dizziness, syncopes and coughing. There may be other apparent symptoms of SVC syndrome, such as oedemas of the face, neck and arms, a noticeable dilatation of neck and arms veins, lips and face cyanosis or even a coma in the final stage (10, 11).

DIAGNOSIS

If the syndrome is fully expressed, the diagnosis is already evident from the medical history and physical examination. The diagnosis can, however, be specified by a quantity of non-invasive and invasive examination methods.

An X-ray of the heart and lungs may reveal a widened mediastinum, pleural exudate and mediastinal or hilar tumour, particularly on the right side (6). Sometimes dilatation of the azygous vein can be apparent (12).

A Duplex ultrasonic examination is a non-invasive method that cannot picture the superior vena cava directly, but it well represents the subclavial and possibly brachiocephalic veins. On the basis of indirect signs, such as breathing variability of blood flow or dilata-

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tion during the Valsalva manoeuvre, a duplex ultrasonic examination can reveal suspicion of central obstruction.

Computer tomography (CT) is a non-invasive examination making it possible to display in detail anatomical structures, the cause and extent of obstruction and of the collateral circulation.

Nuclear magnetic resonance (NMR) provides a higher-quality image of anatomical structures than CT; however, in our context NMR availability is still limited, and it is contraindicated in patients with an implanted pacemaker.

Contrast phlebography remains the gold standard in diagnostics. It makes it possible to display the level and extent of the obstruction and of the collateral circulation, as well as potential presence of thrombi. In specialised departments an endovascular intervention can be carried out simultaneously. The phlebographic findings are the cornerstone of the Standford-Doty classification of the obstruction types: I. type: < 90 % SVC stenosis and a patent azygous vein, II. type: 90-100 % stenosis of SVC and a patent azygous vein, III. type: 90-100 % stenosis of SVC with a reverse circulation in a patent azygous vein and IV. type: occlusion of SVC and its supplying veins (3). The main disadvantage of a phlebographic examination is its invasive character and the necessity use of a contrast agent.

There are other alternatives of examination of the primary disease aetiology, such as bronchoscopy, pleural exudate puncture and mediastinoscopy.

TREATMENT

Treatment of the superior vena cava syndrome depends on the cause of obstruction, gravity of symptoms, the patient's prognosis and wishes.

Pharmacotherapy

In addition to the elevation of the upper part of the body, we use diuretics and corticosteroids to induce regression of the swelling and anticoagulant therapy to prevent thrombosis progression. This treatment, however, has very limited clinical effect (10, 13).

Radiotherapy, chemotherapy

If the aetiogogy of the SVC syndrome is a malignancy, radiotherapy, chemotherapy or a combination of the two - depending on the histological type of the tumour - form the basis of treatment. Most studies are aimed at bronchogenic carcinoma, which is the main cause of SVC stenosis or occlusion. Radiotherapy in radiosensitive and chemotherapy in chemosensitive tumours represents a standard treatment and brings symptomatic relief due to the reduction in tumour tissue volume. The effect of radiotherapy ranges from 46 to 90 % within 2 weeks (14-16) while the effect of chemotherapy oscillates between 62 to 80 % in the treatment of small-cell bronchogenic carcinoma (17, 18). Furthermore, a combination of both treatment methods can be used (17). Analysis from the Cochrane Clinical Trials Register including 2 randomised and 44 non-randomised studies of the treatment of SVC obstruction in bronchogenic lung carcinoma (and more particularly in its small-cell type) notes the effect of chemotherapy and/or radiotherapy on the regression of difficulties caused by the SVC syndrome in 77 % with recurrence in 17 %. In non-small-cell varieties of bronchogenic carcinoma chemotherapy and/or radiotherapy led to the regression of symptoms only in 60 %, with recurrence in 19 %. These traditional methods lead to a clinical effect with a delay of 2-4 weeks (17, 19, 20).

Surgical treatment

Until recently a bypass operation was the only alternative treatment in the event of failure of conservative therapy with a very good and long patency, and it still remains an alternative if there is failure of endovascular treatment or if a radical resection of a tumour can be effected. Owing to a very good and long patency – 88 % of patient bypasses with an average observation period of almost 11 years (20) – some authors prefer this method for cases of benign causes of the SVC syndrome (21-24). In patients suffering from a malignant disease that are in a general bad state, the necessity of sternotomy is the main inconvenience.

