Asian Journal of Medicine and Health

19(4): 10-15, 2021; Article no.AJMAH.67668
ISSN: 2456-8414

Retroperitoneal Primary Cystic Tumours: (Case Series of Seven Patients and Literature Review)

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Authors' contributions

This work was carried out in collaboration among all authors. Author OL Corresponding author writing the paper. Author SREJ writing and correction of the paper. Author ED correction of the paper. Author RB correction of the paper. Author FC correction of the paper. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/AJMAH/2021/v19i430317
Editor(s):
(1) Dr. P. Veera Muthumari, V.V. Vanniaperumal College for Women, India.
Reviewers:
(1) Santanu Bhattacharya, Maharaja Jitendra Narayan Medical College and Hospital, India.
(2) Tcheuwebe Liale Honore, Gatesville University, Cameroon.
Complete Peer review History: http://www.sdiarticle4.com/review-history/67668

Clinical Practice Article

Received 18 February 2021
Accepted 23 April 2021
Published 08 May 2021

ABSTRACT

Primary retroperitoneal cystic tumors include all cystic tumors that arise in the retro and sub peritoneal space independent of the kidney, the excretory tract, the adrenal gland, large vessels and contiguous viscera. We propose a retrospective study of 7 cases of primary retroperitoneal cystic tumors, collected in the department of digestive cancer surgery and liver transplantation at the Ibn Rochd University Hospital in Casablanca over a period of 10 years (2009 to 2019). Our patients are divided into 6 women and 1 man with an average age of 42 years. The diagnostic time varies from 3 months to 3 years with an average of 10 months. The revealing signs are mainly pain (7 cases or 100%) and abdominal mass (3 cases or 43%). The positive diagnosis is based on ultrasound (2 cases or 40%) but above all on computed tomography (7 cases or 100%) which specifies the retroperitoneal site as well as the relationship of the tumor to neighboring organs. Its confirmation can only be histological. Surgery with total resection is the treatment of choice.

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Keywords: Cystic neoplasms; primitive; retroperitoneum.

1. INTRODUCTION

Primary retroperitoneal cystic tumors include all cystic tumors that arise in the retro and sub peritoneal space independently of the kidneys, excretory tract, adrenals, large vessels and attached viscera. These cysts are usually asymptomatic. Their discovery is often late when they reach a very large size, due to their retroperitoneal location and clinical latency. There is significant overlap in the imaging appearance of these tumors. However, familiarity with the most relevant radiological features, combined with clinical information, may suggest a specific diagnosis in some cases [1]. Surgical excision is the only effective treatment, sometimes potentiated by other complementary treatments and the evolution is good in the majority of cases [2].

Retroperitoneal cystic tumours are rare. Their isolated incidences vary from 1/5,750 to 1/250,000 [3,4]. They can be divided into neoplastic and non-neoplastic categories. Neoplastic causes include mucinous cystadenoma, cystic mesothelioma. Non-neoplastic causes include lymphangioma, Mullerian cysts, epidermoid cysts, postoperative lymphoceles and hematoma [5].

Through the study of 7 observations, collected in the department of digestive cancer surgery and liver transplantation at Ibn Rochd hospital in Casablanca, over a period of 10 years (2009-2019) and through a review of the literature, we propose to make a clinical, anatomopathological, therapeutic and evolutionary approach of these tumors.

2. MATERIALS AND METHODS

This is a study of 7 cases of primary retroperitoneal cystic tumors collected in the department of digestive cancer surgery and liver transplantation at Ibn Rochd hospital in Casablanca over a period of 10 years, from 2009 to 2019. The patients recruited in this study were those diagnosed with a primary retroperitoneal cystic tumor during the study period, with histological confirmation, whose age was greater than 14 years. The information gathered for this study was collected from the department's archives, which include epidemiological, clinical, imaging, therapeutic, pathological and evolutionary data.

