G Chir Vol. 39 - n. 2 - pp. 101-106
March-April 2018

clinical practice

Superior vena cava syndrome due to central port catheter thrombosis: a real life-threatening condition

G.D. LIANOS[1,2*], N. HASEMAKI[2*], E. TZIMA[3], G. VANGELIS[2],
A. TSELIOS[3], I. MPAILIS[3], E. LEKKAS[2]

SUMMARY: **Superior vena cava syndrome due to central port catheter thrombosis: a real life-threatening condition.**

G.D. LIANOS, N. HASEMAKI, E. TZIMA, G. VANGELIS, A. TSELIOS, I. MPAILIS, E. LEKKAS

Superior vena cava syndrome (SVCS) represents undoubtedly a rare life-threatening condition. Herein, we present a rare case of a 69-year-old woman, with a history of hepatic flexure tumor and an indwelling central venous port, presenting with acute signs and symptoms of SVCS due to thrombosis of the catheter. The patient was treated with intravenous anticoagulation and fibrinolytic therapy and showed regression of symptoms. It is reported that central venous catheters are routinely used in clinical practice mainly in oncological cases for chemotherapy, parenteral nutrition or dialysis. However, complications related to implantation technique, care, or maintenance of these catheters may arise.

High index of suspicion for SVCS should always arise when a patient presents with common symptoms and long-term central catheters, in order to avoid unfavorable outcomes. Local thrombolysis appears to be a safe and effective therapy for port catheter-associated thrombosis.

KEY WORDS: Central venous catheter - Thrombosis - Superior vena cava syndrome - Port catheter.

Introduction

Superior vena cava syndrome (SVCS) is an extremely rare and life-threatening condition, caused by malignant tumors in 80%-90% of all cases. Interestingly, lung cancer accounts for about 90% of malignant tumors causing SVCS, while the incidence of SVCS in lung cancer patients is 3-5% (1). Other primary mediastinal tumors that may cause SVC syndrome include thymoma, Hodgkin disease, and lymphosarcoma, while metastatic tumors from the breast, thyroid or melanoma, rarely may cause superior vena cava (SVC) obstruction. In addition, all the large benign mediastinal masses and atrial myxoma have been implicated with this rare and life threatening clinical entity (2). Other than malignancies, thrombotic conditions, either idiopathic or associated with polycythemia, mediastinal infection, or indwelling catheters represent unusual causes of SVC syndrome (2). It has to be highlighted that SVCS may present with various symptoms. The complete vein occlusion is associated with arm and facial swelling, stridor, blurred vision, dyspnea, dizziness, positional headache, retroorbital pain, dysphagia, and chest pain, while very rarely the SVC thrombosis is asymptomatic. Herein, we deal with an extremely rare case of SVCS, induced by improper maintenance of central venous port catheter.

Case report

A 69-year-old Caucasian female patient was admitted to our emergency department due to severe headache, dyspnea and shortness of breath of recent onset. The patient was diagnosed three years earlier with a 'large' neoplasm of hepatic flexure. She sub-

[1] Department of Surgery, University Hospital of Ioannina and University of Ioannina, Ioannina, Greece
[2] Department of Surgery, General Hospital of Preveza, Preveza, Greece
[3] Department of Internal Medicine, General Hospital of Preveza, Preveza, Greece
*equal contributors

Corresponding author: Georgios D. Lianos, e-mail: georgiolianos@yahoo.gr

sequently underwent exploratory laparotomy, where an extended right hemicolectomy was performed. The postoperative period was uneventful. Chemotherapy was then followed, for which a central venous port catheter was placed. She maintained regular surveillance, and remained disease-free for the next three years.

Three weeks prior to presentation, the patient stated that she was suffering from head pressure that worsened when lying on the right side, headache when bending forward, orthopnea, nausea and dizziness. The patient denied any history of cough, hemoptysis, chest pain, or wheeze. She also denied receiving any medication. The patient also stated that the central venous port catheter had not been removed after the completion of chemotherapy, and she gave reliable information indicating that it had last been flushed about four-five months earlier.

