Posterior mediastinal angiomyxolipoma with spinal canal extension

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RÉSUMÉ

Prérequis : L'angiomyxolipome est une tumeur bénigne considérée comme une variante du lipome et qui se développe principalement dans le tissu sous-cutané. La localisation médiastinale n'a jamais été rapportée.
But : Décrire les caractéristiques en Imagerie de cette tumeur dans sa localisation médiastinale postérieure et de les confronter aux caractéristiques anatomo-pathologiques.
Observation : Nous rapportons le cas d'une femme de 49 ans admise pour douleur thoracique et troubles neurologiques des deux membres inférieurs. La radiographie du thorax a objectivé une opacité mediastinale postérieure. L'examen Tomodensitométrique a montré une masse médiastinale postérieure avec extension intra canalaire contenant une composante graisseuse et tissulaire fortement rehaussée par l'injection intra veineuse de produit de contraste. L'examen anatomopathologique de la masse excisée a posé le diagnostic d'angiomyxolipome médiastinal postérieur.
Conclusion: La localisation médiastinale postérieure de l'angiomyxolipome n'a jamais été rapportée. Elle peut s'accompagner d'une extension intra canalaire. Ses aspects en imagerie suggèrent le diagnostic.

Mots-clés

Médiastinal angiomyxylipome, Tomodensitométrie.

SUMMARY

Background: Angiomyxolipoma is a benign tumor considered as a variant of lipoma and that occurs mainly in the subcutis. The mediastinal location hasn't been previously reported.
Aim: To describe the radiological features of this tumor in its posterior mediastinal location and to confront them to the pathological features.
Case report: We report the case of a 49-year-old woman who was admitted for chest wall pain and neurologic disturbance of her two lower limbs. The chest X-ray showed a posterior mediastinal opacity. On CT examination, this mass contained some small areas of fat and enhanced intensily. Microscopic examination of the excised mass confirmed the diagnosis of posterior mediastinal angiomyxolipoma.
Conclusion: Mediastinal location of angiomyxolipoma hasn't been previously reported. Clinicians and radiologists should be aware that this diagnosis should be suggested, among others, when there is a posterior mediastinal mass that contains fat and that intensely enhances with a possible spinal cord extension.

Key-words

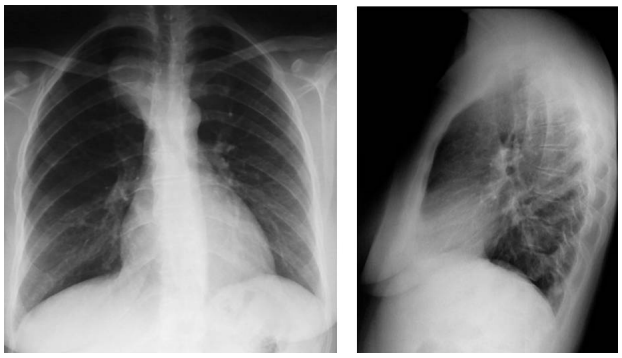
Mediastinal angiomyxolipoma, computed tomography.

There are multiple microscopic types of lipoma. Angiomyxolipoma is considered as one of the rarest of them which mainly occurs in the subcutis. The mediastinal location has never been previously reported.
The aim of this article is to describe the radiological features of this tumor in its posterior mediastinal location and to confront them to the pathological features.

