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Case Report

Malignant Abdominal Mesothelioma Presenting with Features of Superior Vena Cava Syndrome. A Case Report

Athanasia Proklou[1], Evaggelos Maridakis[1], Evaggelia E Vassalou[2], Eumorfia Kondili[*1]

[1]*ICU, University Hospital of Heraklion, Medical School, University of Crete, Heraklion, Crete, Greece*

[2]*Department of Medical Imaging, University Hospital of Heraklion, Medical School, University of Crete, Heraklion, Crete, Greece*

**Corresponding author: Dr. Kondili Eumorfia, Associate Professor of Intensive Care Medicine, ICU, University General Hospital of Heraklion, University of Crete , Greece P.O. Box 1352, 71110 Heraklion, Greece, Tel.: +302810392585; ; E-mail: konde@med.uoc.gr*

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Abstract

Introduction

Superior vena cava (SVC) syndrome is life threatening clinical condition caused by obstruction of the SVC either by extrinsic compression or by internal thrombus. More than 80% of cases of SVCS are caused by malignant mediastinal tumors. Herein we report a rare case of superior vena cava syndrome as first manifestation of abdominal malignant mesothelioma.

Case Presentation

A 73-years-old man presented at the emergency department complaining of acute loss of vision within few hours, sore throat, shortness of breath, and feeling of swelling with signs of cyanosis of the upper limb, head and neck. A chest computed tomography following intravenous contrast administration revealed the presence of a well defined central thrombus in superior vena cava extending from the level of the right pulmonary artery to the proximal right internal jugular vein. A contrast enhanced CT of the abdomen revealed an enhancement of the diaphragmatic surface of the liver by soft tissue, and thickening of the mesentery, with abundant fluid between bowel loops. Histological examination of a specimen obtained by biopsy established the diagnosis of malignant mesothelioma.

Conclusions

We report a case of malignant peritoneal mesothelioma with leading symptoms and signs of Superior vena cava syndrome. Clinicians should be aware that malignant peritoneal mesotheliomas can present without any typical symptoms from the abdomen.

Keywords: Superior Vena Cava Syndrome; Thrombus; Peritoneal Malignant Mesothelioma; Laparoscopy

Introduction

Mesotheliomas are highly aggressive tumors arising from serous surfaces as pleura (65%-70%), peritoneum (30%), tunica vaginalis testis, and pericardium (1%-2%) [1] Peritoneal mesothelioma is a rare malignant tumor with a rapid fatal course and median survival 6-12 months. Asbestos exposure is the principal risk factor for the disease. It can occur in any age group, although the 50 to 69 year age group is the that most affected. It is more common in men, possibly because of the higher male occupational exposure

to asbestos. [2] Only 50% of patients with a peritoneal origin of malignant mesothelioma (MPeM) have a history of asbestos exposure, in contrast to 80% in mesotheliomas with pleural origin. [3]. Clinical presentation of malignant peritoneal mesothelioma includes pain and or swelling , in the abdomen, ascites, nausea ,vomiting, weight loss, fever , bowel obstruction and anemia. Herein we report a rare case of malignant abdominal mesothelioma presenting with features of Superior vena cava syndrome.

Case Presentation

A 73-years-old Caucasian male, lifelong non-smoker, presented at the emergency department complaining of acute loss of vision within few hours, sore throat, shortness of breath, and feeling of swelling with signs of cyanosis of the upper limb, head and neck. His physical examination revealed sinus tachycardia, with 90rpm, arterial hypertension and tachypnea. Laryngoscopy revealed gelatinous uvular edema, swelling of the right arytenoids and aryepiglottic folds without movement deficit.

A chest computed tomography following intravenous contrast administration showed the presence of a well defined central thrombus in superior vena cava extending from the level of the right pulmonary artery to the proximal right internal jugular vein with no signs of pulmonary embolism (Figure 1).Cardiac U/S demonstrated the presence of a mobile thrombus in the entrance of the right atrium and dilatation of the right heart.

