Difficulties in Diagnosis of Primitive Retroperitoneal Tumors

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ABSTRACT Primitive retroperitoneal tumors (PRT) are relatively rare, without having specific signs that will lead to their early recognition and can not be include in the therapeutic schemes. They are diagnosed late, when they have been abdominalized through volume, invading the intra-peritoneal organs. Therefore, we did not hesitate to identify the difficulties in the diagnosis of PRT, where much confusion persists today as everything depends on the precocity of diagnosis.

KEY WORDS retroperitoneal tumors primitive, diagnosis, investigations

Introduction

Primitive retroperitoneal tumors are tumor formations located in the retroperitoneal space, independent of other organs located in this space, which may have very different embryological origins as they can belong to the vascular, lymphoganglionar, muscle, epithelial, vestigial and nervous tissue, contained in the connective tissue of that certain space.

Although there have been major advances in the diagnosis of these tumors, their detection continues to be made in a late moment, in advanced stages of local or systemic evolution. Thus, a significant number of cases can not benefit from complete surgical removal, most times being a real diagnostic and therapeutic challenge for practitioners.

Primitive retroperitoneal tumors represent a difficult chapter of surgical pathology, considering the rarity (0.2% of 60,000 Pack and Tabah tumors), the difficulty of diagnosis, the high percentage of local recurrences after surgical removal and the resistance to the possibilities of adjuvant therapy.

The purpose of this study is to seek answers in order to improve and optimize the diagnosis of patients with primitive retroperitoneal tumors.

Taking age into consideration, statistics performed show a maximum incidence of 82.14% in the age group of patients between 40 and 70 years old, being more frequent after the fourth decade having the extreme ages of 17 and 76 years. This states that, in our study we did not include children as the clinic in which the study was done is addressed to adults only.

Of the 56 total cases studied, 34 involved males (60.71%) and 22 cases (39.29%) were represented by females.

Thus, out of a total of 56 cases, 14 cases (25.00%) were benign primitive retroperitoneal tumors, represented by one hemangioma, one angiomyolipoma, two leiomyomas, five myxomas and five cystic tumors, 31 cases (55.36%) were malignant primitive retroperitoneal tumors represented by eight lymphomas, three fibrosarcomas, three liposarcomas, two myosarcomas, one leiomyosarcoma, one hemangiopericytoma, six schwannomas, three paragangliomas, two adenocarcinomas, one carcinoma, one hepatoma and 11 (19.64%) primitive retroperitoneal tumors with undefined structure.

It should be noted from the start that all retroperitoneal tumors diagnosed in our study were detected in the symptomatic stage.

The clinic was represented by: abdominal pain (was present in 47 cases – 83.93%) and the presence of palpable tumors (40 cases – 71.43%), the moderate increase of abdominal volume (3 cases – 5.36%), followed by a bewildering variety of symptoms belonging to the mechanically or functionally affected

Material and method:

The study presents a retrospective analysis of a group including 56 cases of primitive retroperitoneal tumor, hospitalized and treated in Clinic I Surgery of the Emergency County Hospital in Craiova, between 1999 and 2009.

In the study, data related to age, sex, general and local physical exam were processed together with laboratory tests meant to improve the management of patients with primitive retroperitoneal tumors.

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organs or related to these tumors, as well as urinary manifestations (29 cases – 51.79%), digestive (24 cases – 42.86%), vascular (5 cases – 8.93%) and neoplastic syndrome impregnation (15 cases – 26.79%).

Basically, the clinical data were meant to direct the paraclinical exploration and it cannot be said in any of the cases dealt with that the diagnosis could have been established with certainty on clinical data.

The topography of the tumor was in most cases oriented center-abdominal-right as follows: center-abdominal - 18 cases (32.14%); right hemi-abdominal - 26 cases (46.43%) and, left hemi-abdominal - 11 cases (19.64%) and hypogastrium - 1 case (1.79%).

