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Case Report

# Malignant transformation of a mature retroperitoneal teratoma into adenocarcinoma ☆

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ABSTRACT

Retroperitoneal mature teratoma is a rare congenital tumor, far behind ovarian and testicular localizations. Malignant transformation of teratomas is extremely uncommon. We report a case of a 23-year-old female, who presented with chronic pelvic heaviness, abdominopelvic ultrasound revealed a cystic mass in the right iliac fossa, initially considered as a functional ovarian cyst. Given the persistence of the patient's symptoms a pelvic MRI was requested. It showed a right retro-colic cystic unilocular with parietal nodule. The two ovaries showed normal appearance. A retroperitoneal teratoma was suggested given the retroperitoneal topography of the mass, and the presence of a tissular parietal nodule. She underwent surgery and the diagnosis was confirmed after histological study



## Introduction

Mature cystic teratomas represent a group of tumors derived of tissues from at least 2 of the 3 germ layers [1]. They mainly occur in the gonads, thus, retroperitoneal teratomas are rare, accounting for only 10% of abdominopelvic localizations [2]. They are usually diagnosed in young patients, usually during the first year of life [2]. Malignant transformation is extremely uncommon. It has been reported to occur in 1%-2% of mature cystic teratomas [3].

We report a case of malignant transformation of a mature retroperitoneal teratoma in a 23-year-old female, suspected on magnetic resonance imaging (MRI) and confirmed histologically after surgical removal.

## Observation

We present the case of a 23-year-old female, who presented with chronic pelvic heaviness. No particular medical or surgical history was noted. The clinical and gynecological examination was unremarkable. Abdominopelvic ultrasound revealed a cystic mass in the right iliac fossa, initially considered as a functional ovarian cyst. A follow-up sonography was performed later, which described the same findings. Given the persistence of the patient's symptoms a pelvic MRI was requested. It showed a right retro-colic cystic unilocular, non-infiltrative mass, with high signal intensity content on T2-W images (Fig. 1), surrounded by a thin wall, and containing a parietal nodule with isosignal intensity on T1-W and T2-

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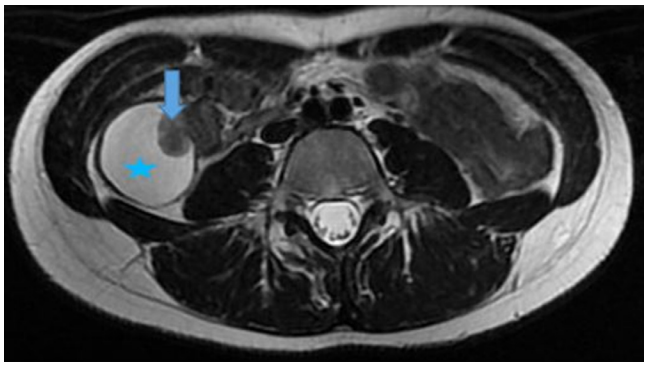

**Fig. 1 – T2 sequence, axial section: unilocular right retroperitoneal cystic mass (star), in clear T2 hypersignal, containing a tissue bud hanging from the wall in T2 isosignal (arrow).**

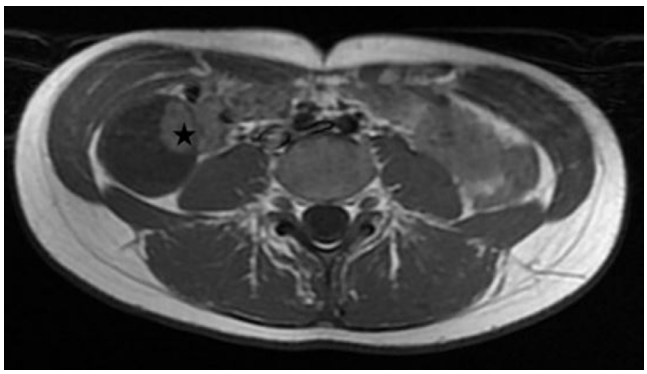

**Fig. 2 – T1 sequence, axial section: unilocular right retroperitoneal cystic mass, in T1 hyposignal, containing a tissue bud hanging from the wall in T1 iso signal (star).**

