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Vascular Surgery Subdivision, The Pope John Paul II Hospital, Zamość
1st Department of Radiology, Medical University of Lublin

MAREK DURAKIEWICZ, ELŻBIETA CZEKAJSKA-CHEHAB,
ROBERT GRAJETA, ANDRZEJ DROP

Paget-Schroetter syndrome in a young female patient. A case report

Paget-Schroetter syndrome, i.e. effort thrombosis of the upper extremity (upper extremity deep vein thrombosis – UEDVT) was described by Paget in 1875 and Schroetter in 1884 independently; Huges, describing another case with analogous clinical symptoms, gave it its name. The disease is rare and its diagnosis still difficult. It is estimated that UEDVT accounts for only 5% of all locations of deep vein thrombosis, however in 10–30% of cases it may be the cause of pulmonary embolism (1, 4-6, 10). Its most common location is the axillary-subclavian part of the venous system, which is apparently connected with higher possibility of compression by the structures surrounding the neurovascular bundle (the subclavian vein and artery and brachial plexus). Based in the pathogenesis, UEDVT should be divided into an extremely rare primary disorder corresponding to Paget-Schroetter syndrome (2 per 100.000 persons per year) and much more common secondary form. Secondary UEDTV has been reported in up to one fourth of patients with upper extremity central venous catheters (for chemotherapy, dialysis, parenteral nutrition), in pacemaker bearing patients, in patients suffering from malignant neoplasms or following trauma of this region (1, 6, 10). Idiopathic thrombosis of the axillary and subclavian veins is extremely rare and its most frequent "cause" found in the history is the heavy exertion (3, 4, 6, 10). A wide range of complaints reported by patients should be stressed, mainly the upper limb swelling and persistent pain. Moreover, in progressing cases, the development of upper vein thrombosis with all known consequences and post-thrombotic syndrome is relevant. Furthermore, possible problems concerning vascular access for dialysis should be mentioned. Idiopathic deep vein thrombosis most often involves the dominating limb and despite the diagnosing procedures carried out, no obvious cause of it is found; it is believed that microtrauma to the vascular intima is likely to be the onset of thrombosis (1, 4, 6, 8).

CASE REPORT

A 28-year-old female patient presented with headache and right upper limb numbness, tingling sensation and periodical finger cyanosis. The progressing complaints occurred for 2–3 days, their onset was associated by the patient with physical effort. Her past history was negative, with two uneventful pregnancies and spontaneous labours. The patient was not on any contraceptive medications. Her family history was negative for venous thrombosis and pulmonary embolism. The neurological and ophthalmological examinations did not reveal any abnormalities, no fever, dyspnoea or thoracic pain were found. Doppler ultrasound of the upper limb veins showed limited resilience of the right axillary vein, the vessel was filled with recent thrombus without flow features. The brachial veins were patent and elastic, the flow maintained although greatly impaired. The physical examination revealed oedema of the forearm and hand, marked cyanosis, dysaesthesia with the features of Raynoud's phenomenon. The pulse was palpable on both right carpal arteries. During hospitalization in the Department of Vascular Surgery the patient underwent thrombolysis. The megadose streptokinase method was used and totally 7.5 mln. units were administered in continuous infusion (infusion pump) to the superficial vein of the right forearm (1.5 mln units/h) for 5 h supplemented with unfractionated heparin in the dose of 1000 units/h. A temporary increase in temperature and short-term episode of chills (which subsided after

the administration of 300 mg of i.v. Hydrocortisone.) were observed in the initial period of streptokinase infusion. After thrombolytic therapy the local complaints completely subsided. The heparin infusion was continued for 96 h and simultaneously oral anticoagulants were administered. The follow-up Doppler ultrasound did not show any symptoms of thrombosis within the brachial, axillary and right subclavian veins. On the 5th day the patient was discharged in good general and local good condition with INR of 2.2. The laboratory tests for congenital thrombophilia (AT III, C and S proteins, APC-resistance) did not confirm its presence.

