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Voluminous Pseudomyxoma of the Thigh Secondary to a Mucinous Tumor of the Appendix

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Authors' contributions

This work was carried out in collaboration among all authors. Authors RB and BD are the operating surgeons. Author BD wrote the protocol and wrote the first draft of the manuscript. Authors AE, BD and MN managed the documentary research, wrote the manuscript and proposed the work for publication. Author FC oversaw all the work. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

A 55 year old woman came to our structure for a swelling of the anterior-internal side of the right thighthat has been evolving for 5 years; creating a taut, firm and slightly painful arch. Soft tissue MRI and abdominal CT scan showed an oblong lesion extending from the right flank to the middle third of the right thigh, the lesion is well limited lobulated with liquid signal. It is extended between the abdomen and the right thigh via the right Psoas and Pectine muscles to infiltrate the anteromedial aspect of the right thigh between the adductors while remaining peripheral, the vascular axes are free. A diagnosis of Pseudomyxoma retroperitonei, probably of appendicular origin, was suspected. An exploratory laparotomy with right hemicolectomy and evacuation of retroperitoneal gelatinous ascites and evacuation of the thigh collection through an incision on the inner side. Pathological examination confirmed the diagnosis of an appendicular mucinous tumour with retroperitoneal pseudomyxoma.

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Keywords: Voluminous pseudomyxoma; mucinous tumor; appendix; abdominal CT scan.

1. INTRODUCTION

The peritoneal pseudomyxoma is a clinico-pathological entity corresponding to diffuse peritoneal involvement, composed of mucinous ascites and multifocal mucinous epithelial implants. It is due to the intraperitoneal rupture of a mucinous tumour, the appendicular or ovarian origin of which has long been discussed [1]. Retroperitoneal pseudomyxoma (RPM) is a rare disease characterized by a mucinous collection in the retroperitoneum, Only 45 documented cases reported in English literature.

PMR (pseudomyxoma retroperitonei) usually presents as a painful abdominal mass and rarely as a mass on the anterior aspect of the thigh [2].

In this study, we report a particular topographical form: the pseudomyxoma of the anteromedial aspect of the right thigh.

2. CASE PRESENTATION

Mrs. A, 55 years old, was hospitalized for swelling of the anteromedial aspect of the right thigh that had been evolving for five years. The patient was apyretic and had swelling on the anterior-internal side of the right thigh measuring 20/10 cm, creating a taut, firm, slightly painful arch, with a mobile liquid-like appearance in relation to the underlying planes.

Clinical examination of the stomach showed sensitivity of the right iliac fossa without irradiation.

Biological examinations (blood cell count, hydro electrolytic and hemostasis assessment), were normal, the sedimentation rate was 25 in the first hour.

Soft tissue ultrasonography had revealed a hypoechoic subcutaneous mass of the soft tissues of the upper and inner side of the right thigh, the inner edge of which touches the femoral neurovascular bundle.

Soft tissue MRI had revealed an oblong lesion extending from the right flank to the middle third of the right thigh, the lesion is well limited lobulated with hyperintense fluid signal in T2 hypointense in T1. It is extended between the abdomen and the right thigh via the right Psoas and Pectine muscles to infiltrate the anteromedial aspect of the right thigh between the adductors while remaining peripheral, the vascular axes are free (Fig. 1).

Thoraco-abdomino-pelvic CT scan had objectified a mass of the right iliac fossa centred on the cecum with ureteral invasion and right hydronephrosis associated with numerous peritoneal nodules with an unseen appendix.

The patient was operated on through a medial laparotomy. On exploration, the presence of a cystic mass at the expense of the caeum of probable appendicular origin associated with a gelatinous ascites partitioned at the level of the retroperitoneum coming into contact with the lower pole of the right kidney, the lower face of the liver and the psoas muscle backwards and descending to the anterior face of the right thigh. The collection was emptied and then washed and trimmed (Fig. 2). A right hemicolectomy was performed with terminolateral ileocolic anastomosis.

A counter-incision on the anteromedial aspect of the right thigh made it possible to complete draining and trimming (Fig. 3). A total of 2000 ml of gelatinous liquid was evacuated. The gelatinous appearance of the liquid suggested an intra-abdominal origin (Fig. 4).

The pathological examination: Histologic appearance in favor of a low-grade mucinous appendicular tumor. Gelatinous material of the thigh: deposit of acellular mucin. The after-effects of the operation were simple.

3. DISCUSSION

Although peritoneal and retroperitoneal pseudomyxoma share similar histopathological features, there is variance in their incidence rates. Peritoneal pseudomyxoma shows a female predominance (female to male ratio 2-3:1) and an average age of 53 years, while PMR shows a female to male ratio of 1:1.22, with a maximum age incidence of 60.9 years [2].