3. RESULTS AND DISCUSSION

3.1 Results

The age of our patients varied between 19 and 71 years, with an average age of 42 years. The standard deviation along with the mean age of the population is 20,17. We noted a female predominance with 6 female patients (86% of cases) and 1 male case (14% of cases), i.e. a sex ratio of 0.17.

The average delay of consultation was 10 months with extremes ranging from 3 months to 3 years. Symptoms (Table 1) were dominated by abdominal pain, of variable intensity and topography, which was present in all our patients, i.e. 100% of cases. Evolving for less than 6 months in 3 cases, i.e. 42%, between 6 and 12 months in 2 cases, i.e. 29% and more than 12 months in 2 cases, i.e. 29%. Associated in 1 case, (14%) with cough and effort dyspnea. Signs of digestive compression were represented by constipation in 2 cases (28.5%) associated in 1 case with nausea and vomiting. No case presented urinary signs or vascular compression. An alteration of the general state of health (AEG) was found in 2 cases (28.5%). The physical examination found a fever in 1 case (14%). The conjunctiva was discolored in 1 case (14%). The mass was palpable in 3 patients (43%) as follows: the right hypochondrium and the right flank in the first case, in the right flank and the right iliac fossa in the second case, and in the left flank in the third patient.

For the radiological assessment, abdominal and pelvic CT scans (Figs. 1,2,3) were performed in all patients, i.e. 100%. It allowed the retroperitoneal location of the mass to be retained in all 7 cases (100%). It specified the cystic nature of the process in 6 patients (86%) and the solid-cystic nature in 1 patient (14%). The primary nature of the masses was specified in all our patients (100%). The size of the tumor was between 10 and 20 cm: in 5 cases, 71% and greater than 20 cm: in 2 cases, 29%. The average size was 17 cm. The relationship of the tumor was specified in 6 cases, i.e. 86%; the tumor came into contact with the psoas muscle in 2 cases. In contact with the head of the pancreas and the duodenum, and pushed the intestinal tract to the contralateral side in 1 case. In contact with the liver and the right kidney, which was pushed up and forward, it was leaning against the right diaphragmatic pillar in 1 case. In one

case, it was in contact with the colonic framework and the small intestines anteriorly and the vascular axes (aorta and inferior vena cava) posteriorly. It was in contact with the right adrenal gland in one case. Only one patient underwent a scan-guided biopsy. The histological examination concluded that it was a cystic lymphangioma. None of our patients had MRI.

The treatment in our series was surgical, in 100% of cases. No patient received neoadjuvant therapy. The approach used was midline transperitoneal laparotomy, which was chosen for all cases (100%). Exploration showed left retroperitoneal location in 4 cases (57%), and right in 3 cases (43%). The tumor structure was cystic in all our patients (100%). The tumors were primitive (independent of retroperitoneal organs) in all patients (100%). With the presence of adhesions to the posterior parietal peritoneum in 1 case, 14%. Removal of the tumor mass was attempted in all patients, and was considered complete in all patients, 100%. The tumor was removed alone, without associated visceral resection.

The histological diagnosis (Table 2) of certainty was only possible after anatomopathological reading of the surgical excision specimen in 6 cases (86%). And of the scan-guided biopsy in 1 case (cystic lymphangioma), i.e. 14%. This study confirmed the primary retroperitoneal origin of the tumors; the results were dominated by cystic lymphangioma, in 5 cases, i.e. 72%. Then mucinous cystadenoma in 1 case (14%). And cystic mesothelioma in 1 case (14%). The removal was complete in all our patients.

The immediate post-operative follow-up was simple in all patients (100%), with uncomplicated skin healing and a functional return to normal life. Post-operative mortality was zero. The average postoperative stay was 4.4 days: 4 days for 4 patients (57%) and 5 days for 3 patients (43%).

The long-term evolution was difficult to assess due to the lack of regular follow-up in most patients, 5 cases were lost to follow-up, i.e. 71%. In 2 cases, the evolution was considered favorable with a 6-month follow-up in one patient and a 10-month follow-up in the other.