Stent implantation, local thrombolysis

The history of metal stents use in the superior vena cava started in 1986, when Charnsangajev carried out successful angioplasty of SVC for the first time, with a stent implantation in 7 dogs with the mediastinal fibrosis (25). Since then the method has become an appropriate alternative to a standard conservative treatment. Its technical success with a subsequent rapid clinical effect has ranged from 90 to 100 % in most of the published studies. In the above-mentioned meta-analysis from the Cochrane Clinical Trials Register, stent implantation led to a regression of symptoms in 95 %, with a recurrence of SVC syndrome in 11 %, but another recanalization was possible in most cases; therefore the long-term patency was 92 % (17). In the event of an extensive SVC thrombosis stent implantation is accompanied by local thrombolysis. In his study, Kee effected local thrombolysis in 27 patients with an acute SVC thrombosis of malignant etiology that in itself led to recanalization in 4 patients (15 %), while the remained patients underwent a stent implantation. During the local thrombolysis one patient with a small-cell carcinoma of lung died of pulmonary embolism of thrombotic and tuberculous masses (26). Other papers show a similar or better technical and clinical effect (27-31). The rapidity of endovascular therapy is also advantageous – the clinical effect comes in a short interval of 2-4 days (26-31). Local thrombolysis effected before proper stent implantation decreases the volume of thrombotic material that could embolise during the procedure. The dissolution of the thrombotic mass also reveals the cause of SVC obstruction, thus reducing the number and length of stents necessary for the recanalization of the obstruction - which, incidentally, brings a considerable economic benefit (26). The evident disadvantage of this therapy is an increased risk of bleeding complications in patients with an increased risk of potentially fatal bleeding due to their principal tumour disease. According to the published studies, fatal bleeding complications during the local thrombolysis occur in 0-5 %; only one small study with 10 patients noted a complication rate of 10 % (1 patient died) (26-31).

OWN EXPERIENCES

During the period of October 2002 to December 2004 three patients with acute superior vena cava syndrome of malignant etiology were admitted to the ward of angiologic intensive care of the 2nd internal clinic of cardiology and angiology, 1st Medical Faculty of Charles University in Prague. Two patients had already undergone unsuccessful treatment in another hospital – 1x systemic thrombolysis and 1x percutaneuous aspiration thrombectomy were performed without any considerable effect. The aetiology was as follows: a relapse of malignant thymoma, Ewing's sarcoma and stomach adenocarcinoma. SVC thrombosis in the patient with Ewing's sarcoma was associated with the central venous catheter.

Intervention performance

After sonographic or CT verification of the SVC obstruction, we bilaterally spiked the brachialis vein and inserted the 6F case by Seldinger method, in local anaesthesia under sonographic or X-ray control. Via the cases we performed a phlebography to reveal the character and the extent of the disease. The thrombosis and the com-

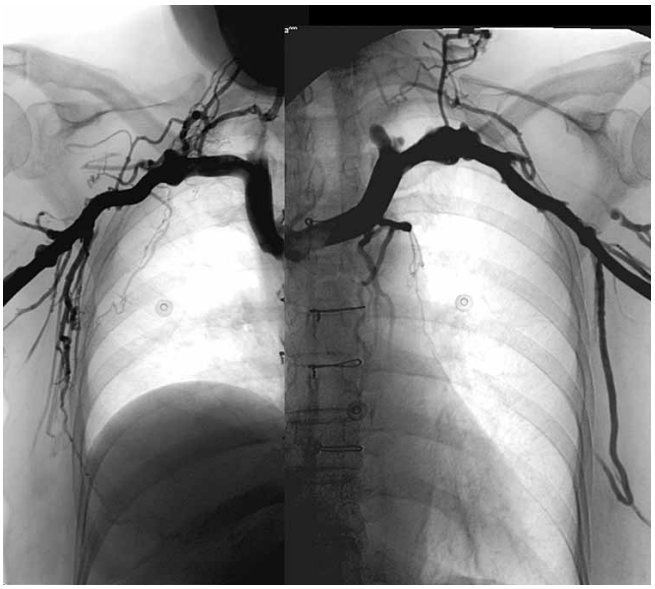

Figure 1. The first patient: phlebography from both brachial veins, a complete occlusion of SVC, there is a thrombus in a junction of brachiocephalic veins