The patient was admitted to the intensive care unit (ICU) and treated with anticoagulants for prevention of clot propagation and corticosteroids for the treatment of the laryngeal edema. A contrast enhanced CT of the abdomen was performed with evidence of encasement of the diaphragmatic surface of the liver by mildly enhancing soft tissue, and thickening of the mesentery. Moreover there was abundant fluid between the bowel loops and in the left paracolic gutter and a multicystic intrapelvic mass with enhancing internal septations (Figures 2, 3). However, accounting on the inconclusive cytologic findings from the diagnostic paracentesis of the exudative ascitic fluid, we decided to proceed with diagnostic laparoscopy. Histological examination of peritoneum specimen revealed scattered epithelioid malignant mesothelioma of abdomen origin.

Figure1: Chest computed tomography following intravenous contrast administration, in transverse planes (a, b,) and coronal reconstruction (c). There is a centrally located superior vena cava thrombus (arrowheads in a) extending to the proximal right internal jugular vein (arrow in c). There is also complete occlusion of the azygous vein (arrows in b). Note the extensive right chest wall and lateral thoracic collateral venous network (arrows in a).

Figure 1a

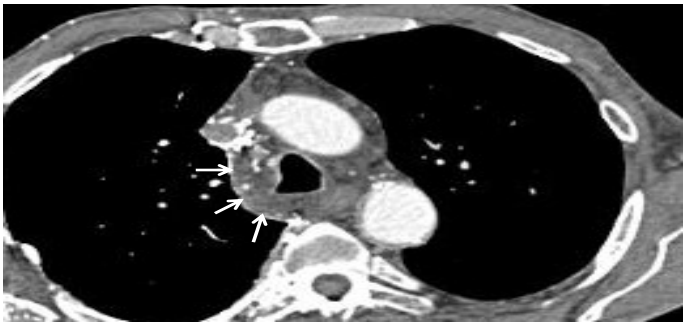


Figure 1b

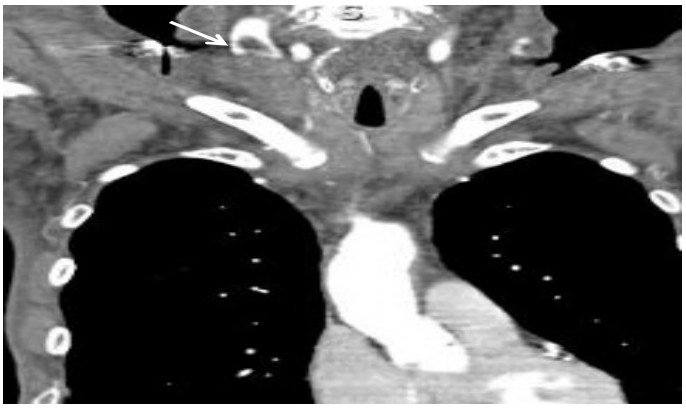
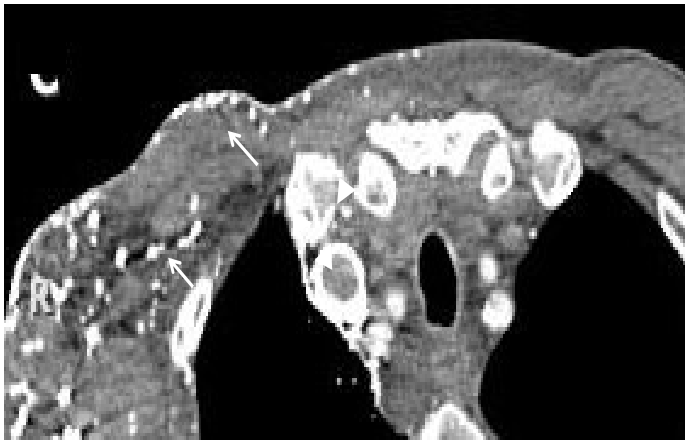


Figure 1c



During the course of hospitalization the patient's medical condition was deteriorated and was complicated with multiple organ failure. A multidisciplinary approach of the ICU with the oncology and surgical department was held. Due to the advanced state of the disease, any treatment approach was considered to be futile.

