CASE REPORTS

NOT ALWAYS A THYMOMA – ABOUT A MEDIASTINAL CAVERNOUS HEMANGIOMA

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Abstract

A mediastinal cavernous hemangioma is difficult to distinguish from other types of mediastinal tumours.

They are usually asymptomatic and incidentally discovered in an imaging study but can present with compressive symptoms or by infiltration of adjacent structures.

A 64-year-old woman with a prior history of triple negative invasive carcinoma of the breast, under surveillance was referred after a Chest CT-scan showed a soft tissue 40x20 mm mediastinal mass, suggestive of a thymoma, and as such no tissue biopsy was obtained.

A right-side uniportal VATS was performed, the anterior mediastinum dissected and the mass was exposed, and several anomalous veins were identified.

Histopathology showed 36x31x15 mm mass, compatible with a cavernous hemangioma of the anterior mediastinum.

This case, whilst not questioning the NCCN statement suggesting not doing a tissue biopsy, points to the fact that rare differential diagnosis, like a Cavernous Hemangioma do exist, and a careful and sound judgement is needed at all times.

Keywords: *Cavernous hemangioma, Anterior mediastinum, Anomalous vein*

INTRODUCTION

The International Society for the Study of Vascular Anomalies (ISSVA) classification of vascular anomalies divides vascular anomalies into vascular tumours and vascular malformations. Cavernous hemangiomas are now considered venous malformation, a type of vascular malformations.[1]

It is considered to be the result of local abnormal morphogenesis of vascular endothelial cells, occurring before birth.[2]

Benign vascular tumours of the mediastinum were first reported in 1914 by Shenan.[3]

They are rare and largely described in case reports, with fewer than 110 cases reported, comprising only 0.5% of mediastinal masses.[4,3]

They can occur anywhere in the mediastinum, most commonly in the anterior compartment (68 %), followed by the posterior (22 %) and middle compartment (10 %).[5]

The first Cavernous hemangioma (CH) was described by Lungenschmid in 1990.[6]

To the best of the author's knowledge, only ten cases of cavernous hemangioma of the anterior mediastinum have been reported.[7]

Whilst the majority of cavernous hemangiomas occur superficially in the cutaneous and mucosal tissues in children, those of the mediastinum present much later in life with a mean age of 35.[7,4]

They are usually asymptomatic and incidentally discovered in an imaging study, but can present with compressive symptoms due to large size or infiltration of adjacent structures causing dysphagia, chest pain or fullness, dyspnoea, haemoptysis or superior vena cava syndrome.[4 7]

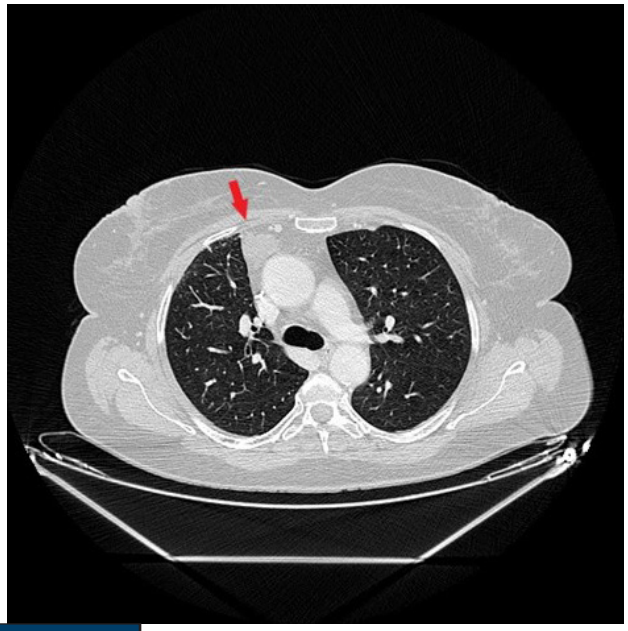

Figure 1 *CT scan Axial View – In the prevascular fat, a 40 x 20 mm mass (red arrow) with tissue density, with calcifications, suggestive of thymoma.*