On physical examination, her pulse was 90/min, her blood pressure (BP) 110/80 mmHg, temperature 35°C, respiratory rate 30/min, O_2 saturation (SpO_2) 93% on room air and 98% with 3 L/min of oxygen by nasal prongs, while she presented jugular venous distention, and mild edema of the upper extremities, head, and neck. There where unremarkable findings by the examination of heart, lung and abdomen. The laboratory examination revealed white blood cell (WBC) count of 3000 x $10^3/\mu L$, while hemoglobin (Hgb), hematocrit (HCT) and the platelet (PLT) count were within normal limits. The plasma coagulation study was normal, and the clinical chemistry study revealed elevated serum creatinine concentration and BUN (1,58 mg/dL and 133 mg/dL respectively). All other values were within normal limits.

During the patient stay at the outpatient clinic, she presented severe edema of the neck and face accompanied by cyanosis of these areas and jugular venous distention (Figure 1). On physical examination, the patient was tachypneic and in moderate respiratory distress, the hemodynamically status of the patient had changed, her BP was now 70/40 mmHg, her pulse 130/min and SpO_2 was 90% with 3L/min of oxygen by nasal prongs. On physical examination she was afebrile, and had regular and rhythmic cardiac activity with facial cyanosis, and edema of head and neck with visible collateral circulation. Arterial blood gas analysis showed decreased oxygen saturation, high level of CO_2, and acidosis. A chest X-ray (posteroanterior view) was performed, showing the central venous port catheter in place in the right subclavian vein, while there was no evidence of pulmonary venous obstruction, mass lesion or hilar enlargement.

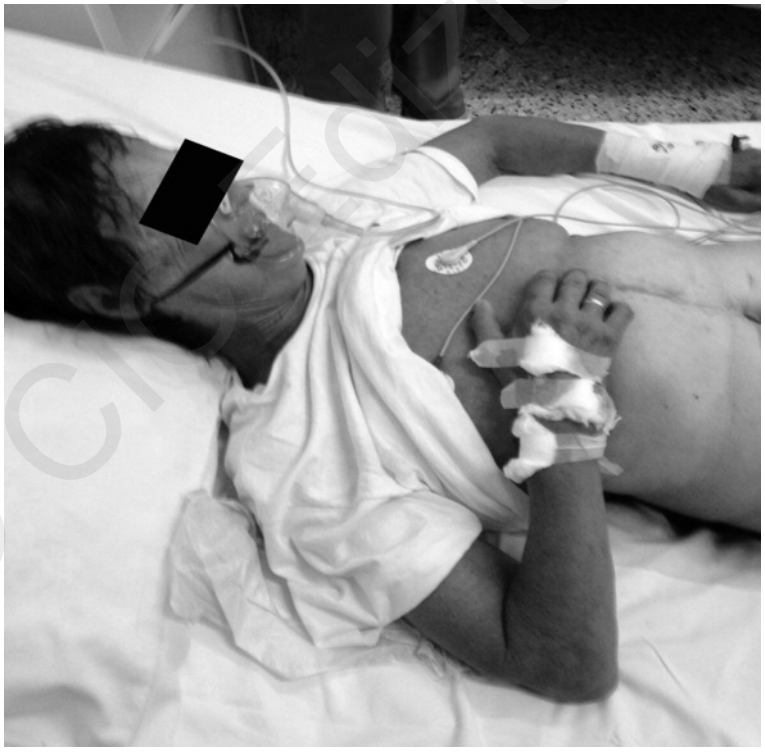

Figure 1 - SVCS (superior vena cava syndrome).

