G Chir Vol. 33 - n. 3 - pp. 62-65
March 2012

clinical practice

Extra-adrenal perirenal myelolipoma. A case report and review of literature

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SUMMARY: **Extra-adrenal perirenal myelolipoma. A case report and review of literature.**

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Myelolipomas are rare tumours which are most commonly found in association with the adrenal glands. However, extra-adrenal sites have been described, but limited to case reports. They are characterized by a normal adrenal gland function and absence of haematopoesis which differentiates them from extramedullary haematopoetic tumours.

We present a rare case of perirenal extra-adrenal myelolipoma and we review the imaging characteristics and management options for this condition.

RIASSUNTO: **Mielolipoma perirenale extra-surrenalico. Caso clinico e revisione della letteratura.**

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I mielolipomi sono tumori rari di solito surrenalici. Sono descritti in letteratura pochi casi a localizzazione extra-surrenalica. A differenza dei tumori emopoietici extramidollari, i mielolipomi sono caratterizzati da una normale funzionalità ghiandolare e dall'assenza appunto di attività ematopoietica.

Riportiamo un raro caso di mielolipoma perirenale extra-surrenalico, analizzandone l'imaging caratteristico e le opzioni terapeutiche.

KEY WORDS: Extra-adrenal myelolipoma - Histology - Clinical presentation.
Mielolipoma extra-surrenalico - Istologia - Clinica.

Introduction

Myelolipomas are rare tumours most commonly found in association with the adrenal glands (1). However, extra-adrenal sites have been described, but limited to case reports (2). These lesions are histologically characterized by myeloid and erythroid cellular precursors interspersed with mature adipose elements. They are further characterized by a normal adrenal gland function and the absence of haematopoesis which differentiates them from extramedullary haematopoetic tumours.

We present a rare case of perirenal extra-adrenal myelolipoma and we review the imaging characteristics and management options for this condition.

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Case report

A sixty-three year old male presented to his general practitioner for a routine physical examination. The only known medical problem was a low testosterone levels for which he was taking supplements.

There was no history of weight loss, early satiety, changes in appetite or bowel habit or abdominal pain. He was also asymptomatic for haematological disorders. His social history was significant for consumption of 30-60 ml of alcohol per day.

Examination revealed a well-looking patient with pink mucous membranes and no lymphadenopathy. The general practitioner did however palpate a vague mass in the abdominal right upper quadrant. The abdominal examination was otherwise normal; liver functions, renal functions, PT/PTT, INR, lipid profile, and haemoglobin were all normal.

A CT scan revealed a 6 × 5.5 cm retroperitoneal mass between the right renal vein superiorly, the inferior vena cava medially and the ureter and kidney laterally. The mass extended retrocaval causing anterior displacement but no compression. There was no identified communication with bone (Fig. 1).

The radiologist's impression was a retroperitoneal sarcoma or lymphoma. On counselling, the patient was given the choice between a 'CT guided biopsy' and 'open surgery'. He opted for the latter.

After induction of general anaesthesia, a right ureteric stent was placed. The patient was positioned supine with a sand bag under the

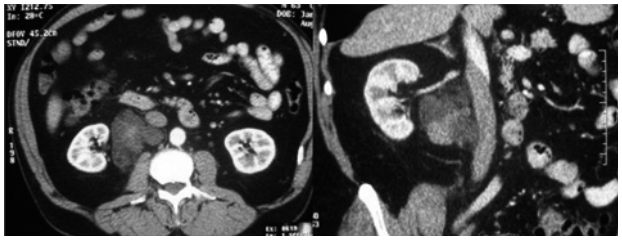

Fig. 1 - CT. Retroperitoneal mass inferior to right renal vein and displacing the vena cava. The ureter is displaced laterally (arrow).

right flank to provide elevation and extension to improve intra operative exposure.