The discussion should not repeat the results, but provide detailed interpretation of data. This should interpret the significance of the findings of the work. Citations should be given in support of the findings. The results and discussion part can also be described as separate, if appropriate.

3.2 Discussion

Our study includes 7 cases of retroperitoneal cystic tumours followed up between 2009 and 2019. Of these, 5 were cystic lymphangioma, 1 was mucinous cystadenoma and 1 was cystic mesothelioma. The mean age of retroperitoneal cystic tumours is 53 years with extremes of 15 to 93 years and a maximum frequency after 40 years [4,6]. In our series, the age of our patients varies between 19 and 71 years with an average of 42 years. The mean age of retroperitoneal cystic tumours is 53 years with extremes of 15 to 93 years and a maximum frequency after 40 years [4,6]. The predominance of one sex or the other varies according to the series and the histological type [2]. A strong female predominance is found in our study. Due to their development in a complacent space, retroperitoneal cystic tumours remain asymptomatic for a long time. The late diagnosis is characteristic of these tumours. The first symptoms only appear when the tumour is large enough to displace or compress the neighbouring organs [7]. The time to diagnosis in our series varies between 3 months and 3 years with an average of 10 months. Symptomatology is predominantly pain, which is present in 50-90% of cases [2,8]. Associated with signs of compression of neighbouring digestive or urinary organs or vascular compression. In our series, they were in the form of digestive signs such as constipation in 2 cases, associated in 1 case with nausea and vomiting. General signs were present in 30% of cases [8,9]. In our series, alteration of the general state, asthenia and weight loss were found in 2 patients (28.5%), and fever in 1 patient (14%). The conjunctiva was slightly discoloured in 1 patient (14%). Abdominal palpation revealed an abdominal mass of variable size in 50% of cases [2,8].

Currently, computed tomography (CT) and magnetic resonance imaging (MRI) are the mainstay of the evaluation of primary retroperitoneal masses by locating and characterizing non-invasively features such as composition (e.g. presence of fat or calcification), size, relationship to adjacent structures [6]. The diagnosis of a primary retroperitoneal mass can be made once the location is confirmed as in the retroperitoneal space and after an organ of origin is excluded. Displacement of normal retroperitoneal structures, such as retroperitoneal organs or vascular structures, suggests that a mass is retroperitoneal in location [10].

Table 1. Distribution of patients according to clinical signs

Clinical signs	Number	Frequency
Abdominal pain	7	100 %
Abdominal masses	3	43 %
Deterioration of general condition	2	28,5 %
Digestive signs	2	28,5 %
Fever	1	14 %
Urinary signs	0	0 %
Asymptomatic	0	0 %

Table 2. Distribution of cases according to pathological findings

Tumor	Potential	Cases	Percentage
Cystic lymphangioma	benign	5	72 %
Mucinous cystadenoma	Malignant	1	14 %
Cystic mesothelioma	uncertain	1	14 %

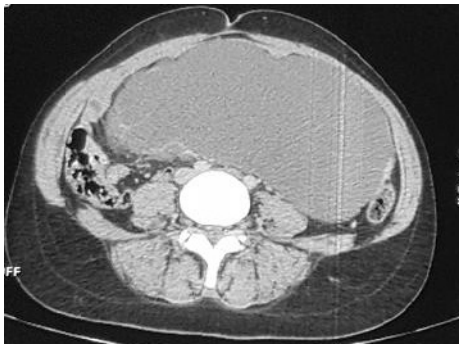

Fig. 1. Abdominopelvic scan: large retroperitoneal cystic formation, pushing backwards and compressing the primary iliac vessels