She was assessed for SVC syndrome, followed by an urgent computed tomography angiography (CTA) scan, revealing thrombus at the distal tip of the catheter, thrombotic plugging of the left subclavian vein, and stenosis of the SVC, due to thrombosis, with near total obstruction of venous return to the chest from SVC (Figures 2, 3, 4, 5). No signs of an acute pulmonary embolism or thoracic aortic aneurysm are detected. Thereby, the patient was finally diagnosed to have SVC syndrome due to

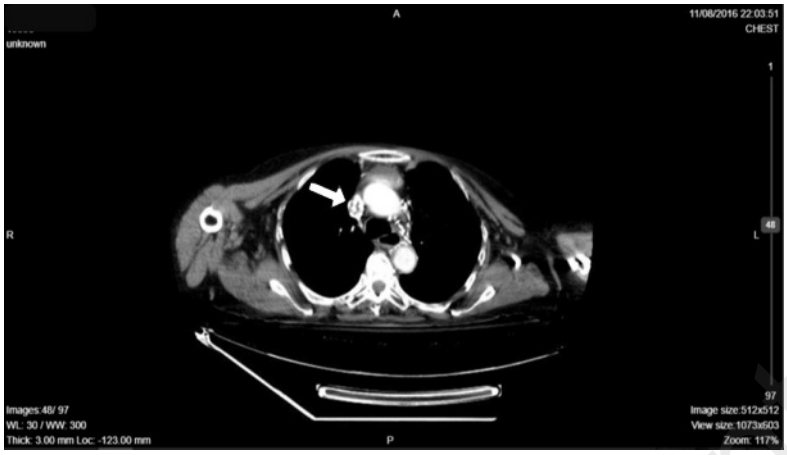

Figure 2 - CT image of superior vena cava thrombosis.

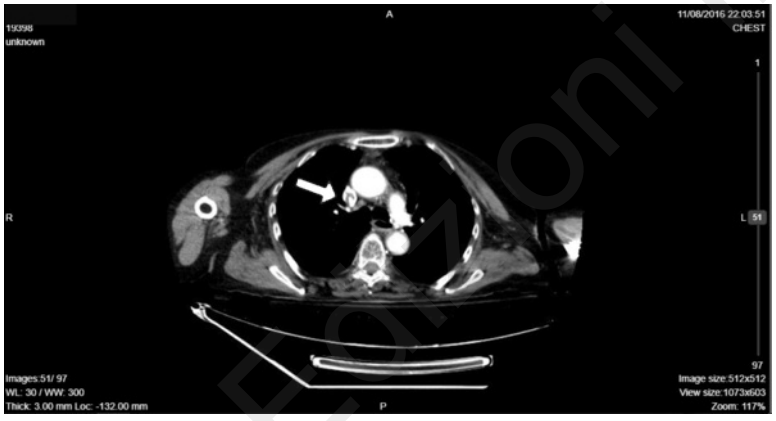

Figure 3 - Superior vena cava trombosis.

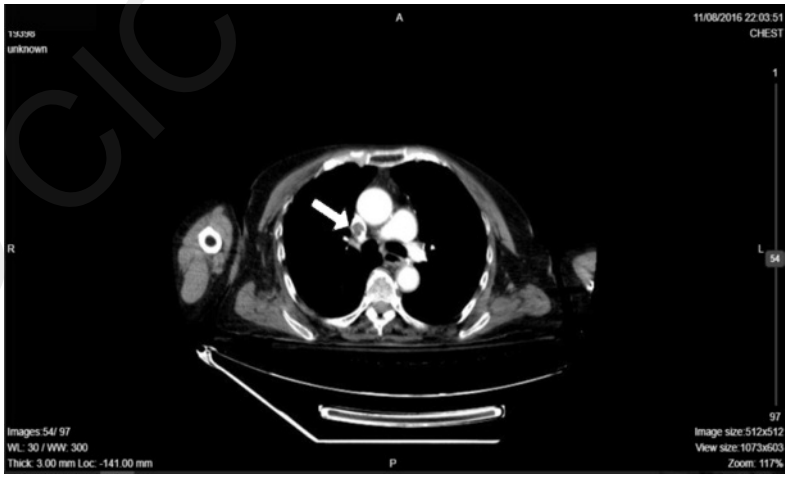

Figure 4 - Superior vena cava with complete trombosis.