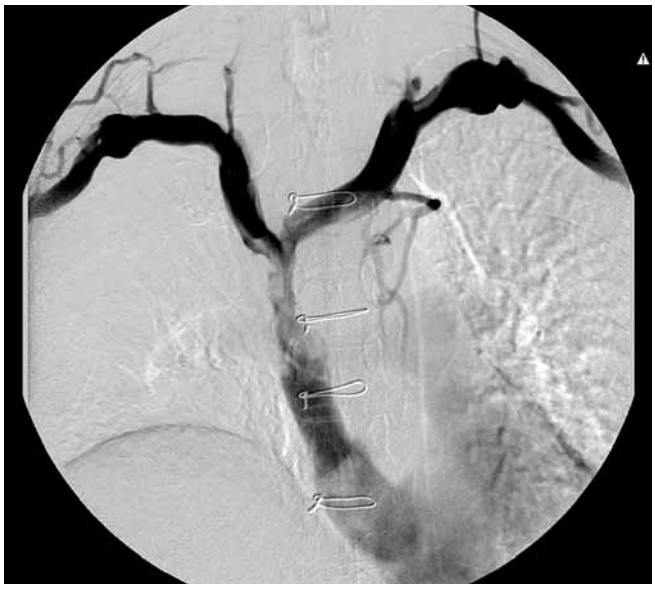

Figure 2. The first patient after 22 hours of thrombolysis (partial SVC recanalization with residual thrombi)

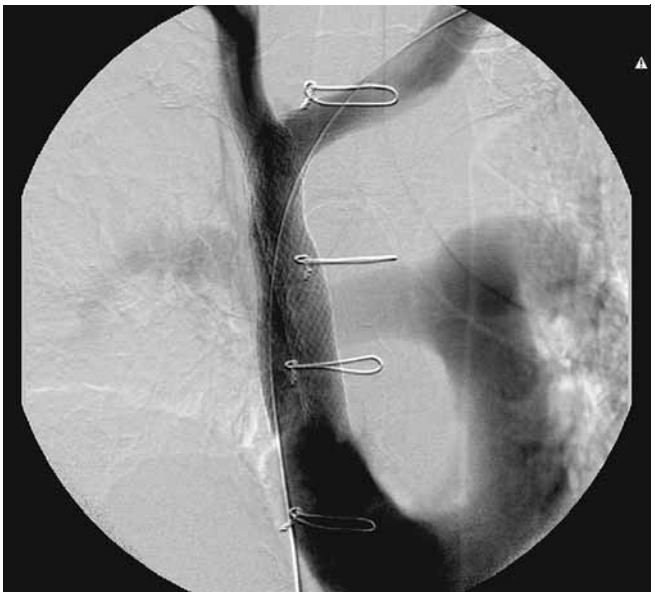

Figure 3. The first patient after 44 hours of local thrombolysis and implantation of 2 self-expanding stents

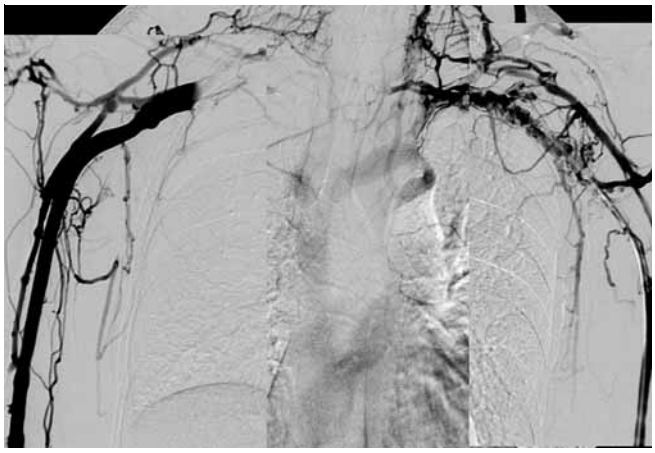

Figure 4. The second patient: phlebography from both brachial veins, thrombosis of axillar vein on the left, both subclavial veins and both brachiocephalic veins are occluded, the superior vein cava fills poorly

