Extra-adrenal mediastinal myelolipoma: A rare cause of posterior mediastinal mass

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Abstract

We report a rare cause of a posterior mediastinal lesion, an Extra Adrenal Myelolipoma (EAM). Myelolipoma commonly occurs in the adrenal gland, is composed of both adipose tissue and normal haematopoietic elements and can occur outside the adrenal gland. A 59-year-old gentleman with palpitations and shortness of breath was found to have a paravertebral lesion with no intraspinal communication. Subsequent surgical removal revealed it to be an EAM. Thoracic EAMs should be considered during the evaluation of a posterior mediastinal mass.

INTRODUCTION

The most common tumours of the posterior mediastinum in adults are metastatic neoplasms, neurogenic tumours, other soft tissue tumours and cystic tumours. We report a rare case of a posterior mediastinal Extra Adrenal Myelolipoma (EAM) in a 59 year-old gentleman.

CASE REPORT

A 59 year-old asthmatic gentleman with a 40-pack year history of smoking presented with palpitations and shortness of breath to his General Practitioner. He was referred to his local hospital for a cardiology opinion. Clinical examination revealed a normal cardiovascular system, a central trachea, clear respiratory tract and a soft non-tender abdomen. Laboratory blood results revealed an elevated haemoglobin at 18.2 g/dl, but no other abnormalities. The Electrocardiogram showed normal sinus rhythm. The plain film radiograph revealed a low posterior mediastinal mass behind the cardiac shadow. Computed tomography (CT) and magnetic resonance imaging (MRI) were performed confirming the presence of a posterior mediastinal mass in the left para-vertebral gutter with no evidence of intraspinal communication [Figures 1 and 2].
The patient was referred to us at the regional thoracic surgical unit where he was counselled and consented for a left thoracotomy and excision biopsy of the mass.

Under general anaesthesia and single lung ventilation with a double lumen endotracheal tube in situ the patient underwent a left thoracotomy. A soft spherical red-bluish vascular mass measuring 7 cm by 6 cm by 4cm was identified in the left para-vertebral gutter with no intra-spinal connection. The lesion was enucleated after gaining control of its vascular supply and sent for histological examination. A single 28FG drain was placed in the pleural cavity and the wound was closed in layers. The patient was nursed in the thoracic high dependency unit with a thoracic epidural for analgesia. His post-operative course was uneventful except an increased serosanguinous chest drainage in the first four days. Biochemical analysis of the fluid ruled out injury to the thoracic duct. The chest drain was removed when the chest radiograph was satisfactory and he was discharged on the fifth post-operative day.

The histo-pathological examination of the lesion was composed of mature adipose tissue admixed with a central cellular area filled with haematopoietic elements including megakaryocytes, accompanied by myeloid and erythroid precursors confirming the presence of myelolipoma [Figure 3].

**DISCUSSION**

Myelolipoma are rare, benign, non-functioning and slow growing lesions arising from reticulum cell metaplasia, which are composed of mature adipose tissue with a variable amount of haematopoietic elements. The incidence is between 0.03 – 0.8%, the majority are found at post mortem. The most common location is the adrenal gland, occurring equally in both sexes on average at 55 years of age. Adrenal myelolipomas are thought to constitute sites of extra medullary haematopoesis. Infection, necrosis and stress promote metaplasia of the reticuloendothelial cells of blood capillaries in the adrenal gland. 15% of myelolipomas exist extra-adrenally and have been reported in the posterior mediastinum, liver, gastric antrum, pre-sacral tissue, peri-renal tissue and the retroperitoneum. Myelolipoma were originally described by Gierke in 1905, and the term, myelolipoma, was first used by Oberling in 1929.

Mediastinal lesions may present with chest pain, shortness of breath or a cough. Myelolipoma can spontaneously haemorrhage, particularly if they are large or vascular in
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nature. Indications for excision include rapidly expanding EAM’s or equivocal lesions that need to be evaluated histologically.

Diagnosis of a myelolipoma by plain film radiography can prove difficult, those which are particularly large or those with a high adipose content may give a suggestive appearance of the lesion. CT demonstrates areas of fatty tissue interspersed with areas of higher-attenuation. Images are dependant on the histological composition and the areas of higher attenuation represent myeloid tissue.

MRI of adrenal myelolipomas characteristically demonstrates a bright signal on TI-weighted and T2-weighted sequences consistent with the presence of fat. The lesion enhances brightly after intravenous administration of gadolinium. The use of fat suppression images will result in a decrease in the signal intensity and help confirm the presence of a myelolipoma. Calcification can be detected in up to 24% of lesions.

Extramedullary haematopoietic lesions probably pose the most difficult differential diagnosis. They are radiologically and histologically very similar. A tumour with its characteristic histological features, without any haematological abnormalities will confirm the diagnosis of EAM. Imaging is helpful, but definitive diagnosis requires cytologic or histologic sampling in conjunction with the appearance of a normal blood film.

This gentleman underwent an excision biopsy of this lesion, which was originally thought to be neurogenic in origin. He will continue to be under surveillance as a similar lesion has been noted in the right hemithorax. This is in keeping with the literature on mediastinal EAM and their frequent bilateral appearance.

Thoracic EAMs should be considered during the evaluation of a posterior mediastinal mass.

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