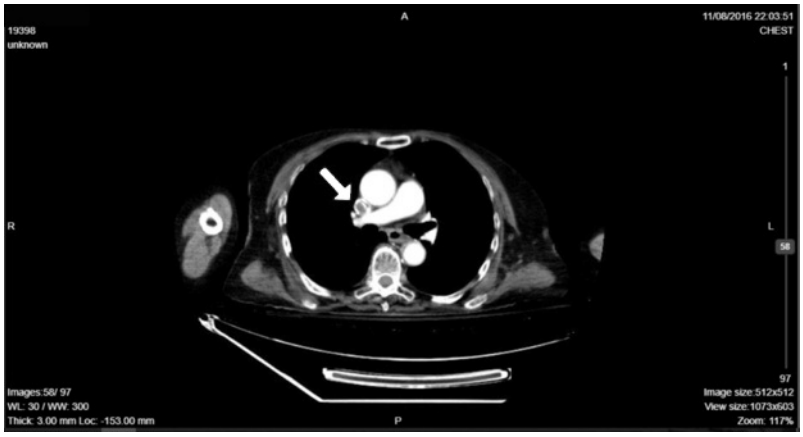

Figure 5 - Another CT image of superior vena cava thrombosis.

thrombosis of the central port venous catheter.

The patient was treated conservatively, with intravenous anticoagulation with continuous heparin infusion, intravenous fibrinolysis with tenecteplase and dexamethasone. She was admitted to the high dependency unit of our hospital, and close monitoring of patient's vital signs was performed. Complete blood count, arterial blood gases, and clotting profile were monitored. The patient showed clinical improvement, with regression of the edema and facial cyanosis, while vital signs returned to normal within several hours. The next day the patient was referred to cardiothoracic surgical department, hemodynamically stable, with complete regression of clinical signs and symptoms of SVC syndrome, in order to assess the need of surgery and catheter removal.

Discussion

Following its first description by William Hunter in 1757 (3), SVCS has multiple varying etiologies. Benign etiologies historically have accounted for up to 22% of cases of superior vena cava (SVC) syndrome (4-6). A recent report suggests that benign etiologies may now comprise up to 40% of cases (7, 8). In the past few decades, permanent indwelling central venous catheters (CVC) attached to an implanted reservoir, or "port systems" have been widely used to facilitate the care of oncological patients. It is believed that this high prevalence of CVC-induced SVC syndrome, can be explained by the fact that a plethora of today's medical treatments necessitate the use of such devices, often in patients with underlying conditions that increase the susceptibility for clot formation.

Despite a low overall risk, it has been reported that approximately 15% of patients have been experienced adverse events following catheter insertion, including infection, hematoma, thrombosis, and pneumothorax (9, 10). The most usual complications after central port system implantation are local infection at puncture site, infection in the subcutaneous pocket around the port, fever, catheter fracture or dislocation, cardiac arrhythmias, extravasation, thrombosis, and pneumothorax. In particular, thrombosis occurs in 2% to 21% of patients (10-12).

The clinical manifestations of SVCS, depend on the abruptness of onset, the location of the obstruction, the completeness of occlusion and the availability of collateral pathways. Clinical signs include cyanosis, plethora, jugular venous distention, and edema of the upper extremities, head and neck (13-16). Edema may compromise the function of the larynx or pharynx, causing dyspnea, stridor, cough, hoarseness, and dysphagia. A more serious consequence is cerebral edema, which may cause confusion, altered state of consciousness, and, possibly, coma. Cough, dyspnea, and orthopnea are common symptoms and may mimic congestive heart failure or pericardial disease. For a more detailed visualization of the SVC and its surrounding structures, a chest computed tomogram (CT) with intravenous contrast medium in the venous phase is highly recommended. CT can be used to diagnose the underlying pathology, including tumor mass size and localization. Superior vena cava diameter and length of SVC stenosis/occlusion can be determined, which constitutes a solid basis for planning a subsequent endovascular treatment.

