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Case Report

Diagnostic difficulties of antero-superior mediastinal masses with cardiac infiltration. Endovascular bioptic alternative approach [☆]

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ABSTRACT

The diagnosis of mediastinal masses is challenging due to the variety of possible pathologies, and its definitive diagnosis is mainly confirmed by histological evaluation. Sometimes some lesions may have a greater intravascular rather than mediastinal development and the collection of a biopsy sample becomes even more complex. In these cases endovascular catheter biopsy is helpful in the collection of the necessary biological material, having to adapt to the type of surface and consistency of the mass to be analyzed.

Endovascular catheter biopsy was performed with a biliary forceps to sample a mediastinal mass with greater endovascular and cardiac development, with a hard and difficult to sample surface. The histological result was diagnosed with non-hodgkins lymphoma.

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Introduction

Mediastinal masses are relatively uncommon yet include a large variety of entities, demonstrating a spectrum of clinical

and pathologic features that pose a diagnostic challenge for clinicians and radiologists [1].

The most frequent mediastinal lesions occur in the anterior compartment and include thymomas and lymphomas (more common in adults), neurogenic tumors, benign cysts,

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and germ cell neoplasms (more typical of childhood), which together account for 60% of patients with mediastinal masses. Typical symptoms at the onset of the disease are cough, chest pain, fever/chills, and dyspnea [2,3].

Imaging plays a crucial role in establishing a presumptive diagnosis in order to establish, if necessary, which type of confirmatory test to use. When the classic features are present, a presumptive diagnosis can be made with a high degree of confidence based on imaging alone. However, the appearance of anterior mediastinal lesions is often less specific. Nevertheless, when combined with a typical clinical presentation, a particular entity can be strongly suggested.

Formulating an appropriate differential diagnosis for a specific type of patient can be very helpful in avoiding unnecessary and sometimes misleading biopsies or additional tests [2].

The diagnostic process of a suspected mediastinal mass begins with an accurate medical history and detailed physical examination, supplemented by imaging and laboratory tests [4]. For this reason, it is imperative that clinicians perform a thorough history, physical examination, and comprehensive analysis of the patient, focusing on the symptoms and signs that are most often associated with mediastinal masses. Tumor markers are especially useful when a thymoma or germ cell cancer is suspected.

The evaluation of the *silhouette sign* on the chest radiography, which describes the loss of normal borders of intrathoracic structures, increases the sensitivity in detecting mediastinal abnormalities.

Computed tomography (CT) of the chest with intravenous contrast is typically used to evaluate abnormalities previously detected on plain radiographs. CT is useful in providing information such as mass location, size, compression or invasion of adjacent structures and tissue characteristics, including the presence of fat, fluid, or calcifications. All previous information can be critical in planning treatment and preparing a patient for possible resection.

Magnetic resonance imaging (MRI) is superior to CT in distinguishing cystic from solid masses (eg, thymic cysts from thymic neoplasms), in discerning cystic/necrotic components within solid masses, and thymic hyperplasia from thymic tumors.

In addition, MRI is useful for distinguishing compression from invasion, particularly in cases of large anterior mediastinal masses that are difficult to evaluate on CT, even when using an intravenous contrast medium.

However, the only way to reach a definite diagnosis or resolve diagnostic doubts is to obtain a histological diagnosis by biopsy of the mediastinal mass.

Ultrasound (US) or CT-guided percutaneous needle biopsy allows access to lesions at essentially all mediastinal sites. A direct mediastinal approach, which allows for extrapleural needle placement, is usually preferred to avoid the risk of pneumothorax and includes parasternal, trans-sternal, suprasternal, and paravertebral approaches depending on the precise location of the lesion detected on CT. An alternative approach involves advancing the needle through a pleural space created by an existing pleural effusion or iatrogenic pneumothorax. Another alternative method could be the transpulmonary approach, which involves the lung and

visceral pleura perforation by the needle but is nevertheless associated with a substantial risk of pneumothorax [5,6]. A further technique, even more rarely used, is the endovascular approach for the biopsy of masses invading the great mediastinal vessels and the heart.

We report here the case of a patient with a mediastinal mass with a major intravascular component, who underwent an endovascular biopsy performed with a biliary tract catheter. Histological examination diagnosed high-grade peripheral B-cell non-Hodgkin's lymphoma (NHL).

Case report

A 76-year-old woman was admitted to our hospital with worsening dyspnea for one month. On a physical examination, she had neck edema, oxygen saturation of 92% with oxygen therapy, blood pressure 127/60 mmHg, sinus tachycardia of 106 bpm and absence of arrhythmia in long-term monitoring.