Pseudomyxoma literally referring to a gelatin or mucinous collection, commonly seen in the peritoneal cavity. Usually the original organ is the appendix, other rare primary sites being the ovaries, colon, stomach and pancreas. Rarely, a pseudomyxoma is limited exclusively to the retroperitoneal compartment and is therefore called a retroperitoneal pseudomyxoma. A

retrocecal appendage accounts for the majority of these cases, but other origins including the ovaries, ascending colon and rectum have also been implicated in its etiology. Primary mucus-secreting tumours can range from low-grade cystadenoma to malignant cystadenocarcinoma [2].

Unique to our case, the PMR can also extend simultaneously in the anterior part of the thigh along the iliopsoas muscle.

Although these have been described in the literature, the delay in diagnosis can be attributed to the rarity of its incidence.

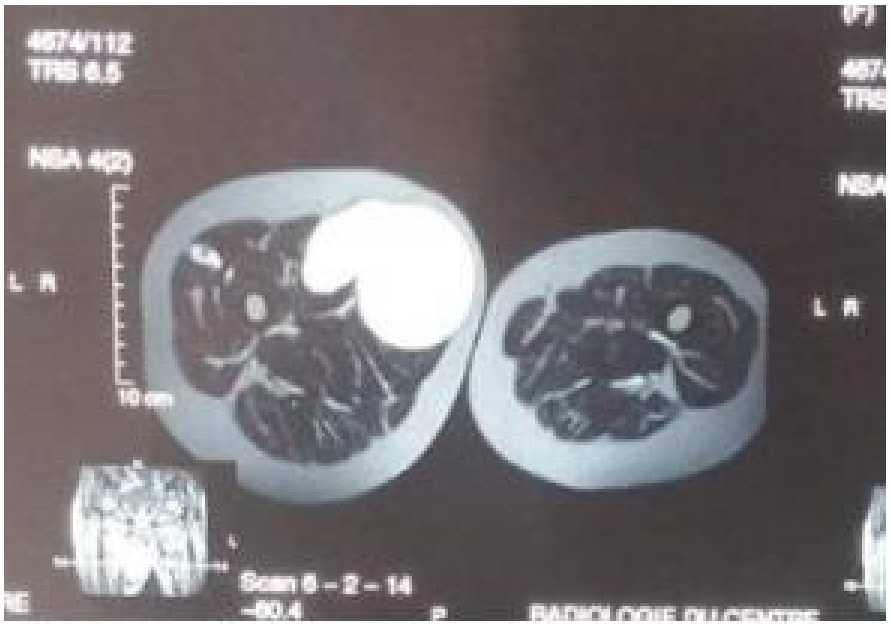

Fig. 1. Oblong lesion of the anteromedial aspect of the right thigh between the fluid signal adductors in hyperintense T2

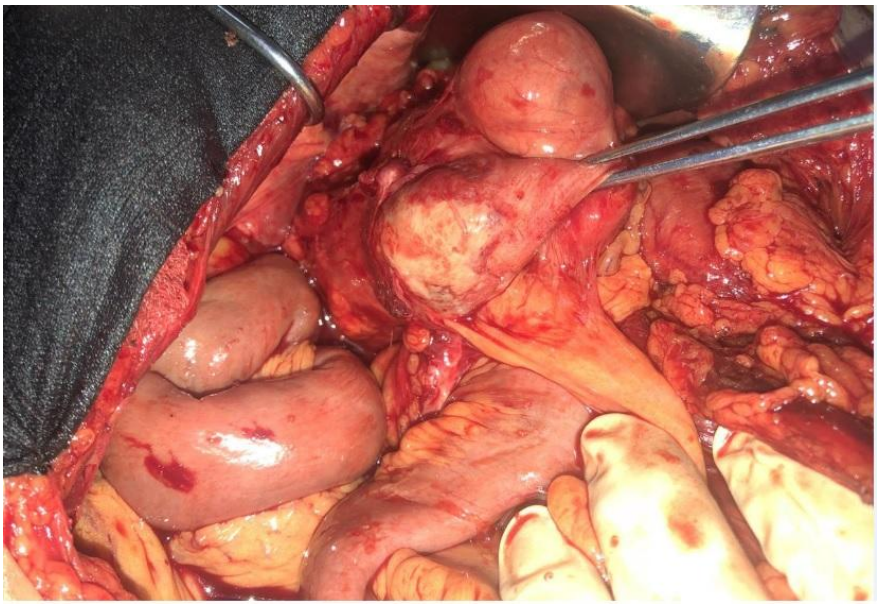

Fig. 2. A cystic mass at the expense of the caeum of probable appendicular origin associated with a gelatinous ascites partitioned in the retroperitoneum