In 1986, Stanford et al. (17) classified patients presenting SVC syndrome in four radiologic stages according to venographic pattern. In particular, type I representing mild SVC obstruction, with vessel obstruction of less than 90%, type II of high-grade SVC stenosis (grade of stenosis 90-100%) with permeable azygos vein and flow toward the right atrium, type III with complete SVC obstruction and prominent flow through collateral veins, but without involvement of the mammary and epigastric veins and type IV with complete SVC obstruction and prominent flow through collateral veins and the mammary and epigastric veins.

The classification of SVC syndrome by Stanford et al. was commonly accepted for the next decades, prior to a new grading system proposed by Yu et al. [18]. In particular, Yu et al. proposed an algorithm for the treatment of SVC syndrome from malignant causes, based on the presence or absence of potential life-threatening symptoms and performance status of the patient.

It has to be highlighted that in any patient with SVCS, the goal is to treat the underlying cause. Management options for patients with SVCS include treatment of the original malignancy, endovascular repair, anticoagulation and surgery (19-21). To the best of our knowledge, the largest series in the literature evaluating thrombolytic therapy for the treatment of thrombosed SVC associated with indwelling central venous catheters was conducted by Gray et al. in 1991 (22). They reported an 88% success rate if treatment was begun less than or equal to 5 days after the symptoms started.

Endovascular treatment and the use of stents is also considered to be an effective first line therapy to provide prompt symptomatic relief in cases, where SVCS is caused by a thrombus or stenosis of a catheter. In a retrospective study, Rizvi et al. (23) claimed that endovascular treatment represents a valid primary intervention in patients with SVCS of benign etiology.

To the best of our knowledge, the literature contains only a very few reports of such severe complications of a port catheter with complete thrombosis of the superior vena cava and development of a superior vena cava syndrome like in our case (24-30). Our case suggests that it is necessary a very high index of suspicion in order to identify SVCS and potentially avoid unfavorable outcomes.

Conclusion

It has to be highlighted that proper handling of a central venous port catheter is of crucial importance so that complications such as thrombosis and infections can be potentially avoided. As it is seen in our case, lack of knowledge about proper port care had serious and life threatening consequences. Although surgical treatment is a well-known strategy for the treatment of SVC occlusions, fibrinolytic and anticoagulation therapy may be an alternative effective strategy for port-associated thrombotic SVCS.

Conflict of interest

The Authors declare no conflict of interest.

References

1. Wilson LD, Detterbeck FC, Yahalom J. Superior vena cava syndrome with malignant causes. The New England Journal of Medicine. 2007;356(18):1862-1869.
2. Rizvi AZ, Kalra M, Bjarnason H, Bower TC, Schleck C, Gloviczki P. Benign superior vena cava syndrome: stenting is now the first line of treatment. Journal of Vascular Surgery. 2008;47(2):372-380.
3. William H. History of aneurysm of the aorta with some remarks on aneurysm in general. Med Observations Inq. 1757;323-357.
4. Klassen KP, Andrews NC, Curtis GM. Diagnosis and treatment of superior-vena-cava obstruction. AMA Arch Surg. 1951;63: 311-25.
5. Nieto AF, Doty DB. Superior vena cava obstruction: clinical syndrome, etiology, and treatment. Curr Probl Cancer. 1986;10:441-84.
6. Parish JM, Marschke RFJr, Dines DE, Lee RE. Etiologic considerations in superior vena cava syndrome. Mayo Clin Proc. 1981;56:407-13.
7. Rice TW, Rodriguez RM, Light RW. The superior vena cava syndrome: clinical characteristics and evolving etiology. Medicine (Baltimore). 2006;85:37-42.
8. Shaheen K, Alraies MC. Superior vena cava syndrome. Cleveland Clinic Journal of Medicine. 2012;79(6):410-412.
9. McGee DC, Gould MK. Preventing complications of central venous catheterization. N Engl J Med. 2003;348:1123-1133.
10. Merrer J, De Jonghe B, Golliot F, Lefrant JY, Raffy B, Barre E, et al. Complications of femoral and subclavian venous catheterization in critically ill patients: a randomized controlled trial. JAMA. 2001;286:700-707.
11. Caers J, Fontaine C, Vinh-Hung V, De Mey J, Ponnet G, Oost C, et al. Catheter tip position as a risk factor for thrombosis associated with the use of subcutaneous infusion ports. Support Care Cancer. 2005;13:325-331.
12. Warren P, Burke C. Endovascular management of chronic upper extremity deep vein thrombosis and superior vena cava syndrome. Seminars in Interventional Radiology. 2011;28(1): 32-38.