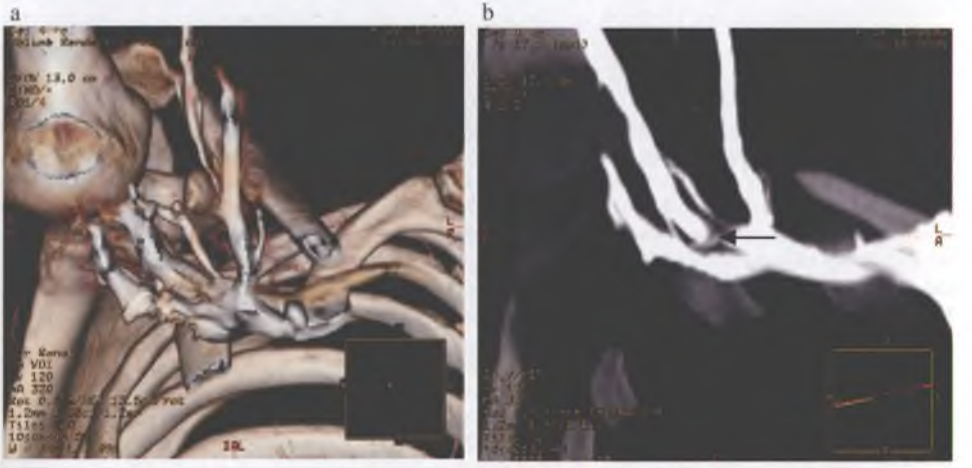

Fig. 1. The 3D view (a) and MIP oblique view (b) of the first phase of the MSCT scanning (postural maneuver, delay time 13 s) show cephalic, basilic, axillary and subclavian veins

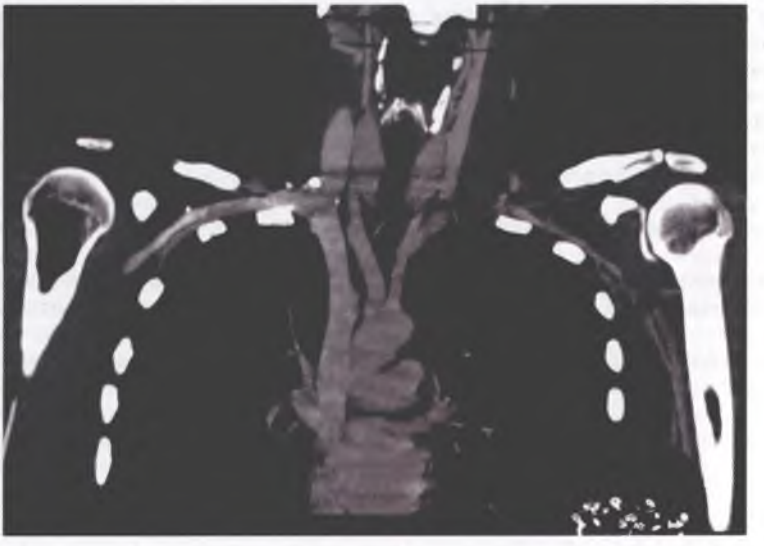

Fig. 2. Coronal oblique reconstruction shows normal thoracic vessels in the phlebographic phase

In order to perform a precise follow-up examination of the local state and find a possible cause of thrombosis, the patient was referred for multislice CT angiography of the thoracic cavity and upper thoracic foramen region. The CT was conducted using the 8-row tomograph LightSpeed

Ultra (General Electric Medical Systems– Milwaukee) with secondary reconstructions on the Workstation Advantage Window 4.2. Following pre-contrast scanning in 5-mm slices, the two-phase 1.2-mm scanning was performed after intravenous bolus of the contract medium (Ultravist 300–100 ml, administered by the automatic power injector to the right basilic vein at the speed of 4 ml/s). The scanning delay time was determined with the bolus tracking technique. Phase I – early arterial (delay time 13 s) – was conducted in 1.2-mm slices with the right upper limb fully raised (Fig. 1 a, b). Phase II – phlebographic (delay time 150 s) included the region of the upper thoracic outlet and was conducted with the upper limbs lowered (Fig. 2). The scans were evaluated using multiformat reconstructions (coronal, sagittal and oblique and 3D volume rendering). No changes within the mediastinal structures were detected, particularly clots in central thoracic veins or symptoms of acute and chronic pulmonary embolism. Bone or muscular developmental variants, which may be the cause of the thoracic outlet syndrome or posttraumatic changes, were excluded. The subclavian arteries were normal. The subclavian and axillary veins were bilaterally patent. The right doubled brachial vein, brachial basilic vein and cephalic vein – with insignificantly widened lumen. In the first scanning phase (with raised right extremity) one of the veins before the junction with the axillary vein and cephalic confluence was bent and constricted by more than 50% at the distance of 10 mm, in the second phase of scanning the lumen was no longer stenosed. No symptoms of developed collateral circulation were found. The patient is further followed up and does not show any post-thrombotic symptoms.