The imaging investigations were represented by:

- Chest radiography used in 100% of cases did not bring significant data in terms of diagnosis, but for 3 cases of diaphragmatic ascension.
- Simple abdominal radiography used in 100% of cases; the most commonly found pathological element was the presence of abdominal opacity with variable density in 40 cases; displacements of the colic frame in 13 cases, 27 cases of psoas shadow removal.
- The abdominal ultrasound, which was the most used investigation from the arsenal of imaging techniques available (100% of cases) had the merit to highlight the tumor, to specify the solid or cystic nature of it, although it could not always specify the association of the tumor with a specific organ and it could not point out its vascular or visceral relationships.
- Much more valuable in this respect was the computer tomography, but because of the available technical means it could not be carried out only in more than 27 cases – 48.21%; the investigation by MRI was not available only in a total of 23 cases – 41.07%.
- Urography was performed in 16 cases – 28.51%. Besides the exclusion of renal association of the tumor formation, it also revealed the relationship between tumor and kidneys, tumor and ureter and, very importantly, where the nephrectomy was required it certified the functionality of the contralateral kidney, this being an essential condition for the surgery in these cases. Thus, the following imaging modifications were highlighted: dislocation of the right kidney and ureter 9 cases, dislocation of the left kidney 3 cases, mute left kidney through inclusion of tumor 1 case, displacement of both ureters 1 case, displacement of left right ureter 1 case, bladder compression 1 case.
- A contrast substance radiological investigation of the digestive tract was performed in 19 cases – 33.93%. Although its importance was disputed by some authors, it had the merit to rule out, on the one hand, the belonging to the digestive tract of the tumor and, on the other hand, it often established the relationship between the tumor and the digestive organs. Barium Contrast Radiography identified the following radiological changes: dislocations of the right colon and hepatic angle 1 case, dislocations of the right colon 8 cases, dislocations of the left colon 3 cases, dislocations of transverse colon 1 case, splenic angle stenosis 2 cases, descending sigmoid colon junction stenosis 1 case, gastric and duodenal movements 3 cases.
- Exploratory laparotomy was indicated and applied in all the cases in order to remove the retroperitoneal tumor or even to obtain specimens for histopathological examination.
- Laboratory explorations have shown: anemia (19 cases – 33.93%) and a mild increase of ESR (36 cases – 64.29%), as well as hypoglycemia (17 cases – 30.36%) and glycosuria (7 cases – 12.50%) in case of insulin sarcomas, increased catecholamine (2 cases – 3.57%).
- Immunological explorations have shown in some histopathological types the presence of antigens CA 19-9 (4 cases – 7.14%) in increased quantities or of other substances such as interleukins has been indicated as well as a decrease in NK lymphocytes (CD16+CD56+) (natural killer) (9 cases – 17.07% or variations) or variations of the number of T helper lymphocytes (CD3+CD4+) (3 cases – 5.36%) or T suppressor / cytotoxic (CD3+CD8+) (5 cases – 8.93%), but without establishing a cause effect relationship between the retroperitoneal tumor and the occurrence of these markers or lymphocyte populations. The fact is that in most cases studied we observed a marked immunologic depression, with a decrease in T
lymphocytes titre. Furthermore, no genetic studies have shown characteristic changes, in most cases the same oncogenes were observed as in other tumor types.

Results

Primitive retroperitoneal tumors belong to the group of tumors that have obscure developing conditions. Specific factors that could cause or facilitate the appearance of tumors in the retroperitoneal space remain unknown.

The incidence in the age groups is maximum in the fifth decade of life, representing 41.07% of all the cases studied.

No special incidence was recorded for groups of patients with a certain profession.

Primitive retroperitoneal tumors are rare tumors and malignant in most cases, 55.36% (31 cases) of all cases, followed by benign tumors 25.00% (14 cases) and tumors with undefined HP structure 19.64% (11 cases).

Most cases of primitive retroperitoneal tumors were developed by males (60.71%), females having a propensity to develop benign tumors, representing 57.14% (8 cases) of all benign tumors. Malignant tumors have a high affinity for males, representing 67.74% (21 cases) of all malignant retroperitoneal tumors.

