European Journal of Radiology 84 (2015) 489–498

Contents lists available at ScienceDirect

European Journal of Radiology

journal homepage: www.elsevier.com/locate/ejrad

Review

Hypervascular mediastinal masses: Action points for radiologists

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ARTICLE INFO

Article history:
Received 1 August 2014
Received in revised form
26 November 2014
Accepted 30 November 2014

Keywords:
Hypervascular mediastinal masses
CT
MR
Functional imaging

ABSTRACT

Hypervascular mediastinal masses are a distinct group of rare diseases that include a subset of benign and malignant entities. Characteristic features and known association with syndromes and genetic mutations assist in achieving a diagnosis. Imaging allows an understanding of the vascularity of the lesion and should alert the radiologist and clinician to potential hemorrhagic complications and avoid percutaneous CT guided biopsies. In such cases, pre-procedure embolization and surgical biopsy maybe considered for better control of post procedure hemorrhage. The purpose of this article is to describe and illustrate the clinical features and radiologic spectrum of hypervascular mediastinal masses, and discuss the associated clinical and genetic syndromes. We will present an imaging algorithm to determine further evaluation and subsequently guide treatment.

1. Introduction (Tables 1 and 2)

Hypervascular mediastinal masses are characterized by intense enhancement after contrast administration often with recruitment of adjacent abnormally enlarged feeding vessels. Hypervascular mediastinal masses are a distinct group of rare diseases. Pathological entities include Castleman disease, paraganglioma, carcinoid, ectopic parathyroid adenoma, vascular malformations, and hypervascular nodal metastatic disease. Accurate localization and characterization of hypervascular masses is important to narrow the differential diagnosis and avoid catastrophic procedure related hemorrhage. The objective of this article is to describe and illustrate the clinical and radiologic features of the hypervascular mediastinal masses, and to discuss the associated clinical and genetic syndromes. We will present an algorithm combining all data to determine further evaluation and subsequently guide treatment.

2. Optimizing imaging for detection of hypervascular masses and proposed diagnostic algorithm (Table 2)

The majority of hypervascular masses are identified as a radiographic abnormality. Alternatively, patients with neuroendocrine tumors may present with clinical symptoms such as flushing, and labile hypertension due to secretion of hormones and other vasoactive products. A third scenario is one in which anatomical localization is sought in a patient in whom diagnosis is suspected clinically such as patients with symptoms of parathyroid hormone excess in whom imaging is done for anatomical localization of the parathyroid adenoma.

2.1. Computed tomography (CT)

Non-contrast CT can highlight important imaging features such as calcifications or cystic change, and may detect presence of fat or hemorrhage. In select cases these may point towards a specific diagnosis, such as presence of arborizing calcification in Castleman disease (Fig. 1A), and phleboliths in vascular malformations (Fig. 2C) [1,2].

Optimal contrast enhancement is important for highlighting the conspicuity and hypervascularity of a mediastinal mass. This contrast opacification is optimized by using a delay time of 30–35 s for image acquisition which corresponds to arterial phase as most mediastinal masses have systemic arterial supply and hence show optimal enhancement at this phase [3]. If image acquisition is performed too early (contrast present only within the pulmonary arterial system) or too late (redistribution of iodinated contrast and opacification of systemic veins) the avid enhancement associated with these masses may be missed or overlooked on imaging. This is an important imaging pitfall which can be avoided by having specific delay time of 30–35 s or alternatively use bolus tracking with region of interest (ROI) placed in the descending thoracic aorta [3].

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http://dx.doi.org/10.1016/j.ejrad.2014.11.039
0720-048X/

Table 1
Hypervascular mediastinal masses by compartment.

Anterior mediastinum	Middle mediastinum	Posterior mediastinum
Castleman disease	Castleman disease	Castleman disease
Ectopic parathyroid adenoma	Paraganglioma	Paraganglioma
Vascular malformation	Vascular malformation	Vascular Malformation
Metastatic disease	Metastatic disease	Metastatic disease
Thymic carcinoid		Neurogenic tumor

The exact anatomical extent and sources of vascularity of complex multicompartmental masses may be incompletely delineated on axial imaging alone. The multiplanar capability of multidetector computed tomography (MDCT) is extremely helpful in delineating the full extent and source(s) of vascularity thus helping in preoperative planning of these patients [4]. Sagittal and coronal reformations allow accurate anatomical localization (Fig. 3B) of these masses within the various mediastinal compartments and also help separate out the mediastinal masses from the intrapericardial masses. Accurate localization is key when planning percutaneous biopsies or procedures of "*low anterior mediastinal masses*" which maybe truly within the pericardium. Biopsy of these masses can be associated with potentially life threatening hemorrhage leading to hemopericardium.

2.2. Magnetic resonance imaging (MRI)

In select cases, chest or cardiac MR may add value due to its superior contrast resolution and the ability to delineate intravascular and intra-cardiac extension. On non-contrast T1-weighted images, hypervascular masses such as paragangliomas may demonstrate a "salt and pepper" appearance where punctate regions of hyperintensity ("salt") are seen along with small flow voids ("pepper") leading to a speckled appearance of tissue (Fig. 3C) [5].