DISCUSSION

Upper extremity deep vein thrombosis is an increasingly important clinical problem with potential for many complications and morbidity. Prandoni et al. (4–6), evaluating the location of UEDVT, detected simultaneous changes in the axillary and subclavian veins in 54.7% of patients, only in the subclavian vein in 26.4%, only in the axillary vein – in 11.3% and isolated changes in the brachial veins in 7.5% in the group of 53 patients examined. The most common risk factors detected included: thrombophilia (22.6%), active neoplastic process (20.7%) and excessive physical exertion (20.7%). In about 15% of patients no cause of the disease was confirmed (1, 4, 5). The data about high risk of pulmonary embolism and significantly higher incidence of postthrombotic syndrome compared to lower limb thrombosis are extremely alarming. The embolic complications are likely to account for more than 36% of cases and are often the first symptom of the disease (4, 6).

At present, the lesions may be detected, their intensity assessed and the post-treatment period followed up using different imaging modalities. Duplex ultrasonography is considered the initial imaging test of choice in the diagnostics of UEDTV. This method is easily accessible, noninvasive, safe, inexpensive and reproducible. The accuracy of Doppler ultrasound for jugular, distal subclavian and axillary UEDTV is high (1), however its limitations concern the assessment of the proximal part of the subclavian vein below the clavicle, no possibilities of visualizing further segments of the thoracic venous system and of detecting embolic complications. Contrast venography provides excellent visualization of the upper extremity and chest veins, yet technical difficulties in cannulating the vein in an edematous arm may occur; moreover, in many centres the examination is not available for 24 h. It is currently believed that contrast venography should be used in cases of negative ultrasound results with high clinical suspicion of UEDTV and before planned interventional procedures. Some centres still use venography for target thrombolytic therapy and follow-up assessment of vascular compression after effective thrombolysis. In the recent years, two quickly developing methods have been increasingly used to diagnose systemic veins – CT angiography, multislice, in particular, and MR angiography. Magnetic resonance angiography (MRA) and multislice computed tomography angiography (MCTA) are extremely accurate methods which additionally enable us to detect thrombi in the central thoracic veins and right heart and visualize normal and accessory flow pathways of the thoracic system veins (1, 2). Furthermore, both examinations have a great value in the assessment of the cause of vessel compression. The main advantage of MRA is the fact that radiation and iodinated contrast agents are not necessary while an important drawback is its relatively long duration, poor availability and

high costs. The method cannot also be used in pacemaker-bearing patients and persons with some types of catheters. Despite its limitations, similar to those of venography, radiation risks and iodine allergy, which should be kept in mind, the advantages of MCTA should be stressed. The main merit of MCTA is its speed, possible multi-phase scanning with simultaneous noninvasive angiography of pulmonary arteries to detect pulmonary embolism often found in such patients, lower costs and better availability compared to MRA. The experience of our centre shows that this examination should play an increasingly bigger role in emergency diagnostics when acute thrombosis of the subclavian or axillary vein is suspected and in follow-ups. Isolated thrombosis of the subclavian and axillary vein is two times rarer than the cases with concomitant brachiocephalic or superior vena cava thrombosis; moreover, secondary UEDVT is much more common than primary UEDVT and in such cases MSCTA is a complex, one-step-shopping method enabling prompt detection of the cause (e.g. compression by muscular or bony structures, mediastinal lymphoma) and life-threatening embolic complications. In many cases, its excellent visualization of collateral circulation pathways and contrasting of the adjacent arterial vessels may be of importance as well as possible digital processing of images and use in teleradiology systems (2).

The disease is treated with various therapeutic methods. In chronic cases, the improvement was observed after intensive heparinization and subsequent treatment with oral anticoagulants. In new cases, thrombolysis of embolic lesions and sometimes thrombectomy is recommended. Stents of venous vessels in this region are not recommended unless decompression was performed (e.g. the first rib resection due to high likelihood of thrombosis recurrence) (4, 9, 10). It was confirmed that the most effective management is primary thrombolysis with surgical decompression irrespective of the interval between both procedures (3, 7).