13. Bell DR, Woods RL, Levi JA. Superior vena caval obstruction: a 10-year experience. Med J Aust. 1986;145:566-568.
14. Lochridge SK, Knibbe WP, Doty DB. Obstruction of the superior vena cava. Surgery. 1979;85:14-24.
15. Palmer KG, King LG, Van Winkle TJ. Clinical manifestations and associated disease syndromes in dogs with cranial vena cava thrombosis: 17 cases (1989–1996). J Am Vet Med Assoc. 1998;213:220-224.
16. Park DR, Pierson DJ. Disorders of the mediastinum. In: Murray JF, Nadel JA, eds. Textbook of Respiratory Medicine. 3rd ed. Philadelphia: WB Saunders; 2000:2079-2094.
17. Stanford W, Doty DB. The Role of Venography and Surgery in the Management of Patients with Superior Vena Cava Obstruction. Ann Thorac Surg. 1986;41:158-163.
18. Yu JB, Wilson LD, Detterbeck FC. Superior vena cava syndrome - a proposed classification system and algorithm for management. J Thorac Oncol. 2008;3:811.
19. Kvale PA, Selecky PA, Prakash UB; for the American College of Chest Physicians. Palliative care in lung cancer: ACCP evidence-based clinical practice guidelines (2nd edition). Chest. 2007;132:368-403.
20. Streiff MB. National Comprehensive Cancer Center Network. The National Comprehensive Cancer Center Network (NCCN) guidelines on the management of venous thromboembolism in cancer patients. Thromb Res. 2010;125:128-133.
21. Pennell N, Hodgson JM, Bahler R, Mueller D, Sisson SD. Superior Vena Cava Syndrome. Elsevier; 2013.
22. Gray BH, Olin JW, Graor RA, Young JR, Bartholomew JR, Ruschhanpt WF. Safety and efficacy of thrombolytic therapy for superior vena cava syndrome. Chest. 1991;99:54-59.
23. Rizvi AZ, Karla M, Bjarnason H, Bower TC, Schleck C, Gloviczki P. Benign superior vena cava syndrome: stenting is now the first line of treatment. J Vasc Surg. 2008 Feb;47(2):372-80.
24. Dumantepe M, Tarhan A, Ozler A. Successful treatment of central venous catheter induced superior vena cava syndrome with ultrasound accelerated catheter-directed thrombolysis. Catheter Cardiovasc Interv. 2013 Jun 1;81(7): E269-73.
25. Shaikh I, Berg K, Kman N. Thrombogenic Catheter-Associated Superior Vena Cava Syndrome. Case Rep Emerg Med. 2013;2013:793054.
26. Thapa S, Terry PB, Kamdar BB. Hemodialysis catheter-associated superior vena cava syndrome and pulmonary embolism: a case report and review of the literature. BMC Res Notes. 2016;9:233.
27. Santos PK, Fernandes AM, Figueiredo V, Janeiro S. Superior vena cava syndrome associated with longstanding implantable central venous port. BMJ Case Rep. 2015;2015:bcr2014206643.
28. Seo M, Shin W-J, Jun I-G. Central venous catheter-related superior vena cava syndrome following renal transplantation - A case report-. Korean J Anesthesiol. 2012 Dec; 63(6):550-554.
29. Venturini E, Becuzzi L, Magni L. Catheter-Induced Thrombosis of the Superior Vena Cava. Case Rep Vasc Med. 2012; 2012:469619.
30. Hasskarl J, Koberich S, Frydrychowicz A, Illerhaus G, Wallwe CF. Complete caval thrombosis secondary to an implanted venous port-a case study. Dtsch Arztebl Int. 2008 Jan;105(1-2):18-21.