Laboratory results were D-dimers 2537 ug/L (normal range, n.r. < 500 ug/L), brain natriuretic peptide 1785 pg/mL (n.r. 0-166 pg/mL), and cardiac troponin I 473.4 pg/mL (n.r. < 51.1 pg/mL).

Contrast-enhanced computed tomography (CT) revealed an anterosuperior mediastinal mass with right atrium involvement and deep vein thrombosis of the superior vena cava and left jugular, left subclavian, left anonymous and azygos veins. Pleural and pericardial effusion were associated (Fig. 1).

The echocardiogram described an atrial mass rather than a thrombus in the atrium. 18F-FDG Positron Emission Tomography (PET)/CT showed radiopharmaceutical uptake in the anterosuperior mediastinum with a maximum standardized uptake value (SUVmax) of 18.0, another area of FDG uptake in the right atrium (SUVmax 14.3) and, with a lower radiopharmaceutical accumulation, in the left armpit, peribronchial in the lower lobe of the right lung and lingula (SUVmax 5.3 to 3.3). No uptake was found in lymph node stations (Fig. 2).

Medical treatment with anticoagulant therapy (Clexane 6000) was therefore recommended.

A follow-up CT performed two days later showed complete reperfusion of the left jugular vein and partial reperfusion of the ipsilateral subclavian vein; however, the filling defect in the right atrium, superior vena cava, left anonymous vein, and azygos vein persisted.

The histological typing of the lesion was of primary importance in order to plan the most appropriate therapeutic strategy for this patient; however, there were no metabolically active lymph nodes on PET/CT examination, classically easier to sample, and the higher FDG uptake was in close proximity to the great mediastinal vessels and in the atrium.

The unusual location of the lesion, therefore, made choosing the correct biopsy approach extremely difficult. Cardiac surgeons did not recommend endovascular sampling, due to the high probability of failure and the risk of injury and perforation of the atrial chamber as the devices used to perform endomyocardial biopsies are intended for samples from the ventricles only and cannot be adapted to other sites.

After consulting with the interventional radiology team, the non-applicability of a classic direct mediastinal approach

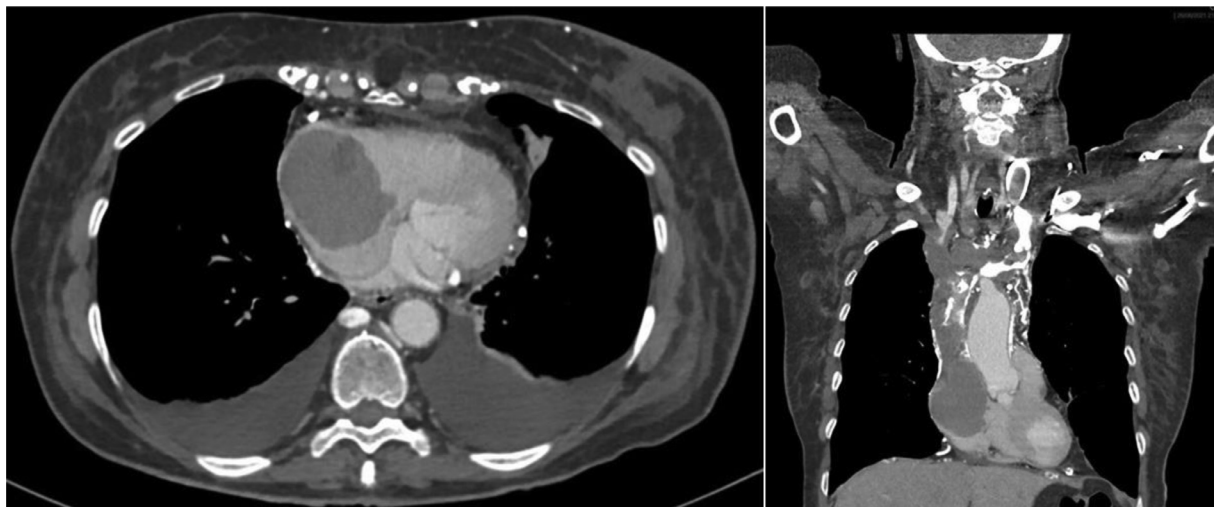


Fig. 1 – (A) Axial contrast-enhanced CT image shows a large filling defect in the right atrium. (B) Coronal contrast-enhanced CT image shows a massive irregularly filling defect involving the anonymous vein and superior vena cava.

