CASE REPORT

THYMOMA PRESENTING AS SUPERIOR VENA CAVA SYNDROME

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ABSTRAK

Pendahuluan

Timoma adalah neoplasma yang jarang terjadi yang berasal dari sel epitel timus dengan prevalensi 0,5-1,5% dari semua keganasan di Amerika Serikat. Pada 2018-2023 ditemukan 20 kasus pasien timoma yang dirawat di RSUP M Djamil. Gejala klinis dapat berupa nyeri dada, dispnea, batuk, atau sindrom vena kava superior akibat efek pendesakan pada organ yang berdekatan.

Ilustrasi Kasus

Laki-laki, 28 tahun dirawat di Bagian Penyakit Dalam RSUP Dr. M. Djamil Padang mengeluhkan sesak nafas yang hebat sejak 3 hari sebelum masuk RS disertai dengan bengkak pada leher, wajah, dan tangan kanan. Pemeriksaan fisik didapatkan keadaan umum berat, takipneu, dan pemberton sign positif. Pada pemeriksaan laboratorium didapatkan hasil dalam batas normal. Pemeriksaan CT Scan thoraks dengan kontras didapatkan tumor mediastinum anterior yang menekan vena kava superior. Dilakukan pemberian kortikosteroid dan radioterapi cito untuk keadaan emergency sindroma vena kava superior (SVKS). Setelah sesak berkurang, dilakukan pemeriksaan transthoracic needle aspiration (TTNA) dan trans thoracic biopsy (TTB) dengan hasil timoma. Selanjutnya pada pasien dilakukan kemoterapi sebagai terapi primer timoma karena massa tumor yang non-resectable.

Diskusi

Kasus ini menarik karena keberhasilan terapi pada pasien. Radioterapi dapat memberikan perbaikan klinis pada pasien SVKS kemudian dilakukan kemoterapi menggunakan regimen cisplatin, doxorubicin, siklofosfamid karena tumor non-resectable. Pada follow up setelah kemoterapi didapatkan perbaikan klinis dan pengecilan massa tumor pada pasien secara radiologis.

Kesimpulan

Timoma adalah tumor yang jarang dengan mortalitas tinggi. Pengobatan timoma harus dilakukan oleh tim multidisiplin untuk menentukan strategi perawatan yang tepat. Pilihan pengobatan kemoterapi memberikan perbaikan klinis pada pasien dengan massa yang non-resectable

Kata kunci: timoma, SVKS, radioterapi, kemoterapi

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INTRODUCTION

Thymoma is considered as a relatively rare type of tumor and has an incidence rate of only 0.15 cases per 100,000 in the global population. However thymoma is the most common primary neoplasm of the anterior mediastinum.[1,2] Thymoma is one of the well-known causes of superior vena cava syndrome. The majority of SVCS cases caused by thymomas are developed by the extrinsic compression of the superior vena cava.[3] It is critical to determine at the early stage whether the tumor is surgically resected or not. Therefore the optimal plan of care for patient should be developed before treatment.[4]

CASE REPORT

A 28-year-old male patient was referred to emergency department with severe shortness of breath in the last 3 days before admission. He presented hoarseness, with severe edema of the face and upper half of the body with numerous dilated cutaneous veins. On general physical examination, patient was conscious ($E_4V_5M_6$), his blood pressure was 110/60 mmHg, heart rate was 110 beats per minute, and respiratory rate was 28 breaths per minute. We found plethora with positive Pemberton's sign accompanied by increased jugular veins and venectations on the chest wall.

On physical examination of the lungs, tactile fremitus decreased in left hemithorax at RIC II-VI levels and right upper hemithorax, dullness to percussion and decreased breath sounds in the left hemithorax at RIC II - VI levels. Laboratory examination results were within normal limits. The initial chest X-ray showed a suspected mass in the mediastinum (Figure 1).

Thoracic CT scan with contrast found an anterior mediastinal tumor with irregular borders with a size of 14.2 cm x 10.9 cm x 10.8 cm pressing on the superior vena cava (Figure 2). Patient was treated with corticosteroids and emergency radiation therapy for an emergency superior vena cava syndrome (SVCS).

Radiation therapy was performed as the fastest way to relieve obstruction in patient with life threatening malignancy related SVCS. The fractional include initial fractions of 3 Gy and followed by daily fractions of 2 Gy. Diagnostic transthoracic needle aspiration (TTNA) and transthoracic biopsy (TTB) were examined after the patient improve clinically by initial fractions of radiation therapy. Pathologic findings confirmed his diagnosis as a stage IV thymoma according to Masaoka staging. Surgery is recommended for thymoma but the tumor location is too closed to vital structures. Chemotherapy was performed as modality treatment for this patient due to a non-resectable tumor. Cisplatin, doxorubicin, cyclophosphamide regimen are administered repeated every 3 weeks. The patient completed a total of 4 cycle of chemotherapy. The patient is doing well now. The plan is to give him two more cycles of chemotherapy and follow up regularly every 6 months for a few years, followed by chest imaging.