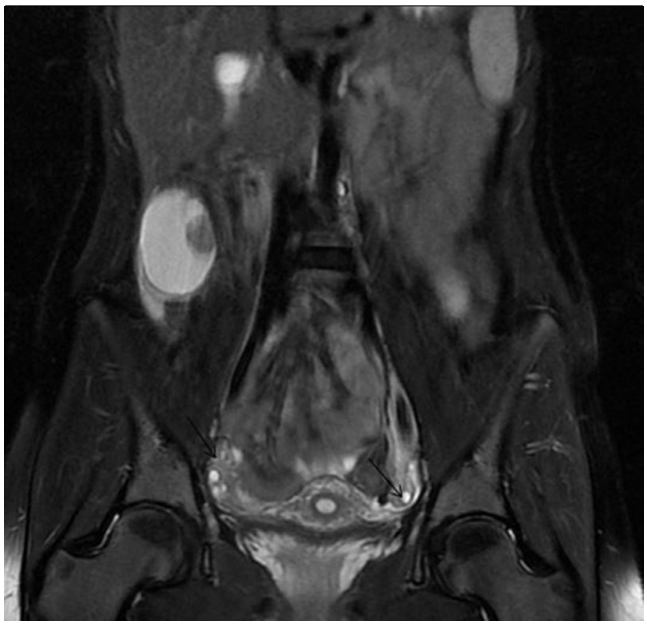

**Fig. 3 – FATASAT T2 sequence: no fatty component in the retroperitoneal mass, the two ovaries are of normal morphology (arrow).**

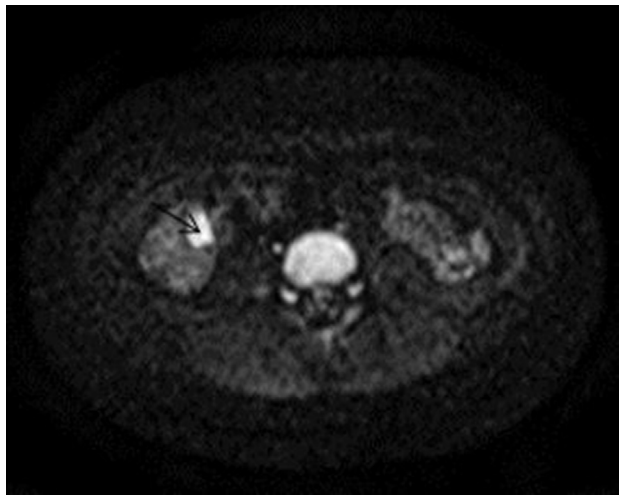

**Fig. 4 – B1000 diffusion sequence: hypersignal of the tissue component (arrow).**

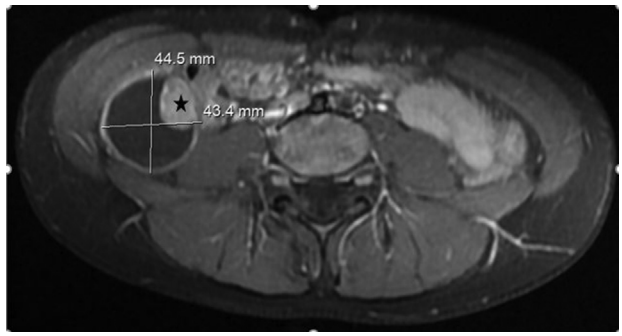


**Fig. 5 – FATSAT T1 sequence with injection of gadolinium: intense enhancement of the tissue bud (star), the cystic mass measures: 44 x 43 mm.**

W sequences (Figs 1 and 2). This nodule showed no signal drop after fat suppression (Fig. 3). However, it demonstrated an important restriction of diffusion (Fig. 4) with marked enhancement after contrast medium injection (Fig. 5). The two ovaries showed normal appearance (Fig. 3). A retroperitoneal teratoma was suggested given the retroperitoneal topography of the mass, and the presence of a tissular parietal nodule. A complementary CT-scan was carried out to look for a fatty component but was equivocal (Fig. 6). Surgical resection was indicated. Microscopic study the specimen demonstrated a mature retroperitoneal teratoma with signs of malignant transformation turning it into an adenocarcinoma (Figs. 7a and 7b).

