

Primary Retroperitoneal Mucinous Cystadenocarcinoma, Cystadenocarcinoma Mucinous Primary Retroperitoneal, Case Presentation

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Cystadenocarcinoma mucinous retroperitoneal;
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1. Summary

1.1. Introduction: Cystadenocarcinoma Primary retroperitoneal mucinous tumors are extremely rare neoplasms.

1.2. Objective: To present the case of a cystadenocarcinoma mucinous primary retroperitoneal .

1.3. Case Presentation: A 49-year-old female patient who came to our institution with recurring abdominal pain.

Discussion: Primary cystadenocarcinomas of the retroperitoneum are rare, being found more frequently in patients between 30 and 50 years old and have a predilection for the female gender. Where its usual form of presentation is abdominal pain or a palpable mass. Its preoperative diagnosis is rare. Well, the tumor markers are normal and the imaging studies only refer to a cystic or mixed lesion that occupies the retroperitoneal . Treatment is eminently surgical.

1.4. Conclusion: Cystadenocarcinoma Primary retroperitoneal mucinous tumors are a rare neoplasm. Where its genesis is still unclear. There is a predominance of the female sex with an age of presentation of 45 years, manifesting as abdominal pain or palpable mass among the cases. Where preoperative diagnosis is extremely rare, where imaging can only diagnose a space-occupying lesion in the retroperitoneum. Its treatment with surgical intent is surgery where tumor excision is the rule.

2. Abstract

2.1. Introduction: Primary retroperitoneal mucinous cystadenocarcinoma are extremely rare neoplasms.

2.2. Objective: To present the case of a primary retroperitoneal mucinous cystadenocarcinoma.

2.3. Case presentation: A 49-year-old female patient who comes

to our institution due to recurrent abdominal pain.

2.4. Discussion: Primary cystadenocarcinoma of the retroperitoneum are rare, being found more frequently in patients aged 30 to 50 years and have a predilection for the female gender. Where its usual form of presentation is abdominal pain or a palpable mass. Its preoperative diagnosis is rare. Well, the tumor markers are normal and the imaging studies only allude to a cystic or mixed lesion that occupies the retroperitoneum. Treatment is eminently surgical.

2.5. Conclusions: Primary retroperitoneal mucinous cystadenocarcinoma is a rare neoplasm. Where its genesis is still unclear. There is a predominance of the female sex with an age of presentation 45 years, manifesting as abdominal pain or palpable mass among cases. Where preoperative diagnosis is extremely rare, where imaging can only diagnose a space-occupying lesion in the retroperitoneum. Being its treatment with surgical intention the surgery where the tumor excision is the rule.

3. Introduction

Primary retroperitoneal mucinous cystadenocarcinomas [RPMC] are a truly rare pathology. Worldwide there are less than 50 reported cases.[1] It is twice as common in women, with abdominal pain being the usual reason for consultation in those who suffer from it. Preoperative diagnosis is made during studies for nonspecific abdominal symptoms or incidentally when performing imaging tests in the study of other diseases. Their differential diagnosis can be difficult and it is not uncommon for them to be confused with renal cysts [2,3] The histogenesis of this unusual neoplasm is unclear. [1,3,4] Surgical exploration is necessary for diagnosis and treatment, and many authors recommend extensive excision that includes total hysterectomy and bilateral salpingo-oophorec-

tomy with enucleation of the retroperitoneal tumor. [5,6] Adjuvant chemotherapy is sometimes given after complete surgical excision. However, the most desirable treatment for this rare tumor remains controversial [4] Here we report on a patient who underwent tumor excision; There is no previous report of this entity in our center.

4. Case presentation

Female patient, white, 49 years old, with medical history. He came to our service due to continuous abdominal pain in the right hypochondrium of moderate intensity radiating to the ipsilateral thigh with slight relief in the supine position and flexion of the right leg, without reporting any other symptoms. Physical examination revealed an ill-defined and mildly tender mass about 10 cm in size over the left lower abdomen. Laboratory tests, including complete blood count, chemistry profile, urinalysis, and chest x-ray, were within normal limits. Abdominopelvic computed tomography revealed a large unilocular cystic mass measuring 7x10 cm, with a solid enhancing portion, probably located in the retroperitoneal space [Figure 1]. There was no evidence of extracystic extension or distant metastasis. With all the above elements, an abdominal examination was performed, confirming the diagnosis of a 7x10 cm retroperitoneal tumor, without observing elements of dissemination or metastasis in the abdominal cavity, uterus and adnexa without macroscopic alterations [Figure 2]. It was decided to remove the tumor. The patient progressed satisfactorily and was discharged after 5 days. A biopsy was received that reported extraovarian primary retroperitoneal mucinous cystadenocarcinoma.



Figure 1: Shows said retroperitoneal tumor unrelated to the ovary or kidney

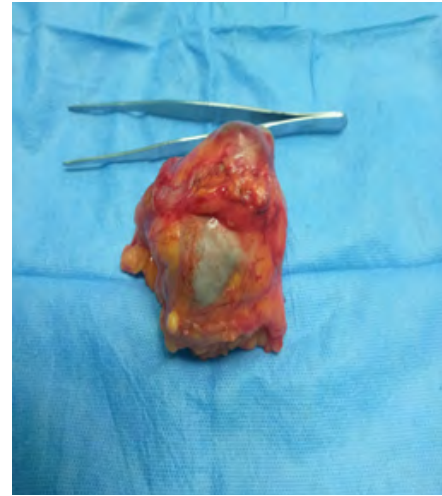


Figure 2: Shows the surgical specimen compatible with primary retroperitoneal mucinous cystadenocarcinoma.