The abdomen was accessed through an extended right **Kocher's incision**. The hepatic flexure was mobilized to expose the duodenum which was subsequently "Kocherized", exposing the inferior vena cava. The dissection was extended proximally to the right renal vein, then medially, dissecting the mass off the inferior vena cava and renal vein. Using the stent for guidance the lateral dissection was then performed. Excision of the mass with intact capsule was accomplished (Fig. 2). The patient's recovery was uneventful and he was discharged on the third post-operative day.

Low power microscopy revealed a well circumscribed mass with a thin fibrous capsule (Fig. 3a). Higher power (×40) showed a mixture of mature lipocytes and marrow elements (Fig. 3b). On magnification (×100) megakaryocytes and other marrow elements were noted. These findings were conclusive for myelolipoma.

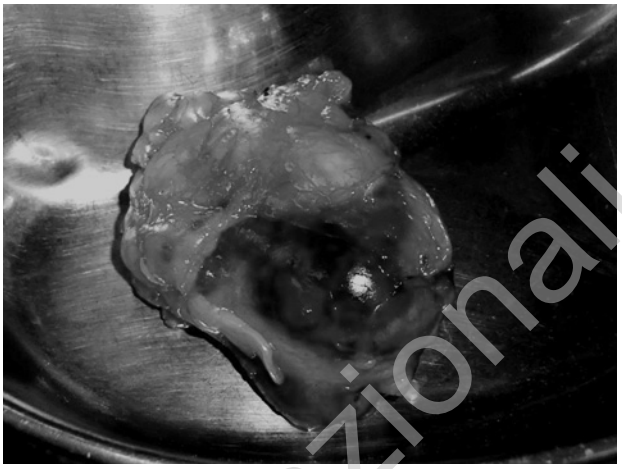

Fig. 2 - Excised mass with intact capsule.

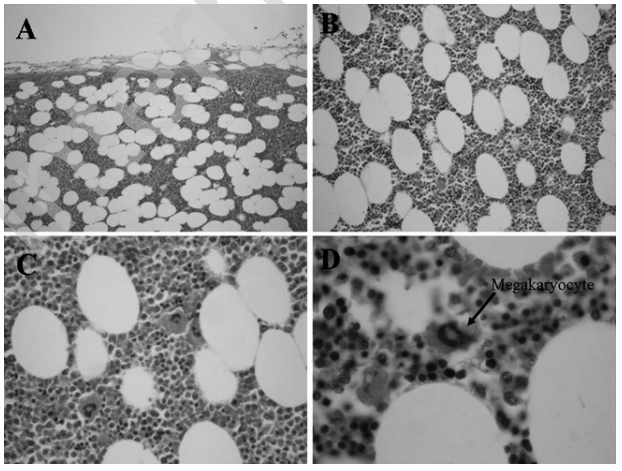

Fig. 3 - Histology. A: low magnification showing well circumscribed mass with loose fibrous capsule. B: higher magnification showing an equivalent mixture. C: ×40 magnification showing lipocytes and mature trilinage haematopoetic cells. D: ×100 magnification, normal megakaryocytes.

Discussion

The histological description of an adrenal mass composed of fat mixed with myeloid and erythroid cells is accredited to Gierke in 1905. Later, in 1929, Oberling coined the term myelolipoma to describe this clinical entity (3). Since the initial description of these rare tumours, case reports have been published of both adrenal and extra-adrenal locations. Olsson reported less than 1% incidence of myelolipomas in an autopsy series in 1973 (4). Of the extra-adrenal locations of the tumours, the presacral region represents 50% (5). Other sites include the stomach, liver, lymph nodes, mediastinum, and cranium (6-9). We present a rare case of perirenal extra-adrenal myelolipoma. Thus far, there have been few reported similar cases including the report of bilateral extra-adrenal myelolipomas, both completely enveloping the kidney by Kumar et al (10). A similar unilateral lesion was described by Brietta and Watkins in 1994 (11).

