Presacral myelolipoma: Imaging features

Myélolipome présacré : aspect en imagerie

We report the case of a 40-year-old woman who was followed in the gastro-enterology department for coeliac disease and who consulted recently with abdominal pain, loss of weight that had been evolving for three months.

Non-contrast and contrast-enhanced computed tomography (CT) of the abdomen and pelvis showed a presacral 11.5 x 8.5 x 5 cm mass with fatty and soft-tissue components. Abdominal MRI was performed. It did not provide any new semiotic element but confirmed the coexistence of a fatty component with another quantity of tissue forming zones of moderate enhancement. The patient underwent CT-guided percutaneous micro-biopsy.

The histopathologic study showed predominantly mature adipose tissue intermixed with extramedullary hematopoietic components. This was consistent with the diagnosis of myelolipoma.

Discussion

This paper is a case report of a presacral myelolipoma. There are only 75 papers in PubMed with the words extra-adrenal myelolipoma.

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. Since the initial description of these rare tumors, case reports have been published for both adrenal and extra-adrenal locations [1].

It is a relatively uncommon benign tumor composed of mature adipose tissue admixed with benign mature hematopoietic elements in varying proportions.

It arises in males and females with approximately equal frequency. Average patient age at discovery is about 50 years (range 17 to 93) but myelolipoma is uncommon in patients younger than 30 years. The most common site of involvement is the adrenal gland but may rarely occur in extra-adrenal locations [2]. Other reported sites include the presacral areas of the retroperitoneum, renal hilum, lung, mediastinum, liver, leptomeninges and stomach [1,2]. Previous reports of extra-adrenal myelolipomas revealed that more than half of these cases occurred in the presacral region [3].

The etiology is still obscure. Some reports proposed that endocrine dysfunction could be included in the etiology of adrenal and extra-adrenal myelolipoma. Clinical examples like Cushing’s syndrome, Addison’s disease, adrenal hyperplasia, and chronic use of exogenous steroids have been associated with myelolipoma [3].

Singla et al. [4] summarized 37 reported cases of extra-adrenal myelolipomas. Eight patients had a history of diabetes mellitus (21.6%), seven had a past history of cancer (18.9%), and six had a past history of steroid use (16.2%) [3]. It is classically asymptomatic and found in a normal adrenal gland [5]. Presacral myelolipomas are usually asymptomatic also but may manifest with pressure symptoms due to mass effect on the rectum, ureters, or urinary bladder.

At CT, presacral myelolipomas appears generally well-circumscribed heterogeneous mass with predominantly fat-containing masses that contain areas of enhancing soft-tissue (figure 1).

Extra-adrenal lesions contain less fat than the adrenal masses (50 versus 90%) [1]. At MR imaging, areas of high T1 signal intensity and intermediate to high T2 signal intensity with loss of signal intensity on fat-suppressed images are characteristic and confirm the presence of intratumoral fat [5] (figure 2). We noted on MRI images signs consistent with coeliac disease such as the jejunisation of ileal wire loops.

The differential diagnosis for fat-containing presacral masses includes lipoma, liposarcoma, teratoma, and lipomatosis. Lipo-sarcoma typically exhibits infiltrative growth [5], which is different from the well-circumscribed aspect of the tumor in our case. Teratoma is usually not median. The differentiation of presacral myelolipoma from other fat-containing lesions may not always be
Extra-adrenal myelolipomas need to be confirmed by biopsy if a conservative approach and a follow up are contemplated. Most lesions in previous publications underwent surgical excision [1] to rule out malignancies. In our case, the patient with the presacral mass was asymptomatic and the follow up was decided after a collegiate decision.

**Conclusion**

Extra-adrenal myelolipomas are rare lesions universally benign. Sometimes, the diagnosis is suggested by imaging but need at least the practice of core needle biopsy to rule out malignancies. Key messages:

- myelolipoma is a rare tumor that can have both locations: adrenal and extra-adrenal;
- imaging of myelolipomas reveals typically an heterogeneous lesion with predominantly fat-containing masses that contain areas of enhancing soft-tissue;
- an histological proof is mandatory for extra-adrenal myelolipoma mainly to rule out malignancies such as liposarcoma.

**Disclosure of interest:** the authors declare that they have no competing interest.

**References**