REFERENCES

1. Joffe H.V., Goldhaber S.Z.: Upper-extremity deep vein thrombosis. Circulation, 106, 1874, 2002.
2. Lawler L.P. et al.: Multi-detector row and volume-rendered CT of the normal and accessory flow pathways of the thoracic systemic and pulmonary veins. Radiographics, 22, S45, 2002.
3. Lindbald B. et al.: Deep vein thrombosis of the axillary-subclavian veins: epidemiologic data, effects of different types of treatment and late sequelae. Eur.J. Vasc. Surg., 2, 161, 1988.
4. Prandoni P., Bernardi E.: Upper extremity deep vein thrombosis. Curr. Opin. Pulm. Med., 5, 222, 1999.
5. Prandoni P. et al.: The long term clinical course of acute deep vein thrombosis of the arm: prospective cohort study. BMJ, 329, 484, 2004.
6. Prandoni P. et al. Upper extremity deep-vein thrombosis: Risk factors, diagnosis and complication. Arch. Inter. Med., 157, 57, 1997.
7. Pupka A. et al.: Zmiana strategii leczenia zakrzepicy żyły podobojczykowej w przebiegu zespołu uciskowego górnego otworu klatki piersiowej – rekomendacja kliniczna. Przegląd Flebologiczny, 11, 37, 2003.
8. Szarnecka-Sojda A.: Thoracic outlet syndrome. Acta Angiol., 10, 99, 2004.
9. Urschel H.C. Jr., Patel A.N.: Paget-Schroetter syndrome therapy: failure of intravenous stents. Ann. Thorac. Surg., 75, 1693, 2003.
10. Vijaysadan V. et al: Paget-Schroetter syndrome in the young and active. JABFP, 18, 314, 2005.

SUMMARY

Primary subclavian-axillary venous thrombosis is a rare disease occurring in young and healthy individuals. It is called Paget-Schroetter syndrome if the cause considered is microtrauma to the vascular wall related to physical effort or anatomical abnormalities of the superior aperture of the thoracic cavity found in such patients. The authors present the case of thrombosis of the

right axillary vein in a 28-year-old healthy female patient after heavy exertion. The diagnosis was based on a characteristic clinical picture and Doppler ultrasound findings. The treatment involved thrombolysis with a high dose (7.5 mln) of streptokinase administered for 5 h with the concurrent unfractionated heparin infusion continued for 96 h. The venous patency was restored and the symptoms subsided. In order to exclude the anatomical cause of thrombosis, the two-phase multislice CT angiography was performed with limbs in neutral position and on hyperabduction. During postural maneuver the subclavian vein in the costoclavicular space was found to be bent, compressed and constricted and the flow hampered. In our opinion, dynamic multislice CT is a useful method for evaluating the venous structures of the superior aperture of the thoracic cavity and may be applied in the diagnostics of Paget-Schroetter syndrome. Its additional advantage is the possibility of simultaneous assessment of the pulmonary arteries for embolism.

Zespół Paget-Schroettera u młodej kobiety. Opis przypadku

Pierwotna zakrzepica żylna podobojczykowo-pachowa jest rzadką chorobą, występującą u młodych, zdrowych ludzi. Jest określana jako zespół Paget-Schroettera, jeśli jako czynnik przyczynowy brane są pod uwagę mikrourazy ściany naczynia związane z wysiłkiem fizycznym oraz spotykane u tych chorych nieprawidłowości anatomiczne otworu górnego klatki piersiowej. Autorzy przedstawiają opis zakrzepicy prawej żyły pachowej, która wystąpiła u 28-letniej, dotychczas zdrowej kobiety po intensywnym wysiłku. Rozpoznanie postawiono na podstawie charakterystycznego obrazu klinicznego oraz ultrasonografii dopplerowskiej. W leczeniu zastosowano trombolizę dużą dawką (7,5 mln) streptokinazy, podaną w ciągu 5 godzin z towarzyszącym wlewem niefrakcjonowanej heparyny utrzymywanym przez 96 godzin. Uzyskano pełne udrożnienie żyły i cofnięcie objawów. W celu wykluczenia anatomicznej przyczyny zakrzepicy wykonano dwufazową angiografię metodą wielorzędowej tomografii komputerowej przy neutralnej pozycji kończyny oraz podczas hiperabdukcji. W trakcie fazy badania wykonanej z uniesioną kończyną stwierdzono zagięcie, ucisk i zwężenie żyły podobojczykowej w przestrzeni trójkąta żebrowo-obojczykowego z objawami utrudnionego przepływu. Dynamiczna wielorzędowa tomografia komputerowa w opinii autorów jest przydatną metodą oceny struktur żylnych otworu górnego klatki piersiowej i może być wykorzystana w diagnostyce zespołu Paget-Schroettera. Dodatkową zaletą jest możliwość równoczesnej oceny tętnic płucnych w kierunku zatorowości.