OBSERVATION

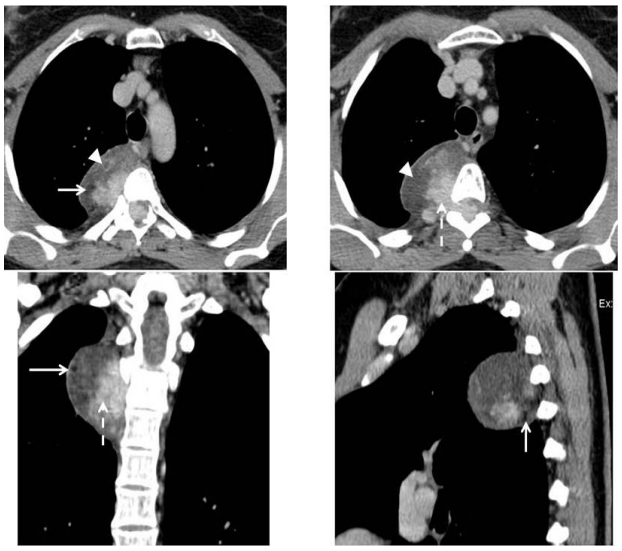
A 49-year-old woman was admitted in our hospital for investigation of an ill-defined right-sided posterior chest wall pain which had been evolving for a few weeks. She had never smoked, had no history of a previous infection, no cough, no hemoptysis and no dyspnea but she noticed a progressive weakness and numbness of her two lower limbs. The examination of her chest didn't show any mass or cutaneous abnormality. Her electrocardiogram was normal as were all the hematologic and biologic parameters.
The chest radiograph at admission identified a homogeneous right opacity with smooth borders making an obtuse angle with mediastinum in its lower part reflecting its mediastinal origin. This opacity extended above the clavicle on the postero-anterior film. The central mediastinum structures were in place. There were no lung parenchyma lesions. No osseous anomalies were found (figure 1).

Figure 1 : Posteranterior and profile chest plain radiographs showing a well-limited and homogeneous right posterior mediastinal mass. There is no parenchymal lesion.



A subsequent enhanced chest and abdominal computed tomography has been performed. It showed the presence of a heterogeneous right posterior mediastinal mass located in the costovertebral gutter with a vascular blush and small fat areas. The mass extended into the vertebral canal through an intervertebral foramen and squeezed the thoracic spinal cord (figure 2). No further thoracic or extra-thoracic lesions were detected. We couldn't perform MRI exploration as the patient was claustrophobic. We didn't perform a trans-thoracic biopsy of the mass because of its large vascular component shown on CT examination. The mediastinal mass was surgically removed.

Figure 2 : Enhanced computed tomography showing a right posterior mediastinal mass. The mass is heterogeneous. It contains small fat areas (arrows), low density areas (arrowheads) and areas with intense enhancement (dashed arrows). This latter component extends in the vertebral canal through an inter vertebral foramen.



Pathologic evaluation demonstrated a well-circumscribed mass, with yellowish and gelatinous cut surface (figure 3). Microscopic examination showed that it was composed of abundant myxoid stroma, blood vessels and mature adipocytes with lobulated architecture confirming the mediastinal angiomyxolipoma diagnosis (figure 4).

Figure 3 : The excised mass is well-circumscribed, with yellowish and gelatinous cut surface.