## Discussion

Retroperitoneal teratoma is a rare congenital tumor, representing only 1% to 10% of primary retroperitoneal tumors [2], far behind ovarian and testicular localizations. Malignant transformation of teratomas is extremely uncommon and represents only 1% to 2% of cases.

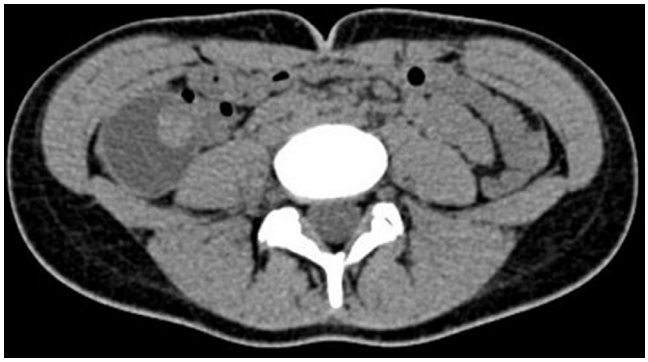

**Fig. 6 – abdominal CT scan without injection of contrast product, axial section: absence of fatty component of the cystic mass.**

The clinical presentation depends of the tumor size and the patients' age. Retroperitoneal teratoma can be diagnosed at any age, with a first peak before 3 years and a second after twelve. They are usually benign tumors that mainly affect female patients [4].

The particularity of our observation is the advanced age of the patient (27 years), the retroperitoneal topography and the carcinogenesis of the teratoma, which is an extremely rare association.

The tumor is asymptomatic in 15%-30% of cases. Sometimes, a vague abdominal pain is reported. Signs of compression of the neighboring organs, especially urinary and digestive, may occur in the case of large tumors [5].

There are 2 essential tumor markers: alpha-fetoprotein and Human chorionic gonadotropin (Beta-hCG), which are essential for therapy indications and post-therapeutic monitoring [6].

Abdominal ultrasound may demonstrate a mass with variable echogenicity depending on the predominance of the sebaceous or lipidic component. The tumor may contain echogenic foci with acoustic attenuation (hairs, calcifications teeth), or a densely echogenic shadowing nodule projecting into lumen. In other cases, the tumor appears to be heterogenous, which reflects the complexity of the structures that make up the tumor (fat, sebum, serosity, hair, calcifications, cartilage, and variable tissues) [7,8], Malignant transformation cannot be evoked on ultrasound. However, color Doppler may reveal the presence of a suspicious tissue associating an intratumoral blood flow, a high pulsatility index, and a lowered mean resistance [3].

CT is performed mainly for its excellent spatial resolution to confirm the retroperitoneal location of the tumor and also to look for fatty or calcified contingents.

MRI gives better assessment of retroperitoneal masses as it confirms the cystic nature of the lesion and characterizes its tissular component. Malignancy can be suspected in the presence of an invasive tissue growth with irregular borders crossing the cyst wall, a marked enhancement after the contrast medium injection and a diffusion restriction [8].

In our case, there were 2 MRI signs that suggested malignancy; marked restriction on diffusion weighted images and intense contrast enhancement of the nodule. However, the regular appearance of the nodule borders, as well as the integrity of the cyst wall was argument against malignant transformation theory.