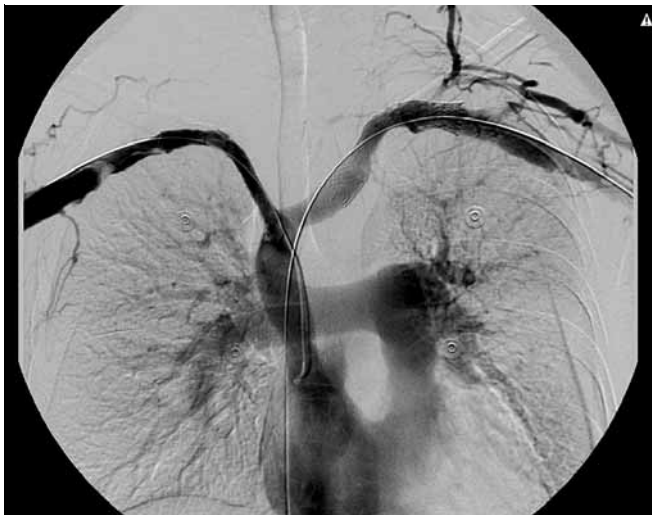

Figure 5. The second patient after 60 hours of local thrombolysis and bilateral implantation of 2 self-expanding stents on boundary of subclavial and brachiocephalic veins (residual thrombosis in the subclavial vein on the right, contrast agent however flows away very well from the periphery)

plete SVC obstruction were present in all three cases. The first patient had patent brachiocephalic veins (Fig. 1, 2, 3). The second patient had a thrombosis of both SVC and left brachiocephalic vein (Fig. 4, 5) and the third patient had a thrombosis of SVC, both brachiocephalic and subclavia veins and a unilateral thrombosis of the left axillaris vein. We administered Heparin 5000 U IV and via the cases, by means of soft hydrophilic conductors, we inserted local thrombolysis catheters with lateral orifices into the coagulum. We commonly use the recombinant tissue-plasminogen activator (rt-PA, Actilyse®, alteplasum) in the total dose of 1 mg/h into the catheters, and we applied heparin in an initial dose of 1250 U/h into the inserted case, with the dose adjustment according to APTT. The target value should be between 2-3 multiples of normal values. We performed a control phlebographic examination at intervals of 12 to 24 hours to consider the treatment effect and reposition of inserted catheters. The total time of local thrombolysis was on average 57.6 hours (38-60 hours); in one case the local thrombolysis alone led to

a complete opening of the venous vascular system, while in the two remaining cases an intervention performance followed – the implantation of stents into the SVC or into the brachiocephalic trunks and the subclavian vein. A quick regression of the swelling of the upper part of body and the regression of subjective symptoms came in all cases. In all patients we continued anticoagulant treatment by low-molecular heparin and a subsequent warfarinization after the performance. No complications appeared in our patients during the local thrombolysis and during their stay in our ward (on average 6.6 days). Subsequently 2 patients died during the following observation – one died of intracranial bleeding caused by brain metastases (using the therapy of low-molecular heparin) 2 weeks after the operation. The second patient died of the progression of his malignant disease after 3 months. The third patient has been without clinical signs of relapse of SVC syndrome, and the intervented veins have still been patent according to the executed examination as well.

CONCLUSION

Superior vena cava syndrome is a relatively frequent complication in patients with lung or mediastinal malignancy. The standard treatment on the basis of radiotherapy or chemotherapy is successful in a wide range of 45-80 %; however the clinical effect comes with a delay of 2-4 weeks. Endovascular treatment – stent implantation – is a highly effective method in short lesions, with a technical and clinical successfulness of 90-100 % and a quick effect. Local thrombolysis followed by angioplasty and possibly stent implantation is an appropriate method in an extensive thrombosis of SVC and other veins draining the upper part of body. The risk of fatal bleeding in local thrombolysis is 4-10 % according to the published papers. In contrast to radiotherapy, endovascular treatment allows reintervention in cases of relapse of SVC syndrome. The high cost of the intervention performance is a disadvantage, but it is still a fraction of sum expended for the total treatment cost of patients with the malignant disease.

The surgical treatment is an alternative for the cases of failure of conservative and intervention therapy or for patients with benign causes of the superior vena cava obstruction.

Abbreviations

aPTT – Activated Partial Tromboplastin Time
CT – computer tomography
SVC – superior vena cava
NHL – non-Hodgkin lymphoma
rt-PA – recombinant tissue-type plasminogen activator

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