The malignant / benign ratio in the PRT group is ~ 2.21 to 1 in favor of malignant tumors.

The clinic of these tumors is very uncharacteristic and misleading. The clinic of these tumors is very uncharacteristic and misleading. Except for the presence of pain (83.93%) and of bulky tumor masses which can be felt through lumbo-abdominal palpation (71.43%) the symptoms and signs interpreted by an uninformed specialist and / or unwittingly can easily lead to a wrong diagnosis.

Laboratory explorations were only indicative. Immunologically speaking, there is no characteristic tumor marker at the present stage of knowledge. Furthermore, not even genetic studies have shown characteristic changes being observed, in most cases, the same oncogenes present and in other tumor types.

Imaging explorations are the ones that suggest the diagnosis, but cannot confirm the nature of the histological tumor (benign or malignant), the value of computed tomography is superior to all others, and the value of the histopathological examination is unquestionable in assessing the diagnosis of a patient with retroperitoneal tumor.

Discussions

Retroperitoneal tumors are considered malignant not only from the histological point of view but also by the pathological impact that they have on other anatomical intra-and retroperitoneal structures, having as main clinic characteristic the discrepancy between tumor size and symptoms: because of their possibility of development in the retroperitoneal space, they evolve for a long time without showing clinical symptoms and even when they reach significant dimensions, they manifest through nonspecific symptoms due to the complications they generate.

The clinical manifestations of retroperitoneal tumors are polymorphous and unspecific as they reveal themselves after a long latency period, in which the tumor had to develop enough to cause significant effects depending on its location and volume by compression, displacement, deformation or invasion of neighboring structures.

Depending on the topography of the tumor formation the following can be mentioned: tumors developed on the median line, center-abdominal can be pulsing, delivering pulses to the aorta (18 cases – 32.14%), in case the tumor evolves from the retroperitoneal space towards the pelvis (1 case – 1.79%), its lower extremity may be palpated through vaginal or ano-rectal examination as a rough formation, fixed, latero-pelvic, retro-rectal, interrecto-vaginal; sometimes it can cause rectal stenosis.

The characteristics of the tumor do not allow, only in a small number of cases, conclusions regarding its benign or malignant nature, as some data obtained by palpation are similar (hardness, fixity). In general, as the tumor gets more stable, the possibility of it being malignant increases.

Echography was the one that highlighted the tumor and specified the solid or cystic nature of it, but could not always specify the membership of the tumor with an organ or the vascular and visceral relations.

The essential data provided by the computed tomography were represented by the highlighting of the retroperitoneal tumor and the exclusion of membership with an organ, the tumor size, and especially the clarification of the relationships with adjacent structures; so, although there was no case in which the tomographic examination certified the futility of the laparotomy, the surgeon was advised of visceral and vascular risks existing in the given case, a very important element in determining the operative tactics and technology.
The desire of some of the authors of this article is to highlight, with the help of specialists in genetics and medical immunology, the existence of several markers with tumor predictive capacity and to determine the presence / absence of proto-oncogenes able to induce the tumor process or to stop its development.

Conclusion:

- The diagnosis is not easy because of the great number of organs which are located in the retroperitoneal space and of the organs whose tumors can evolve in the retroperitoneal space.
- The diagnosis is established most often late because of the discrepancy between tumor size and symptoms, which may lead to illogical or inaccurate therapeutic gestures, which may also contribute to delayed diagnosis.
- The share of diagnosis increases with imaging techniques like: ultrasound imaging, CAT scan and MRI; with mention to the fact that the ultrasound exam reveals the presence of abdominal tumor formations without specifying the association with a certain organ, as they only suggest the diagnosis without confirming it. The value of the laparotomy and of the histopathological examination is undeniable.
- The prognosis of these diseases is influenced by the time of diagnosis, the histopathological type and the topography of the PRT
- Expectancy in early diagnosis and even treatment now rests on the shoulders of geneticists.

References

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