Additionally, use of automated post contrast MR subtraction sequences is extremely useful in distinguishing proteinaceous or hemorrhagic T1 hyperintense lesions from masses with true enhancement on post contrast images [6].

Hypervascular masses may appear to be inseparable from adjacent large mediastinal vessels (Fig. 3B) and cardiac chambers and thus present a special challenge for the radiologist and the surgeon as it is frequently difficult to delineate vascular and cardiac extension on contrast-enhanced chest CT. Incorporation of EKG-gated black-blood double inversion recovery (DIR) fast spin-echo cardiac-MR sequences null signal from flowing blood and are best suited for determining the extent of invasion of adjacent mediastinal vascular structures and, thus, resectability (Fig. 3C) [7].

2.3. Functional imaging

Functional imaging techniques are especially helpful in patients with history of symptoms related to secretion of vasoactive substances. Scintigraphy with I-123 or I-131 metaiodobenzylguanidine (MIBG) or Octreotide is most frequently used in patients with suspected paraganglioma. This is helpful in localization and staging of tumors and are particularly recommended for evaluation of patients with genetic predisposition to familial paraganglioma such as mutation carriers of succinate dehydrogenase enzyme B (SDHB) due to high risk of multifocality. MIBG is a norepinephrine analog taken up by some neuroendocrine tumors as it is incorporated into neurosecretory vesicles [8] while Octreotide is a somatostatin analog that binds to the somatostatin receptors expressed by some neuroendocrine tumors. Octreotide scintigraphy is also used in patients with suspected primary or metastatic carcinoid and has a high detection rate

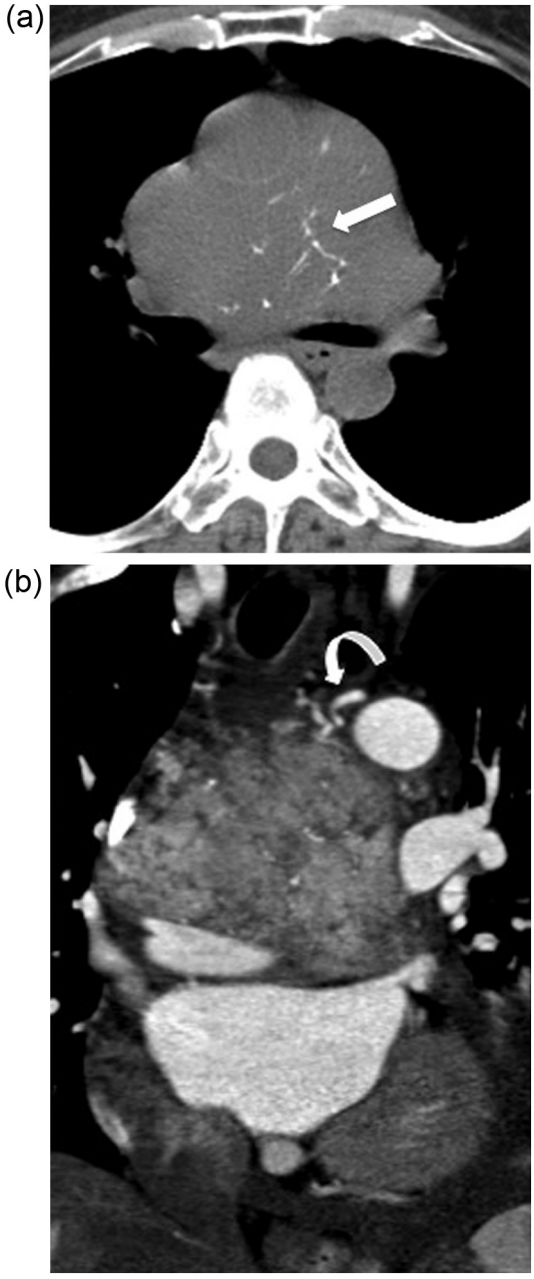

Fig. 1. 59-year-old woman with unicentric Castleman disease, hyaline subtype. (A) Unenhanced axial CT scan shows large middle mediastinal mass with central arborizing calcification (arrow). (B) Coronal contrast-enhanced CT image demonstrates intense enhancement within the mass which appears to compress and displace the pulmonary arteries and abuts the aortic arch. Multiple feeding vessels are noted in its periphery (curved arrow) due to the significant vascularity. Video-assisted thoracoscopic surgery (VATS) biopsy was performed, resection was not feasible due to the extreme vascularity.

of up to 90% [9]. In patients with symptoms of hyperparathyroidism and an enhancing mass in the mediastinum, further evaluation maybe performed with Technetium (99mTc) Sestamibi scan and a diagnosis of parathyroid adenoma can be confirmed (Fig. 4).

PET CT has a role in staging of malignant hypervascular tumors such as paragangliomas and Castleman disease which may be associated with other lymphoproliferative disorders such as lymphoma. However, PET CT is unable to distinguish between various types of hypervascular mediastinal tumors or distinguish malignant from benign entities in absence of distant metastases [1].