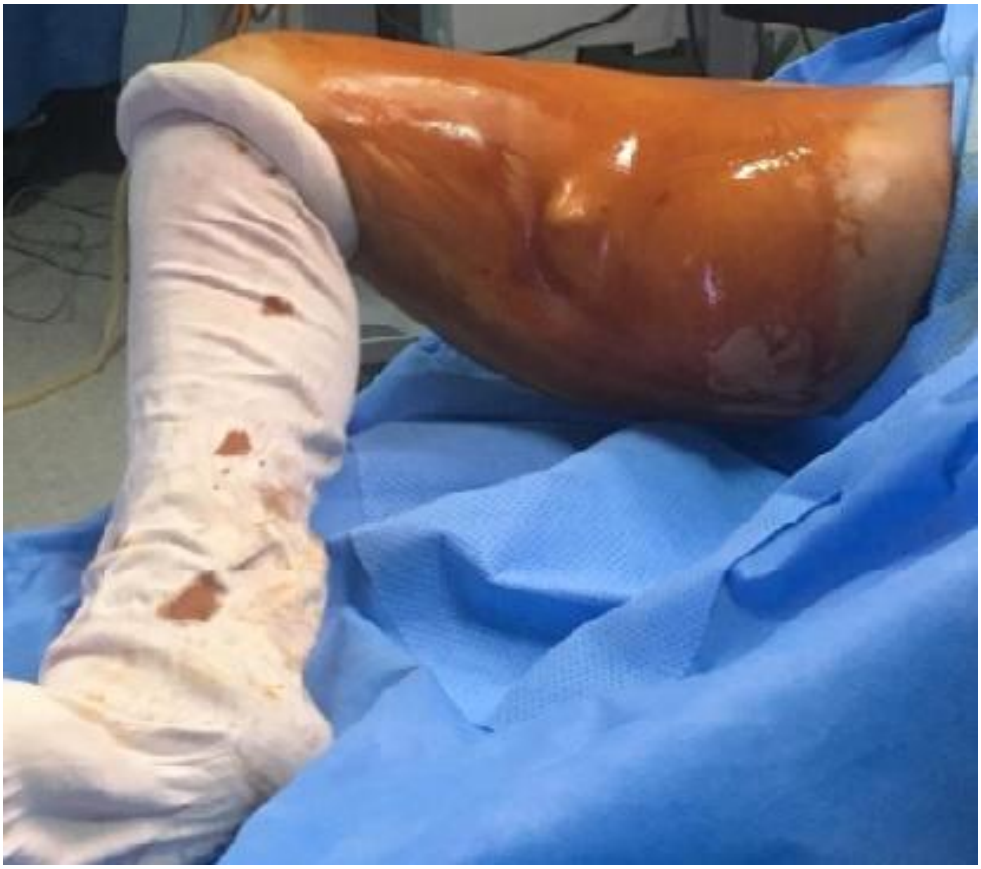

Fig. 3. Swelling of the anteromedial aspect of the right thigh

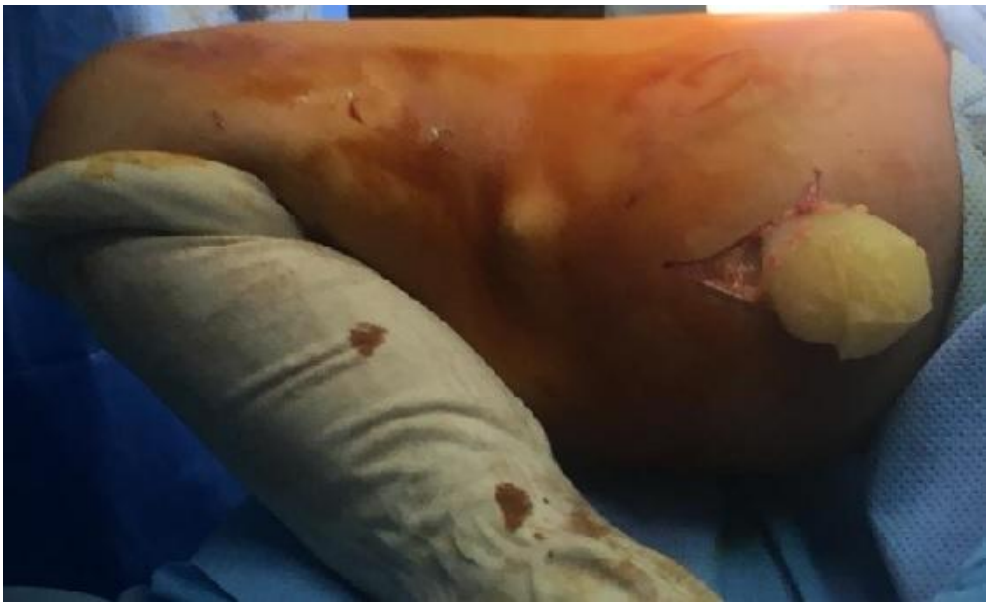

Fig. 4. Evacuation of a gelatinous liquid through an incision on the anteromedial surface of the right cuticle