Due to its rarity the pathogenesis of these tumors has not been adequately studied and several theories have been proposed. Metaplasia of mesenchymal adrenal cells in the bud stage of embryonic development is one suggested mechanism (12). The similarity of lymphocytes in the adrenal cortex to haematopoetic stem cells has also generated the theory that these lesions are the result of adrenocortical remains (5).

Peritoneal foci of extramedullary haematopoiesis are a normal part of embryonic development; these foci can become haematopoietic in certain pathologic situations such as sepsis, severe anaemia, myeloproliferative diseases and skeletal disorders. A mechanism akin to that of extramedullary haematopoiesis has been suggested as the embryonic origin of the myelolipoma (13).

Like its pathogenesis, the natural history of myelolipomas is not well defined. They occur predominantly in females (2:1) and in middle aged to elderly patients (mean age 60 years). Lesions also vary in size from 2 to 26 cm (2). Large tumours can present with a mass effect resulting in abdominal pain, weight loss, gastrointestinal complaints, back pain, a palpable abdominal mass and retroperitoneal haemorrhage. The most common presentation is however, as in our patient, an asymptomatic incidental finding (8).

On gross examination these tumours display features of a lipoma but those in which the myeloid component predominates assume a greyish-red appearance. The cut surface has a variegated appearance consisting of clearly separated fatty and marrow-like areas (14). Microscopic examination reveals a mixture of bone marrow elements and mature lipocytes in varying proportions. The marrow elements consist of a variable mixture of myeloid and erythroid cells, megakaryocytes and occasionaly lymphocytes (15). Cytogenetic analysis of one case revealed a t (3;21) (q25;p11) translocation suggesting a neoplastic process. Further support for this mechanism of pathogenesis is lent by the presence of non-random X-chromosome inactivation displayed by most myelolipomas (16).

The finding of a retroperitoneal mass on imaging carries multiple differential diagnoses. Of these differentials, benign lesions account for only 13.8% (8). This highlights the importance of a thorough evaluation to rule out malignancy.

Imaging characteristics unique to myelolipomas may potentially allow a conservative approach to management. In a series of 67 patients CT showed large amounts of fat with interspersed "smoky" areas having attenuation values of 20-30 HU (17). This density reflects the admixture of fat and marrow-like elements. These imaging findings do not distinguish extra-adrenal lesions from other fat containing retroperitoneal tumours but a percutaneous needle biopsy showing a mixture of marrow-like elements and fat is diagnostic and may negate the need for surgery (14). Our patient's CT did show attenuation values which varied from 70 (fat) to 30-40 (soft tissue) confirming an admixture consistent with a myelolipomas. A conservative approach with CT-guided biopsy could therefore have been used considering the patient was asymptomatic. Tumours such as liposarcomas and myolipomas represent the group of lesions resembling myelolipomas on imaging which can be distinguished by biopsy. Bieko, in April 2010 presented an extra-adrenal myelolipoma for which imaging characteristics suggested a retroperitoneal liposarcoma. Preoperative fine needle aspiration suggested myelolipoma which was confirmed on final pathology (18). Along similar lines Zeiker described the case of a simultaneous extra-adrenal perirenal lesion and a retroperitoneal lesion both with imaging characteristics suggestive of liposarcomas; pathologic analysis was conclusive for a myelolipoma. Final pathology usually clears preoperative ambiguity related to imaging characteristics (19). Pathologic distinction between myelolipomas and extramedullary haematopoietic tissue can however pose a challenge. The latter is more often multiple and associated with hepatosplenomegaly. Myeloproliferative disorders need to be ruled out by bone marrow biopsy (17).

Conclusion

In summary, myelolipomas are rare lesions most commonly associated with the adrenal glands. These lesions are universally benign and therefore can be treated conservatively if diagnosed accurately. When associated with the adrenal gland the diagnosis can be assumed based on imaging characteristics. Extra-adrenal lesions need to be confirmed by biopsy if conservative care is contemplated. Because of the diagnostic dilemma and the need to rule out malignancy most lesions are definitively diagnosed following surgical excision.

