Underestimated awakening face-edema caused by a mediastinal syndrome

Abstract

The "mediastinal syndrome" is an oncological urgency but, in some situations as in the case report described below, it can present itself in a subtle and inextricable way with symptoms and signs that, if singularly evaluated, can be nonspecific. In particular, we report the case of facial edema and a rash that, developing in an unusual and progressive way during about 2 months, concealed a voluminous mediastinal formation due to the localization of non-Hodgkin B-cell lymphoma that caused a "superior caval vein syndrome".

Introduction

Mediastinal syndromes are a group of diseases characterized by infiltration, entrapment or compression of the mediastinal structures; anatomically the mediastinum is divided into anterior, middle and posterior and the symptoms that characterize mediastinal syndromes are related to the involved structures. The causes of malignant origin, generally, include lymphomas, thymomas, germ cell tumors, thyroid neoplasms, and metastases from serous or mucinous tumors such as ovarian, gastrointestinal and small pulmonary cells; non-malignant causes include goiter or large aortic aneurysms. In particular, we speak of superior caval vein syndrome when the superior caval vein and nerves can be compressed or trapped, resulting in venous ectasia, edema of the face and upper extremities. Superior caval vein syndrome is the most severe complication of mediastinal syndromes and is considered a medical emergency [1-3].

Case Presentation

The patient, a 50-year-old woman with a history of pollen and food allergies, was suffering from morning episodes of facial edema that, for about 2 months, were treated with corticosteroids and antihistamines with initial apparent benefit in the suspicion of the allergic syndrome. The patient performed initial dermatological counseling after evaluation by her attending physician and over time subsequent dermatological and otorinolaringoiatric (ENT) evaluations that had always confirmed the ongoing therapy and was awaiting specific allergological evaluations. Over the weeks the patient noticed a worsening of the face and the appearance of a fleeting rash, despite therapy, so one night she turned to our Emergency Department where we only saw slight cutaneous hyperemia of the face without particular edema and gave her antihistamines and corticosteroids again with the following benefit. A chest radiograph was also performed and showed "...apical outcomes to the right with minimal pleural thickening associated and in-axis mediastinum..."; blood tests were essentially normal, including the leukocyte formula, for which patient was discharged with a diagnosis of "suspected urticaria regressed after medical treatment" and sent, by protected discharge, to allergological assessment.

About a week later, due to subjective dyspnea and increased rash despite ongoing therapy, the patient was again admitted to our Emergency Department. Se reported that the feeling of lack of air worsened by lying down and improved while standing during the day, as well as facial edema and neck rash. The objective examination, this time, proved jugular turgidity, redness, and edema of neck and shoulders; the patient showed some photos showing much more pronounced swelling of the face present above all on waking (after prolonged clinostatism) and a clear picture of mantle edema.

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Bed-side ultrasound was performed showing dilatation of jugulars with the slowed flow and slight circumferential pericardial effusion. At this point, mediastinal pathology was promptly suspected and so new thorax X-ray was performed and highlighted a "... flare of anterosuperior mediastinum with homogeneous opacity on retrosternal site on the LL projection..." **(FIGURE 1),** besides dosage of lactic-dehydrogenase, D-Dimer, and blood count. A chest CT scan with iodinate contrast was urgently performed and therefore was made a provisional diagnosis of "...massive expansive formation in the upper mediastinal site which infiltrates superior vena cava and anonymous left vein..." (**FIGURES 2 and 3**). Therefore patient was hospitalized for the appropriate assessments.

During the hospitalization, the patient was subjected to two subsequent needle aspirations which were "non-diagnostic" due to the presence of colliquated material. The final diagnosis of Non-Hodgkin B-Cell Lymphoma was performed by cytological and cytofluorimetric analysis of tissue taken by echoendoscopy (clone lambda B cells) for which the patient was transferred to the Hematology Department to carry out PET-TAC and the 1st cycle of R-CHOP chemoimmunotherapy.

Discussion

Symptoms of the mediastinal syndrome may often be nonspecific, including a dry cough, dyspnea, hemoptysis, chest pain, fatigue, and weight loss, especially when they occur gradually over many days (even over 2 months as in our patient). Although in this case, the symptoms were quite indicative of mediastinal pathology

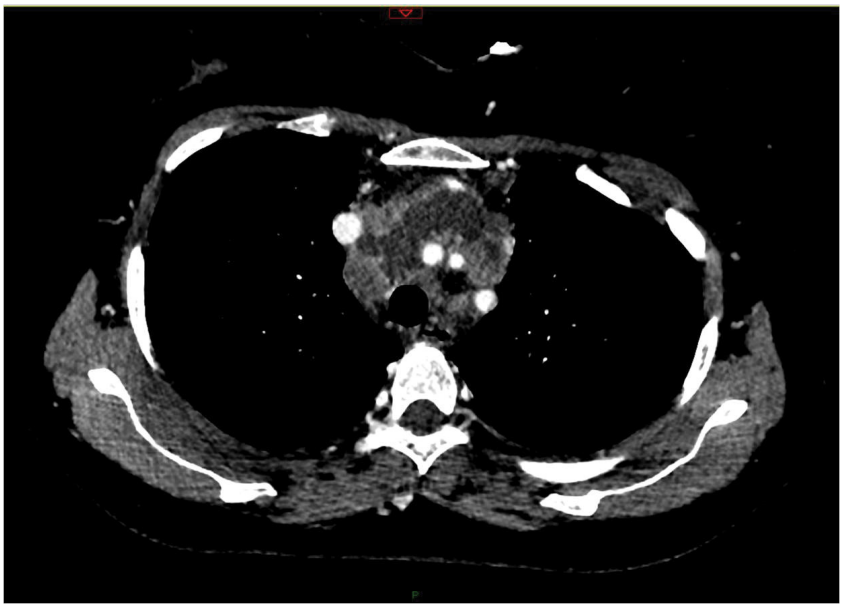

FIGURE 1. Flare of antero-superior mediastinum with homogeneous opacity on retrosternal site on the LL projection.