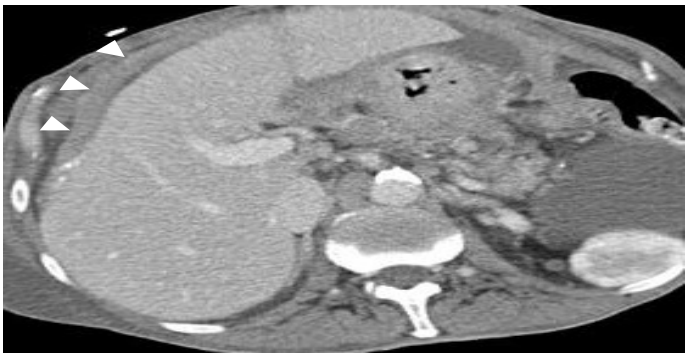


Figure 2. Abdominal contrast enhanced computed tomography images in transverse plane. There is enhancement of the diaphragmatic surface of the liver by mildly enhancing soft tissue (arrowheads)

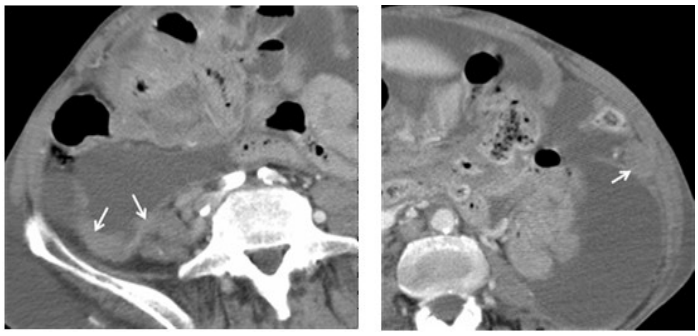


Figure3. Abdominal contrast enhanced computed tomography images in transverse plane shows multiple sites of nodular peritoneal thickening (arrows in a, b).

Discussion

Malignant peritoneal mesothelioma (MPM) is a rare aggressive tumor of the peritoneum, regarded as a fatal disease. The knowledge of its natural history is very limited. Occupational and environmental asbestos exposure still remains a public health problem around the world. However, only a small fraction (< 5%) of those heavily exposed to asbestos will develop MM, yet the disease has been observed to cluster within families.

Diagnosis of MPM may be suspected based on findings from computerized tomography (CT) scan or MRI findings, but a definitive diagnosis is confirmed either by cytology of ascitic effusion with 32% to 76% sensitivity, or histological examination of peritoneum biopsy.[4]

There are three main histological types of malignant mesothelioma. Epithelioid mesothelioma (56% of the cases), Sarcomatoid histotype (32%) and Biphasic (mixed) mesothelioma (13% of the total account) which usually shows both epithelioid and sarcomatoid components. Furthermore, a borderline malignant potential exists divided to well-differentiated papillary mesothelioma (WDPM) and benign multicystic mesothelioma (BMM). [5] MPM usually presents with abdominal pain, nausea, vomiting, ascites and a rapid weight loss fever and bowel obstruction. In our case there were no clinical signs from the abdomen and the leading symptoms were from the SCV syndrome.

Superior vena cava (SVC) syndrome life threatening clinical condition with characteristic and often striking clinical presentation. Up to 80 % of cases are related to the existence of malignant tumors of the mediastinum, mainly bronchial lung cancer and lymphoma and rarely metastatic malignasies of breast (Table 1). The presence of obstruction from extrinsic or intrinsic compression or luminal obstruction from neoplastic infiltration or in situ thrombosis are the cornerstone in the pathogenesis of the syndrome. As a consequence, the development of a collateral venous system is present arising from azygous, internal thoracic, lateral thoracic, paraspinous and esophageal venous. [6]