References

1. Prahlow JA, Loggie BW, Cappellari JO, Scharling ES, Teot LA, Iskandar SS. Extra-adrenal myelolipoma: report of two cases. South Med J 1995; 88(6):639-43.
2. Kamen BF, Elder DE, Fraker DL, Sigelman ES. Extra-adrenal myelolipoma: MR imaging findings. AJR Am J Roentgenol 1998; 171(3):721-3.
3. Wani NA, Kosar T, Rawa IA, Qayum A. Giant adrenal myelolipoma: incidentaloma with a rare incidental association. Urol Ann 2010;2(3):130-3.
4. Olsson CA Krane RJ, Klugo RC, Selikowitz SM. Adrenal myelolipoma. Surgery 1973;73(5);665-70.
5. Pascual Garcia X, Bunjons Tur A, Rodriguez Faba O, Gomez Ruiz JJ, Palou Redorta J, Villavicencio Mavrich H. Extra-adrenal perirenal myelolipoma: case report and literature review. Actas Urol Esp 2007;31(8):932-4.
6. Le Bodic MF, Mussini-Montpellier J, Magois JY, Lepère J. Myelolipome a localisation gastrique. Arch Anat Path (Paris) 1974; 22(2):119-22.
7. Gao B, Sugimura H, Sugimura S, Hattori Y, Iriyama T, Kano H. Mediastinal myelolipoma. Asian Cardiovasc Thorac Ann 2002;10(2):189-90.
8. Sneiders A, Zhang G, Gordon BE. Extra-adrenal perirenal myelolipoma. J Urol 1993;150(5 Pt 1):1496-7.
9. Nalesnik MA, Martinez AJ, Heros RC. Intracranial lipoma with hematopoietic elements (myelolipoma): report of a case with successful surgical resection. Cancer1982;50(2):295-9.
10. Kumar M. Duerinckx AJ. Bilateral extra.adrenal perirenal myelolipomas: an imaging challenge. AJR Am J Roentgenol 2004;183(3):833-6.
11. Brietta LK, Watkins D. Giant extra-adrenal myelolipoma. Arch Pathol Lab Med 1994;118(2):188-90.
12. Boudreaux D, Waisman J, Skinner DG, Low R. Giant adrenal myelolipoma and testicular interstitial cell tumour in a man with congenital 21-hydroxylase deficiency. Am J Surg Path 1979;3(2):109-23.

13. Amin MB, Tickoo SK, Schultz D. Myelolipoma of the renal sinus: an unusual site of for a rare extra-adrenal tumour. Arch Pathol Lab Med. 1999; 123(7):631-4.
14. Rao P, Kenny PJ, Wagner BJ, Davidson AJ. Imaging and pathologic features of myelolipoma. Radiographics 1997;17(6):1373-85.
15. Plaut A. Myelolipoma in the adrenal cortex ; myeloadipose structures. Am J Pathol 1958;34(3):487-515.
16. Bishop E, Eble JN, Cheng LWang M, Chase DR, Orazi A, O'Malley DP. Adrenal myelolipomas show non-random X-chromosome inactivation in hematoportic elements and fat: support for a clonal origin of myelolipomas. Am J Surg Path 2006;30(7):838-43.
17. Bartumeus Martínez P, Ripollés González T. Extra-adrenal retroperitoneal myelolipoma. Actas Urol Esp 2009;33(4):439-42.
18. Beiko D, Roldan H, Sengupta SK, George RL. Laparoscopic excision of a large extra-adrenal perirenal myelolipoma. Can Urol Assoc J 2010;4(2):E39-E41.
19. Zieker D, Königsrainer I, Miller S, Vogel U, Sotlar K, Steurer W, Königsrainer A, Lehmann TG. Simultaneous adrenal and extra-adrenal myelolipoma – an uncommon incident: case report and review of the literature. World J Surg Oncol 2008;6:72.