Surgery is certainly the most important treatment for retroperitoneal cystic tumours [11]. In general, the possibilities of excision are dictated by the size of the tumour and its relationship with the neighbouring organs. In our series the treatment was surgical. None of our patients received additional treatment (radio-chemotherapy). Multiple approaches can be used, depending mainly on the volume and location of the tumour, but also on the habits of the operator [12]. The transperitoneal surgical approach by a median incision is the most commonly used, extraperitoneal by lobotomy is reserved for lateral and small tumours of very probable benignity [2]. In our series, this was the route chosen in all our patients (100%). Complete excision is the method of choice. Retroperitoneal cystic tumours, mainly lymphangiomas and vestigial cysts, are almost always accessible to complete removal, facilitated by evacuation. However, these tumours can form dangerous adhesions with large vessels, and given their usual benignity, it is better to abandon a fragment fixed in depth than to take an inordinate risk of haemorrhage [11,13]. In our series, all our patients benefited from a complete exegesis with respect of the vascular and neural elements. The prognosis of retroperitoneal cystic lymphangiomas is excellent when the resection is complete. Almost all authors agree that it is a benign tumour. The malignant transformation remains widely debated [14]. The malignant potential of mucinous cystadenoma is explained by the simultaneous presence of benign, malignant and borderline foci, which would require the early diagnosis and treatment of these tumours. The evolution after treatment depends on the benignity of the tumour at the time of treatment [11]. Complete removal of mucinous cystadenoma is generally sufficient to obtain a cure; [15] however, recurrence is possible after treatment [16]. The pathogenesis of cystic mesothelioma is controversial, as it has traditionally been considered a benign lesion, but has a potential risk of malignancy [17]. Monitoring of patients is initially clinical by abdominal palpation and pelvic touch. A thoracic-abdominal-pelvic scan every 4 months for the first year and then every 6 months followed by an annual examination for at least 5 years is recommended [18].

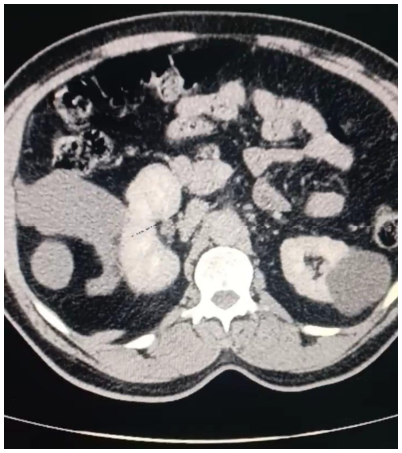

Fig. 2. Abdominal-pelvic CT scan: multi-loculated retroperitoneal cystic formation, coming into contact with the right kidney with preservation of a fatty separation line. This mass connects with the right kidney at an obtuse angle, and pushes it forward and inwards

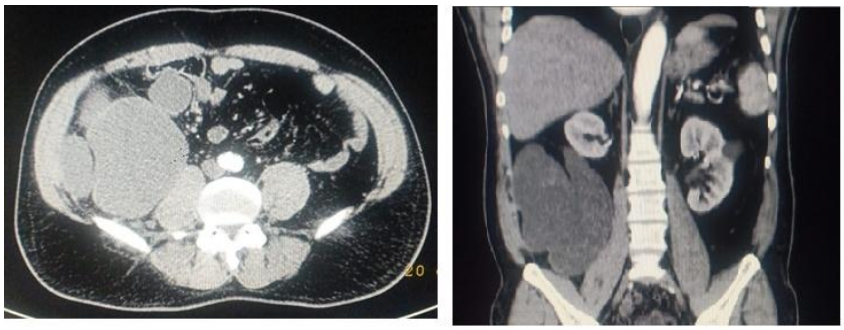

Fig. 3. Abdominal-pelvic CT scan in axial sections with coronal reconstruction: Large retroperitoneal cystic formation, pushing the right colon superiorly and anteriorly, and the lumbar psoas muscle medially, as well as the right kidney superiorly and medially, without any signs of invasion of neighboring organs

4. CONCLUSION

The histological nature of Retroperitoneal cystic tumors these is varied, they are most often benign in nature. A complete imaging work-up including ultrasound, CT and MRI is required preoperatively. Precise histological examination of the surgical specimen allows the diagnosis to be confirmed. Surgical removal is the only effective treatment. It must be codified and complete. The prognosis, although dominated by the risk of recurrence, is excellent.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/67668