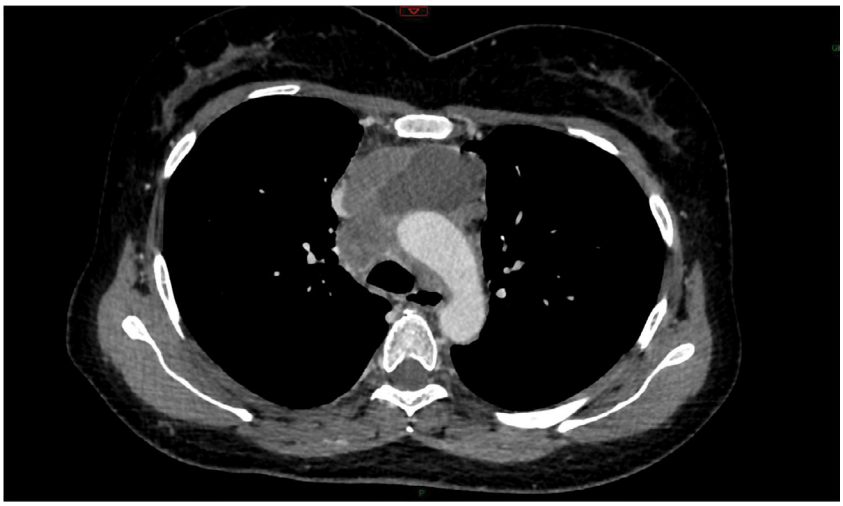

FIGURE 2. Massive expansive formation in upper mediastinal site which infiltrates superior vena cava and anonymous left vein.

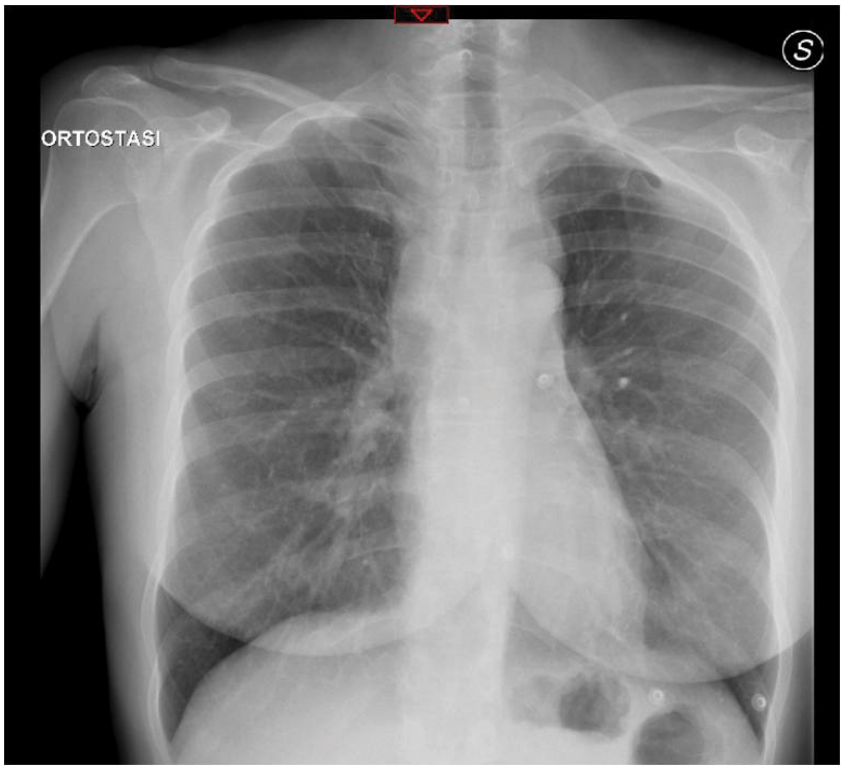

FIGURE 3. Massive expansive formation.

(edema and mantle rash), what probably led to not recognize them promptly was the fact that they presented themselves subtly and gradually, moreover in a young patient, and without comorbidity if not an allergic diathesis. The symptoms were assessed individually by the various specialists without framing the set of signs and symptoms in its entirety.

In conclusion, the patient was discharged from the Hematology Department and sent to Day Hospital to perform the 2nd cycle of chemoimmunotherapy with Rituximab subcutis which is currently well tolerated.

Acknowledgment

We deeply want to thank Mrs. M. who gave us permission to report her case. The patient explicitly asked that the photos she gave us should not be published, photos in which the edema and the rash she showed in the very first minutes after waking up were very evident and really impressive.

We thank the lady and wish her well for her therapy.

References

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