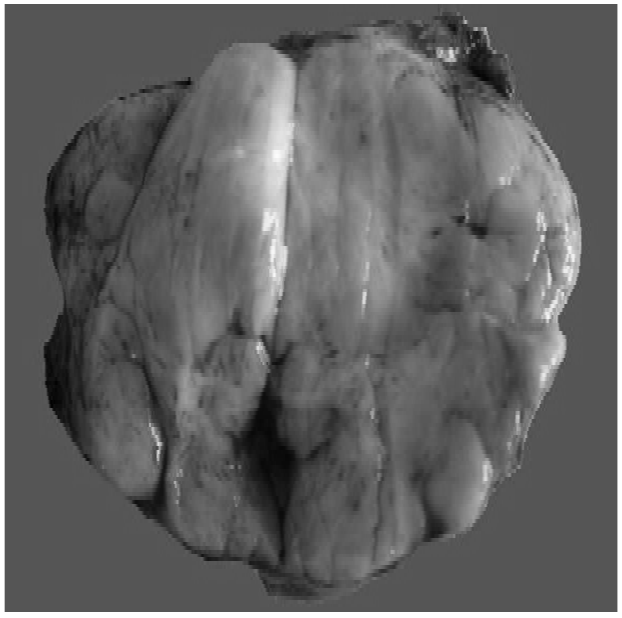
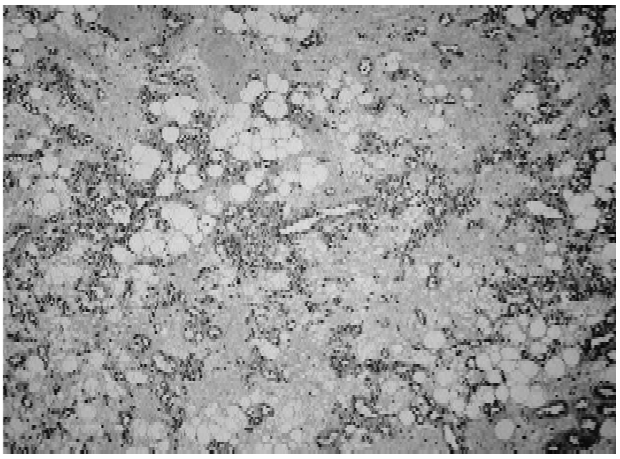


Figure 4 : Photomicrograph showing myxoid stroma, blood vessels and mature adipocytes (Hematoxylin Eosin: original x 20)



DISCUSSION

Angiomyxolipoma is an extremely rare benign tumor. It is recognized as a variant of lipoma [1]. Twelve cases have been reported in the literature up to now [2-4]. This tumor occurs mainly in the subcutis [1, 5, 6]. Other locations have been reported such as the spermatic cord [7], the subungual area and the knee [3, 4]. Mediastinal angiomyxolipoma hasn't been previously reported. Only some cases of mediastinal angiolipoma are found in the literature.

The mediastinal angiomyxolipoma we are reporting is diagnosed after the occurrence of symptoms related to spine cord compression.

Angiomyxolipoma has no specific radiological signs on plain chest radiography. On CT, the tumor is heterogeneous. The vascular component is predominant and shows an intense enhancement. This feature can suggest the diagnosis of angioma. The fatty component appears as small spots with low density. Multiplanar reconstructions of the data of our patient showed extension to the vertebral canal through the fourth right intervertebral foramen without osseous destruction. This extension explained the sensitive disturbance and the lower limbs failure that occurred in our patient. It can also be seen in peripheral nerves tumors such as schwannoma and neurofibroma [1, 7]. The analysis of the pulmonary parenchyma didn't show any anomaly.

When practiced, MRI shows that the fatty component has a high signal on T1-weighted images that is attenuated on sequences with fat signal saturation. It allows a better evaluation of the extent into the spinal canal and shows signs of spinal cord injury. There are few tumors of the posterior mediastinum containing fat component. They include teratomas, myxoïd liposarcoma and liposarcoma [1, 5]. Angiolipoma and angiomyolipoma could even be discussed in case of an intense enhancement.

The excised specimen was yellow, well limited with a 6,5cm maximal diameter. On microscopic examination, the tumor showed typical features of an angiomyxolipoma with lobulated architecture, myxoïd stroma, mature adipocytes and blood vessels.

Angiomyxolipoma is considered to be a rare variant of lipoma characterized by an admixture of mature adipose tissue, paucicellular myxoid stroma, and an abundance of thin- and thick-walled blood vessels [8]. Immunohistochemical studies show that the myxoïd component expresses vimentin and CD34. The blood vessels are positive for vimentin and SMA whereas adipocytes are positive for S-100 protein [1].

CONCLUSION

Mediastinal location of angiomyxolipoma hasn't been previously reported. Clinicians and radiologists should be aware that this diagnosis should be suggested, among others, when there is a posterior mediastinal mass that contains fat and that intensely enhances with a possible spinal cord extension.

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