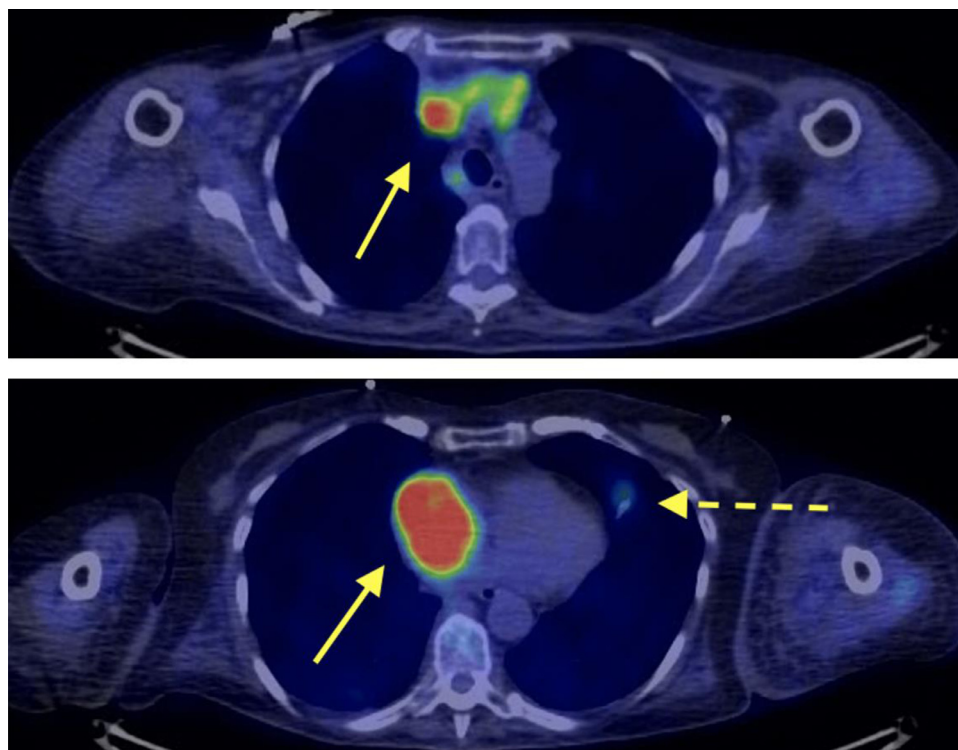


Fig. 2 – Axial FDG PET/CT images show radiotracer uptake within (A) the superior vena cava and (B) atrium and lingual, the latter evaluated as phlogistics on the co-registration CT images. FDG, fluorodeoxyglucose; PET, positron emission tomography; SUVmax, maximum standardized.

was established, due to the risk linked to the proximity of the mass to the cardiac vascular pedicle and to the high likelihood of failure as the lesion had too little extravascular extent compared to the intravascular one. Therefore, an endovascular approach was considered the best option.

Since it was necessary to use a device capable to overcome the hardness of the mass and collect enough sample, after a careful evaluation of the available equipment, the choice was

a transluminal Biliary Biopsy Forceps Set (Cook Medical) including Flexor Check-Flo Introducer Sheath (7 Fr, length 30 cm, with radiopaque band) and Flexible biliary biopsy forceps (5.2 Fr, length 60 cm, cup volume 2.25 mm³), commonly used for bile ducts biopsy. This could allow us to abrade the mass and obtain a good sample.

Preliminary angiography showed a massive opacification defect in the right atrium due to the mass (Fig. 3).