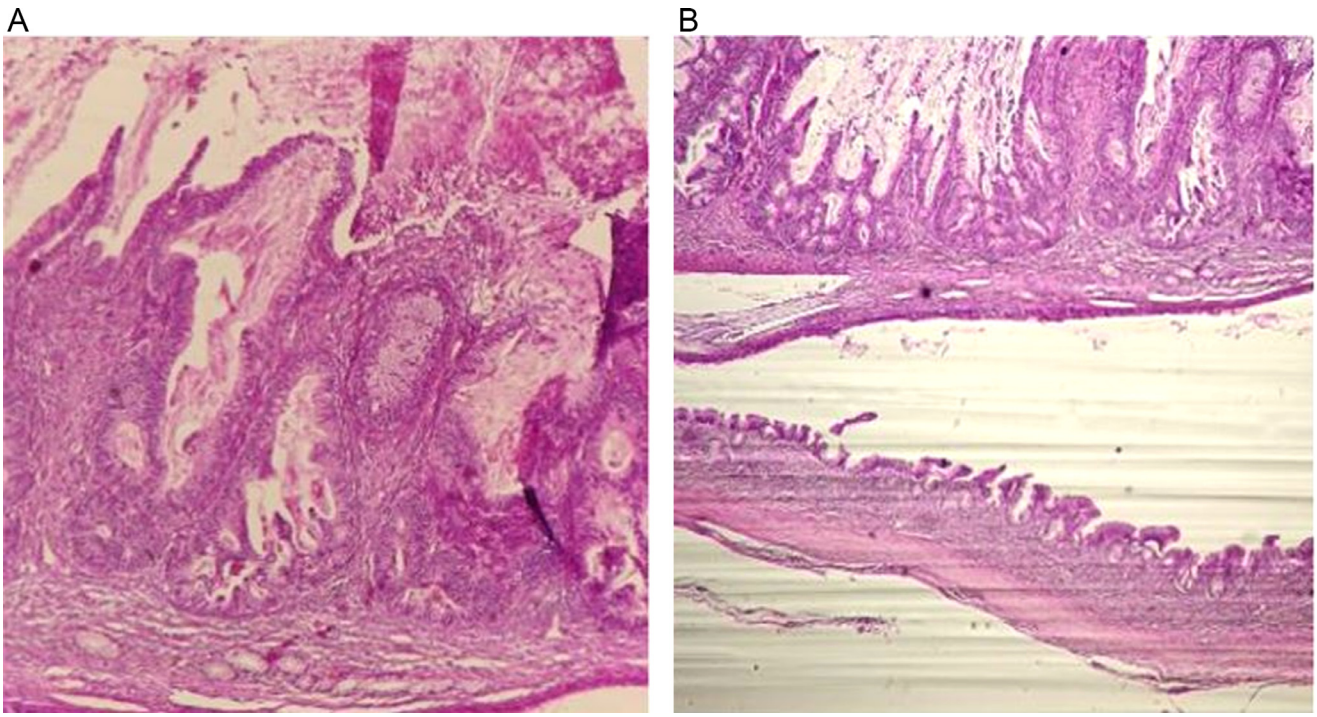


**Fig. 7 –** (a, b): **histological sections, Hemalun Eosine HE x25, HEx50: mature cystic teratoma with respiratory epithelium and an enteric-type adenocarcinoma.**

The treatment of retroperitoneal teratoma is exclusively surgical [3]. In our patient the diagnosis of a malignant contingent was confirmed on histology. Fortunately, the surgical team was informed of suspected malignancy, thus, the surgical resection was performed with safe margins.

## Conclusion

Retroperitoneal teratoma is a rare congenital tumor, let alone its malignant transformation. Imaging plays a crucial role in therapeutic management, by specifying the location and characterization of the tumor. MRI remains the key examination used to demonstrate signs of malignant transformation. Treatment is exclusively surgical with anatomopathological study of the surgical specimen to confirm carcinogenesis. Postoperative monitoring is based on clinical, biological, and radiological examination.

## Authors' contributions

All authors contributed equally to this work.

## Patient consent

Written informed patient consent for publication has been obtained.

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