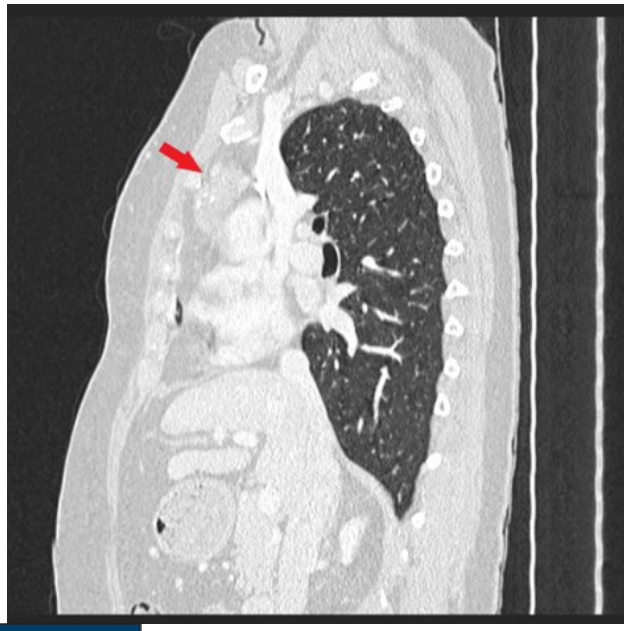

Figure 2 *CT scan Sagittal View – In the prevascular fat, a 40 x 20 mm mass (red arrow) with tissue density, with calcifications, suggestive of thymoma.*

A mediastinal cavernous hemangioma is difficult to distinguish from other types of mediastinal tumours, and differential diagnosis includes numerous malignant and benign tumours, such as thymoma, lymphoma, teratoma, neuroma, angiolipoma, goitre, and cystic lymphangioma.[7]

CASE REPORT

A 64-year-old asymptomatic woman with a prior history of triple negative invasive carcinoma of the breast, under surveillance after surgery and adjuvant chemo and radiotherapy. She was referred to the Centre's Cardiothoracic department after a Chest CT-scan showed a soft tissue 40x20 mm mediastinal mass, suggestive of a thymoma or an enlarged lymph node. (Image 1-2)

An MRI as performed, but showed only an expansive, heterogenous, prevascular lesion of the mediastinum, with no invasion of surrounding structures or vessels. (Image 3)

A right-side uniportal VATS was performed, the anterior mediastinum dissected, and the mass was exposed. Of notice, the right internal thoracic vein followed an anomalous path over the anterior surface of the mass, entering the superior vena cava (SVC) at the level of the azygous vein.

During the dissection another anomalous vein posterior to the mass was avulsed on its entry onto the SVC, leading to a significant bleeding, and conversion to thoracotomy.

The SVC was tangentially clamped, the mass was removed en-block with surrounding thymic fat, and with a better exposure the lesion sutured with a 4/0 prolene.

The remainder of the surgery and postoperative period were uneventful, and the patient was discharged on the 3rd postoperative day.

Histopathology showed a 36x31x15 mm mass with cavernous vascular spaces, overlaid by endothelial cells, with no signs of malignity, compatible with a cavernous hemangioma of the anterior mediastinum. (Image 4)

The patient having made a full recovery remains under surveillance.

DISCUSSION

The diagnosis of a Cavernous Hemangioma can be made by CT scan. It may show multiple venous lakes, calcified phleboliths, complex multiple venous channels, distant feeding veins, and delayed enhancement. Calcified phleboliths are the characteristic manifestation of cavernous hemangioma of the mediastinum, however, are only observed in 10% of the cases.[7]

As in all mediastinal masses detection of thymic malignancy versus thymic cyst or thymic hyperplasia can be better discriminated with chest MRI[8].

Ideally, in vascular lesions a combination of CT and MRI is ideal, as MRI is more sensitive in detecting the components of an MH than CT, whilst contrast CT is more sensitive in detecting phleboliths and shows a greater advantage over MRI in detecting feeding arteries and draining veins.[5]

Large aberrant draining veins are a specific

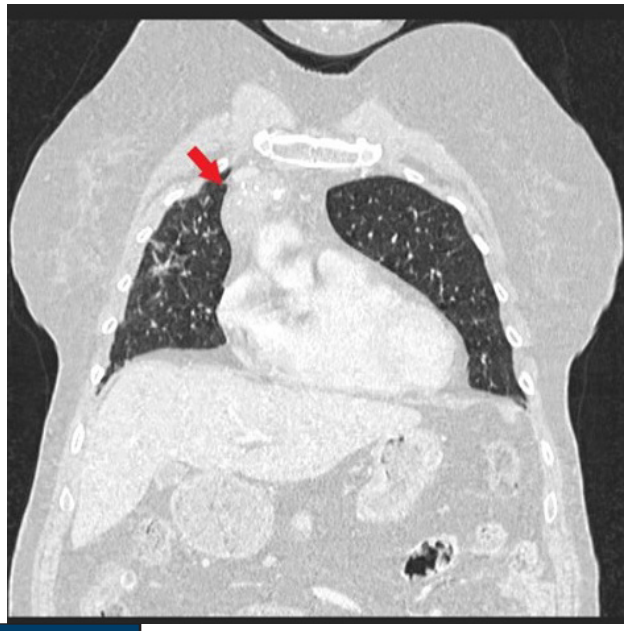

Figure 3 *CT Scan Coronal View – In the prevascular fat, a 40 x 20 mm mass (red arrow) with tissue density, with calcifications, suggestive of thymoma.*

imaging feature and are vital for surgical planning, as they may represent there is or was high blood flow, shunts, or arteriovenous fistulas within lesions. Furthermore, significant draining veins with local dilation or venous aneurysms, and smooth, regular walls of draining veins and feeding arteries within lesions represent no tumoral invasion.[5]