5. Discussion

Primary retroperitoneal mucinous cystadenocarcinoma is a rare tumor. The first case was described in 1977 by Roth et al 2, and to date about 30 cases have been described in the English medical literature and less than 50 cases worldwide. Mucinous cystadenocarcinomas can originate in the pancreas, ovary and retroperitoneum [1,2,3] The present case was located in the retroperitoneum in the absence of adhesion to neighboring organs. Due to the integrity of the pancreas, digestive tract, and ovaries, this tumor was considered primary retroperitoneum. Several hypotheses [1,3,4,6] have been suggested to explain the histogenesis of primary retroperitoneal mucinous cystic neoplasia, and they are heterotopic or supernumerary ovary, retroperitoneal teratoma, intestinal duplication and coelomic metaplasia. To date, the hypothesis that has gained increasing support is coelomic metaplasia [4,5], that is, retroperitoneal mucinous cystadenocarcinomas arise from invaginations of the peritoneal mesothelium, with subsequent mucinous metaplasia. Ultrastructural findings and immunohistochemical observations support this hypothesis [5,7,8].

Retroperitoneal tumors of epithelial origin are a rare entity. The presentation predominates in the fourth-sixth decade in women [range, 33-86 years; mean, 42.4 years; in men, the presentation tends to be late [64-83 years], with a mean age of 46 years, where only 2 of the reported cases were men [4,7,9] Patients generally present to the hospital with a complaint of abdominal pain or a palpable mass. Radiological tests, such as ultrasound, computed tomography, or magnetic resonance imaging, are performed. used to locate papillary nodules demonstrated within a cyst by ultrasound or computed tomography have been reported to suggest malignancy.2,3,4 However, it is often difficult to differentiate a benign neoplasm from a malignant one, or even determine the site of malignancy. origin of the tumor with the use of preoperative radiological images. Tumor markers, in most cases, are normal.

Isolated cases of elevation of CA19.9, CA125 and serum alpha-fetoprotein, and cases of elevation in the cystic fluid from CEA and CA 19.9. The latter could be useful if preoperative FNAC is performed. [2,3,5,6] Serum tumor markers rarely help in diagnosis or follow-up. Sangeetha et al 5 suggested that the presence of glandular epithelial cells and elevated levels of CEA in the cystic fluid were helpful in making the diagnosis. Laparotomy, tumor removal and pathological examination are inevitable to make a correct diagnosis. [3,5,6,9]

The treatment of primary retroperitoneal mucinous cystadenocarcinoma remains controversial. Treatment with curative intent is surgical excision. Some authors recommend combining hysterectomy and oophorectomy. However, in the published cases the female internal genitalia did not present macroscopic or microscopic alterations; Therefore, this attitude could only be justified in postmenopausal patients or those who have satisfied their reproductive desires [5,6,8,9] The association of adjuvant chemotherapy is not part of the standard treatment. There are authors who recommend their association in cases of rupture of the lesion or invasion of neighboring structures. Mikami et al.4 observed a patient who experienced rupture of the tumor capsule intraoperatively with subsequent peritoneal implantation of the tumor, and the patient died 18 months after the initial surgery. Song et al.10, in their series, a laparoscopic approach was performed in three cases; reported cases did not undergo extensive surgical excision, including hysterectomy and bilateral salpingo-oophorectomy. I only mention the recurrence of the disease in the event that the tumor ruptured during extraction. The immunohistochemical findings of this neoplasm resemble those of its ovarian counterpart; Thus, a large number of authors recommend management according to the treatment protocol for ovarian neoplasms, including the staging procedure [3,5,7,9]

The tumor was surgically removed in almost all previously reported cases. Roth et al.1 reported on a patient who had only one tumor removed and died from disease spread 6 months postoperatively. Two patients received tumor excision and adjuvant chemotherapy, but one patient had a paraovarian recurrence 21 months after surgery [10] and another patient died from disease dissemination 4 months after surgery [11]. The patient with histologically borderline malignancy reported by Banerjee et al.3 underwent tumor removal, left salpingo-oophorectomy, and descending colon resection, and mediastinal metastasis developed 4 years later [14]. Hysterectomy and bilateral salpingo-oophorectomy were performed after tumor removal in approximately half of the reported cases, and these cases were alive without evidence of recurrence 3 to 36 months postoperatively. In our case, complete surgical resection was performed, without intraoperative rupture. No macroscopic alterations were evident in the internal genitalia. Our department and the oncology department at our institution decided to use adjuvant chemotherapy. Currently, 8 months after the intervention, as

demonstrated by imaging studies, the patient is disease-free.

6. Conclusion

Primary retroperitoneal mucinous cystadenocarcinoma is a rare neoplasm. It is still unclear where it originated. There is a predominance of the female sex with an age of presentation of 45 years, manifesting as abdominal pain or palpable mass among cases. Where preoperative diagnosis is extremely rare, where imaging can only diagnose a space-occupying lesion in the retroperitoneum. Its treatment with surgical intent is surgery where removal of the tumor is the rule.

7. Conflicts of Interest

The authors declare that there are no conflicts of interest in this article.

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