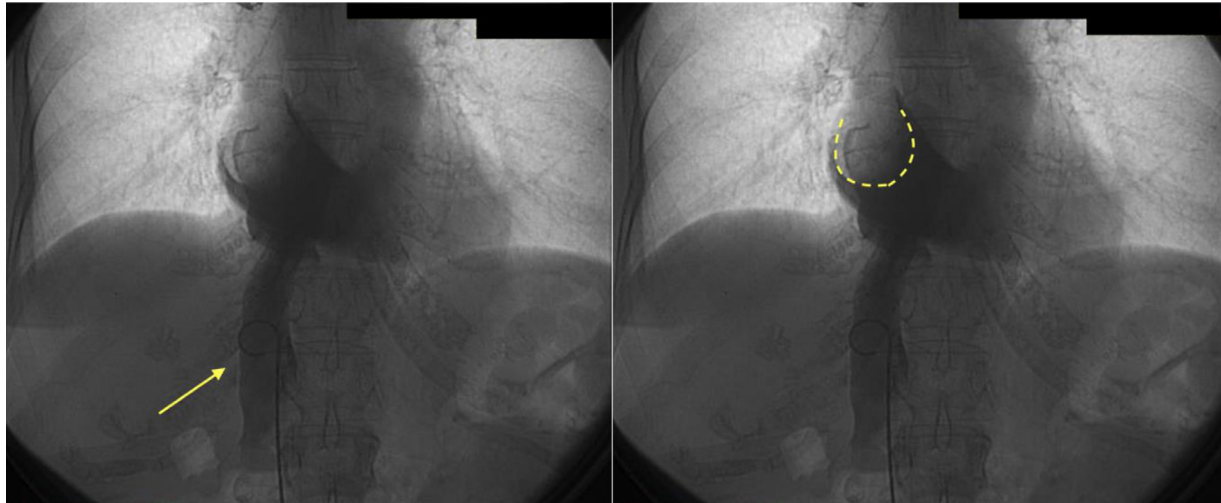


Fig. 3 – Frontal cardiac angiography revealed massive occlusion of the right atrium (dashed line).

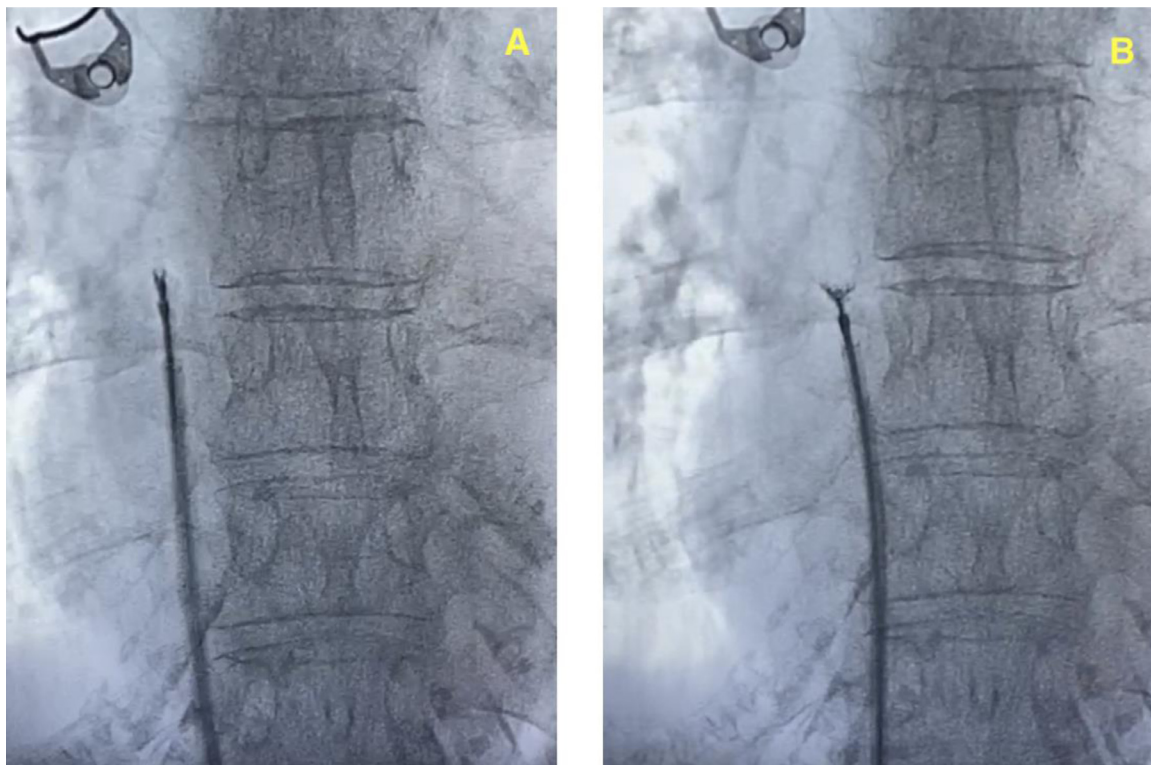


Fig. 4 – (A, B) Frontal cardiac angiography during the sampling with the miniforceps for the biliary tract.

An endovascular biopsy was performed under angiographic guidance. The biopsy forceps were advanced through the 7-Fr guide catheter to the right atrium. The mass was hard and resistant, but the biopsy was performed using the miniforceps for the biliary tract, stronger than other catheters. Three millimetric samples (2.25 mm³) were collected (Fig. 4).