The management of this type of mediastinal mass is controversial. Typically, it involves total resection, or subtotal resection for lesions invading an adjacent great vessel or the heart, both with excellent prognosis. Still the risk of haemorrhage must be considered, and subtotal excision can be an acceptable treatment to avoid substantial blood loss in severe cases.[9,5]

In the presented case, the preoperative exams pointed to a likely to either a thymoma or a lymph node metastasis of the breast and as such a tissue biopsy was not performed.[8]

A retrospective review did not show features suggesting a cavernous hemangioma, nor aberrant vessels to and from the mass, except for some calcifications.

Still, upon identifying the anomalous path of the right internal thoracic over the anterior surface of the mass, the diagnosis should have been raised, and extra care taken.

The anomalous vein posterior to the mass that was avulsed, was most likely a branch draining the Cavernous Hemangioma.

Whilst not questioning the NCCN statement suggesting not doing a tissue biopsy when there is a high likelihood of a thymoma, this case points to the fact that there rare differential diagnosis, like a Cavernous Hemangioma, even in these cases.

Even if there is a preoperative suspicion, performing a biopsy in a vascular malformation, such as this one is risky, but still, can be done.

In conclusion, preoperative study didn't point to a Cavernous Hemangioma, and a biopsy was not warranted, but still a careful and sound judgement and surgical technique are needed at all times.

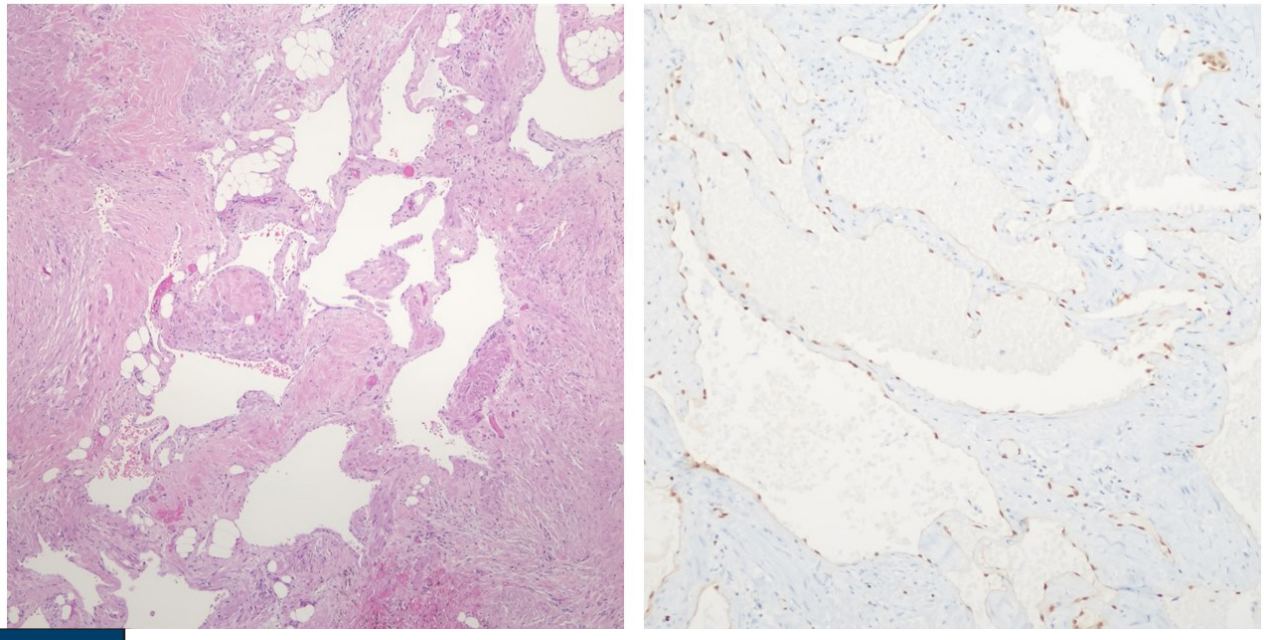

Figure 4 *Circumscribed proliferation of variably sized, dilated and thin walled vessels lined by a single layer of flat endothelial cells (ERG+) without atypia or mitosis.*

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