Myelolipoma in the paravesical space - case report

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Summary

Myelolipomas are benign tumours composed of haematopoietic tissue and mature adipocytes. They occur predominantly in the adrenal glands although rarely extra adrenal sites have been described. Their presentation and location can cause them to be easily confused with a malignant tumour. Here we present a myelolipoma in the paravesical space. To the best of our knowledge this location has not been reported before. The growing use of imaging will lead to an increase in incidence of these tumours. Understanding the natural history and their differentiation from other tumours is therefore important. The purpose of the present report is to raise awareness for this unusual tumour type in this location.

Key words: Myelolipoma; Cancer; Diagnosis; Management; Location.

Introduction

Myelolipomas were first described as distinct benign entities in 1929 [1]. Generally they are asymptomatic and present as solitary well described nodules. Microscopically they consist of fat, myeloid, and erythroid marrow elements [2-4]. Sometimes internal areas of haemorrhage and subsequent calcification are seen. Except for the extra-medullary location, the histopathology resembles mature bone marrow, with differing proportions of haematopoietic lineage [5]. However the typically reticular sinusoids or bone spicules are not present [6]. The incidence in autopsy series varies between 0.08% and 0.2% [6, 7]. The most common location is the adrenal glands. Less frequently extra adrenal locations are seen such as in the liver, stomach, mesentery, spleen, mediastinum, presacrum, leptomeninges, and pleural cavity [2-9]. Due to their presentation they can form a diagnostic challenge in differentiation from a malignant tumour. The present report describes for the first time localisation of a myelolipoma in the paravesical area.

Case Report

A 43-year-old woman with a complex medical history came under our care. At the age of 20 she underwent a kidney transplant. Shortly afterwards septic bone necrosis of both hips and the left shoulder followed. Due to her cortical steroid intake she developed osteoporosis at age 28. The latter was managed by vitamin D, calcium supplements and a reduction of the corticosteroid therapy. When she reached 32 she developed a secondary hyperparathyroidism, which was treated by increased vitamin D intake. A few months later she developed a vitamin B12 deficiency. At the age of 34 she was anaemic and treatment with erythropoietin was started. At the age of 35 she had regular recurrent skin mycosis which was treated locally with terbinafine cream. At the age of 36 she started to suffer from migraines. Initial treatment with clonidine gave no response, but treatment with oxetoron fumarate did (3 tablets daily). However after eight months there was a slight reduction in kidney function and a tendency to hypotension (HT), therefore the dose was reduced to two tablets. Due to severe metrorrhagia she had curettage at age 36. The pathology report revealed a benign endocervial polyp and a hormonal intrauterine device (R/mirena) was installed, which led to the expected amenorrhea. When she reached the age of 38 diabetes mellitus was diagnosed. The diabetes was likely induced by corticosteroids. Initially diabetic measurements were sufficient, however later on insulin was needed to control HT glycaemia. She developed a squamous cancer of the right middle finger at age 42.

Over time she had multiple examinations which revealed a slowly growing retroperitoneal mass at the left pelvic side wall. Initial computed tomography (CT) suggested a haematoma. On the most recent computerised tomography, age 43, the mass had reached a size of 9 cm and had a pushing effect on the bladder. Imaging suggested a partly solid and cystic mass. On clinical examination it was a non mobile mass stuck against the pelvic sidewall. CA125 at that time was 15.7 (= normal). Due to the growth and the size explorative laparotomy was performed and the retroperitoneal mass was excised. The lesion was located mainly in the paravesical fossa, with a small part hanging over the uterine artery in the pararectal fossa. Macroscopically it was a well defined mass of 9 x 8 cm and on section the colour was dark red to brown. Frozen section was performed and revealed that it was a myelolipoma; no further surgery was done. The patient made an uneventful recovery and could be discharged after five days.

Discussion

Myelolipomas are typically found in middle and in older age, with a slight female predominance [3, 6]. They have a benign clinical course, however large tumours carry a risk of spontaneous rupture and bleeding [5].

The aetiology is unknown and no satisfactory explanation can been given for the presence of haematopoietic elements in these lipomatous tumours. It has been postulated that these lesions develop from embryonic mesenchymal rests in the adrenal gland, bone marrow tissue

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embolism with subsequent hypertrophic, stress-induced reticuloendothelial cell metaplasia, and proliferation of embryonic rests of haematopoietic stem cells or autonomous growth [3, 4, 6, 8].

Others have suggested a possible association with certain endocrine or hormonal disorders. Conditions like Conn's syndrome, Cushing's disease, congenital adrenal hyperplasia, and conditions associated with increased cortisol production, such as diabetes, hypertension, and obesity, as well as exogenous steroid use have been described in the literature [3, 4, 6, 8]. The hypothesis of chronic corticosteroid use fits the present case.

The differential diagnosis of retroperitoneal tumours should include benign lipomatous lesions, angiomyolipoma, extramedullary haematopoiesis in soft tissues, carcinoma and more common tumours of soft tissue or lymphoid origin like sarcomas and lymphomas [5].

Myelipomas should be differentiated from angiomyolipomas. Angiomyolipomas contain vascular and leiomyomatous elements in addition to adipose tissue [5]. Extramedullary haematopoiesis in soft tissues is usually a response to a primary pathology, such as myeloproliferative disorders or haemolytic anaemia. They differ from myelolipoma by the involvement of other haematopoiesis organs like the liver and spleen. Furthermore there are no lipomatous elements or encapsulation [5].

Imaging studies like CT or magnetic resonance imaging (MRI) may give characteristic findings of a completely or partially encapsulated fatty mass, with typically low density. Additional image-guided final needle aspiration cytology can be of help in further differentiating between a benign lipomatous lesion or an other retroperitoneal tumours.

Myelolipomas are benign lesions, which can be managed conservatively. Only in case of doubt or when there is a risk of rupture (lesions > 4 cm) and haemorrhage or when they start to grow and compress on adjacent structures should they be removed [2]. A simple resection is sufficient. No recurrence or malignant degeneration has been reported so far.

Awareness of this tumour is important thus the growing use of imaging (MRI and CT scanning), will probably lead to an increase in the diagnosis of these tumours.

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