After the procedure, angiography confirmed the absence of vascular injury (Fig. 5).

The samples obtained were diagnostic and the histological evaluation showed infiltration of lymphoid elements with marked nuclear coarctation and with a positive immunophe-

notype for CD20 + and negative for CK8 / 18-, CD3- and TdT-, very high (> 80%) proliferating ki67 + fraction. Immunomorphologic findings were suggestive of high-grade peripheral B-cell NHL.

Discussion

Primary mediastinal NHLs (PM-NHLs) are a relatively rare disease while classical Hodgkin's lymphoma is by far the most

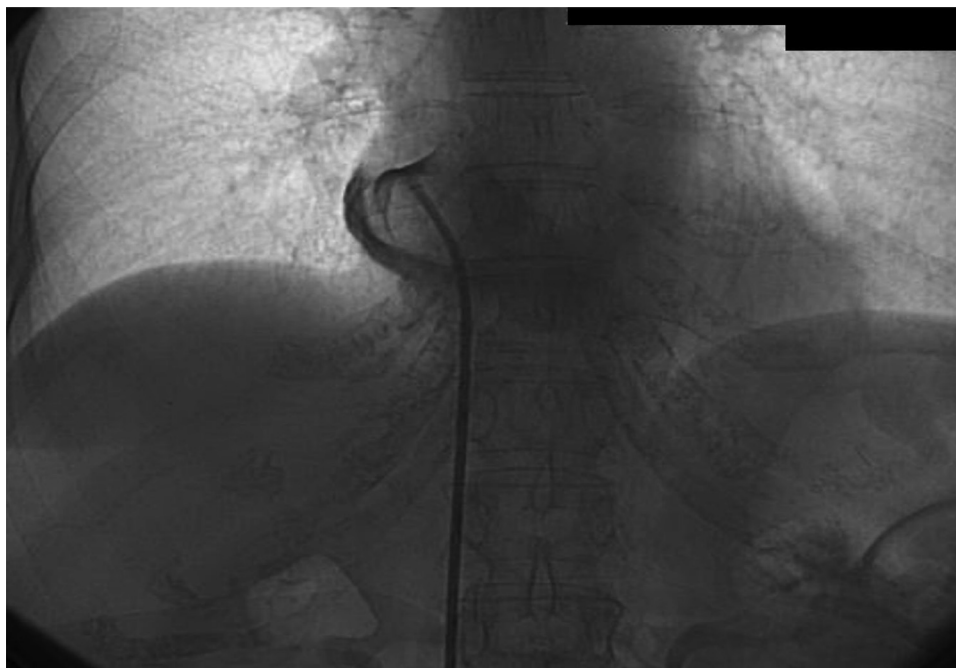


Fig. 5 – Post-procedural control; absence of extravasation of contrast material.

frequent lymphoma in the mediastinum, predominantly in the nodular sclerosis subtype [6].

The radiologic presentation of PM-NHL is also variable, ranging from a mediastinal mass with or without superior vena cava syndrome to a pleural or a cardiac mass associated with effusion or as an effusion only. The diagnosis of PM-NHL can only be established by microscopic evaluation [6].

Considering our case, its uniqueness and rarity lies in the unusual fact that the major component of the lesion was within the atrium, with little extravascular involvement, making the traditional percutaneous methods possibly inadequate.

In our case, due to the massive infiltration into the right atrium, the endovascular approach was proven reasonably effective for the diagnosis of NHL.

Another important aspect to take into consideration is the consistence of the lesion; in fact, the surface of the mass was fibrous and hard.

Certainly, a classic aspiration biopsy is less invasive and easier to perform but it carries a higher risk of obtaining a non-diagnostic sample. In our case, the best choice was to use biopsy forceps, classically used for biliary tracts, due to their robustness and possibility of abrading the tumor even if it is hard.

Although this procedure carried the risk of missing the lesion (eg, collecting thrombotic material), in our patient, the procedure was performed successfully, and the material collected was found to be diagnostic.

In this case, the mass almost completely occupied the right atrium, and this facilitated the biopsy sampling; however, the effectiveness of the approach should be discussed on a case-by-case basis, especially as regards the location and size of the cardiac mass.

In our patient, cardiac forceps biopsy occurred without complications and the endovascular approach proved to be a valid and alternative method for the diagnosis of intracardiac NHL.

Patient consent

Informed written consent was obtained from the patient for publication of the case report and all imaging studies. Consent form on record.

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