Table 1. Most common causes of Superior Vena Cava Syndrome

Malignant
• Lung cancer • Lymphomas • Thymoma • Mediastinal metastases • Mediastinal germ cell tumors • Mesothelioma • Leiomyosarcoma • Neoplastic thrombi • Thyroid cancer
Benign
• After radiation/idiopathic • Infectious disease: TBC, Histoplasmosis, Echinococcosis, Syphilis, Aspergillosis, Blastomycosis, Nocardiosis • Aortic aneurysm • Thrombosis/non malignant • Lymphadenopathies: sarcoidosis, Castelman's disease • Bronchogenic, pericardial, thymic cysts • Substernal goiter • Iatrogenic: Central venous catheters, pacemaker-defibrillator placement

In our case we believe that the SVC syndrome was due to in situ thrombosis due to malignant hypercoagulopathy state or thromboembolic disease. Hypercoagulopathy is common complication in patients with malignant disease. The incidence of clinical episodes of thromboembolism in patients with cancer varies from 1% to 11%[7]. In cases of malignant mesothelioma, is described that clotting abnormalities occur in 10-20%. [8]

Antman et al., have previously reported incidences of clotting abnormalities complicating the course of five of 23 patients with malignant peritoneal mesothelioma: disseminated intravascular coagulation in two, extensive thrombosis and concurrent pulmonary emboli in two, hemolytic anemia and

phlebitis in one. Carrington and Adams also reported a case of jugular vein thrombosis. [9]

Patients with confirmed MPeM, should undergo radical resection which is associated with better prognosis when possible. Other treatment options for peritoneal mesothelioma include intensive loco-regional therapeutic strategies: aggressive cytoreductive surgery, intraoperatively hyperthermic intraperitoneal chemotherapy (HIPEC), early postoperative intraperitoneal chemotherapy (EPIC), whole-abdominal radiation or adjuvant systemic chemotherapy and immunotherapy. [2] In this case the treatment decision was palliative care due to the patient's multiorgan failure and advanced phase of the disease.

Conclusions

The clinical presentation of our case is of interesting because the patient, although affected by a peritoneal mesothelioma, presented with SVC syndrome without complaining of symptoms referable to the abdominal disease. The prognosis in this case was poor, according to the data related to the SVC syndrome and the abdominal disease separately; median survival of untreated malignant SVC syndrome is ~30 days and for the untreated MPeM is 6 months.

References

1. Raptopoulos V. Peritoneal mesothelioma. Crit Rev Diagn Imaging. 1985, 24(4): 293–328.

2. Bridda A, Padoan I, Mencarelli R, Frego M. Peritoneal Mesothelioma: A Review. MedGenMed. 2007, 9(2): 32.

3. Busch JM, Kruskal JB, Wu B, Armed Forces Institute of Pathology Best cases from the AFIP. Malignant peritoneal mesothelioma. Radiographics. 2002, 22(6): 1511–1515.

4. Husain AN, Colby T, Ordonez N, Krausz T, Attanoos R et al. Guidelines for pathologic diagnosis of malignant mesothelioma. 2012 update of the consensus statement from the International Mesothelioma Interest Group. Arch Pathol Lab Med. 2013, 137(5): 647-67.

5. Tanida S, Kataoka H, Kubota E, Mori Y, Sasaki M et al. Combination chemotherapy with cisplatin and gemcitabine in malignant peritoneal mesothelioma. Int J Clin Oncol. 2009, 14(3): 266-269.

6. Rice TW, Rodriguez RM, Light RW. The superior vena cava syndrome: clinical characteristics and evolving etiology. Medicine (Baltimore). 2006, 85(1): 37-42.

7. Neschitz JE, Yeshurun D, Lev LM. Thromboembolism in cancer. Changing trends. Cancer.1993, 71(4): 1383-1390.

8. Antman KH, SciffPB, Pass HI. Benign and malignant mesothelioma. in: Cancer: principles and practice of oncology. 5th ed. De Vita VT Jr, Hellman S, Rosenberg SA, Eds. JB Lippincott, Philadelphia, 1997: 1853-1878.

9. Carrington BM, Adams JE. Jugular vein thrombosis associated with distant malignancy. Postgrad Med J. 1988, 64(752): 455-458.