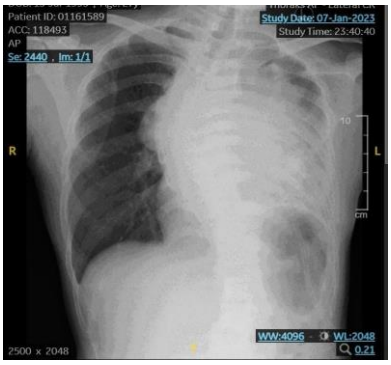

Figure 1. Chest X-ray with suspected mediastinum mass

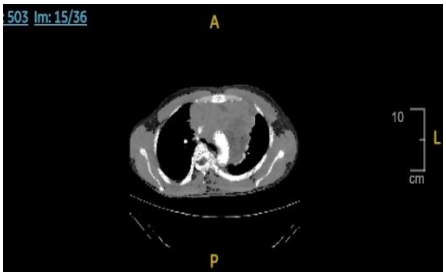

Figure 2. CT Scan Thorax demonstrates compression of SVC

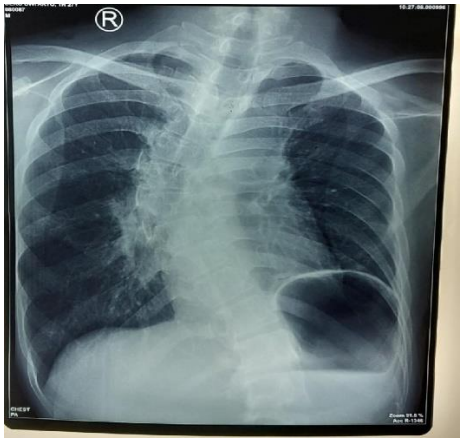

Figure 3. Chest X-ray post 4th cycle of chemotheraphy

DISCUSSION

The superior vena cava is formed by the junction of the left and right innominate (brachiocephalic) veins and is tasked with returning blood from the head, neck, upper extremities, and torso back to the heart.[5] Anatomically, the superior vena cava is located in the middle of the mediastinum, slightly across to the right of the midline and is surrounded by rigid structures such as the sternum, trachea, right bronchus, aorta, pulmonary artery, and paratracheal and perihilar. Superior vena cava carry blood from the head, neck, upper extremities, and chest to the right atrium with 1/3 of the total volume of venous return to the heart. The thin walls of the superior vena cava make their walls easy compressed. Acute obstruction of the superior vena cava can cause an increase significant intracranial pressure up to 40 mm Hg from normal pressure 2-8mm Hg. SVCS is the condition of emergency that occurs when there is obstruction of blood flow in the path from great veins draining into the superior vena cava.[6] This syndrome is most commonly secondary to malignancy.[7]

Thymoma presents uncommonly as SVCS. Thymoma in this patient was confirmed by Thoracic computed tomography examination with contrast. Computed tomography is the most common imaging procedure to detect thymoma and can help differentiate thymomas of other anterior mediastinal disorders.[1]

Thoracic CT scan with contrast found an anterior mediastinal tumor with irregular borders with a size of 14.2 cm x 10.9 cm x 10.8 cm pressing on the superior vena cava. Cause of the obstruction in SVCS may vary widely such as extrinsic compression, intrinsic stenosis, or thrombosis of the vein.[6] In this case, the obstruction caused by thymoma. Radiation therapy can provide clinical improvement in SVCS patients.

Evidenced based guidelines for the management of SVCS are not available.[8] Most of the options regarding the therapeutic approach SVC obstruction are obtained from case series and randomized trials.[9] Radiation therapy is the most important therapy and is widely recommended in patients with superior vena cava syndrome. Initiation of radiation therapy is the fastest way to relieve obstruction in patients with life-threatening symptoms.[10] The patient experienced clinical improvement after initial radiation theraphy. The reduction of post-radiation symptoms can begin felt within 72 hours and usually maximized in 2 weeks, on average 5-9 days.

The initial radiation therapy used larger dose fractions of 3 Gy for the first 2-3 days, followed by conventional fractions of 2 Gy to finally reach the total required dose.[11] Diagnostic transthoracic needle aspiration (TTNA) and trans thoracic biopsy (TTB) were examined with thymoma as a result.

The optimal plan of care for patient with thymoma should be developed by radiation oncologist, internal medicine, thoracic surgeon, respirologist, medical oncologist, and diagnostic imaging specialist.[12] The treatment of choice for thymoma in this patient is chemotherapy. This matter consider whether the mass of a thymoma is resectable or non-resectable. Therapy of thymoma should be based on the resectability of tumor. If the resection can be

performed, then surgery is the main option therapy, followed by postoperative radiotherapy or chemotherapy.

Primary chemotherapy is standard in non-resectable tumor. No randomised studies have been conducted and which regimen should be considered standard remains unknown. Multiagent combination regimens and anthracycline-based regimens appear to have improved response rates compared with etoposide-based regimens. Combination of cisplatin, doxorubicin and cyclophosphamide is preferred.[13]

Cisplatin, doxorubicin, cyclophosphamide (PAC) regimen was chosen because it causes few side effects. [14,15] At follow-up after chemotherapy, clinical improvement and radiological reduction of the tumor mass were obtained.

CONCLUSION

Thymomas are rare tumors with high mortality. Several possibilities can cause SVCS. The management of thymoma can be challenging. The primary therapy for thymoma is surgery, however, in non-resectable tumor, chemotherapy can be used. Treatment of thymoma must be carried out by a multidisciplinary team to determine the appropriate treatment strategy. Chemotherapy treatment provides clinical improvement and radiological reduction in patients.

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