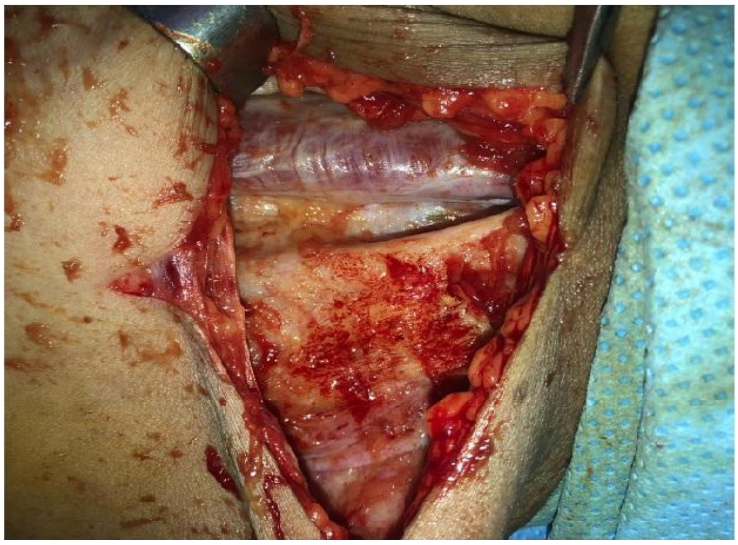

Fig. 5. Residual cavity after evacuation of the mucous liquid from the thigh

In ultrasound as in CT scans,the diagnosis of pseudomyxoma and is based on the identification of three lesions: micinous ascites and its characteristics, peritoneal nodular implants and the primary tumour, which is only exceptionally visualised.

3.1 Ultrasonography

Mucinous ascites is typically heterogeneous, hypoechoic, not very mobile with more or less stratified content; it can be partitioned with multiple septa and fine septal calcifications.

3.2 Abdominal CT Scan

Mucinous ascites of the pseudomyxoma is hypodense, often discreetly more hyperdense than a simple transsudate. It can be partitioned and contain fine curvilinear calcifications [1].

A fairly specific feature of hepatic scalloping is the discrepancy between the presence of numerous or bulky peripheral hypodense lesions and the absence of centrohepatic hypodense lesions at a distance from the capsule.

Peritoneal implants are visible as heterogeneous nodules, which can be raised after injection of contrast material [1].

PMR may also appear as parietal calcifications. An abdominal CT scan can also help to identify the primary pathology in the appendix by demonstrating the thickening of its wall and its retrocecal position.

The cystic nature of a PRM is best demonstrated by its classical signal characteristic of hypo density in T1 and hyperintensity T2 on MRI [2].

The diagnosis of PMR has been largely confused with psoas muscle abscess [3], the latter is usually associated with spondylodiscitis of adjacent vertebrae, and has a homogeneous peripheral elevation.

The absence of vertebral damage, with morphological signs of erasure, the absence of psoas muscle invasion and the improvement of irregular internal partitioning within the collection, ruled out the possibility of a psoas abscess and favoured the diagnosis of PMR [4].

The treatment must meet two objectives: A complete removal of the initial tumour and a thorough cleaning of the cavity. The laparoscopic approach is not recommended [5].

Treatment of unruptured appendicular mucocele is surgical, preferably by laparotomy rather than laparoscopy; in the case of laparoscopy, a collection bag must be used to avoid peritoneal dissemination. Appendectomy is performed without rupture of the appendix with complete removal of the meso appendicular and sampling for cytology of the peritoneal fluid.

Depending on the result of the cytology, possible rupture of the mucocele and involvement of the meso-appendix, further treatment may be carried out: right hemicolectomy, tumour cytoreduction, intraperitoneal chemotherapy.

The treatment of peritoneal pseudomyxoma is similar regardless of the form: Tumour reduction surgery consisting of the removal of the maximum amount of mucus and tumour formations with omentectomy, localised peritonectomies, right hemicolectomy and bilateral oophorectomy in women, combined with postoperative intraperitoneal chemotherapy (CHIP) or immediate postoperative intraperitoneal chemotherapy, which improves the prognosis by acting on the microscopic residual disease [1].

4. CONCLUSION

In summary, the clinician must be attentive to the rare Possibility of retroperitoneal pseudomyxoma. A thorough physical examination should be carried out, especially in the inguinal region, to look for extra-pelvic extensions of such retroperitoneal diseases, which can serve as clues,to the underlying disease.

Surgical treatment involves an elective approach to the retroperitoneal cavity to evacuate the gelatinous substance and clean the cavity, followed by an intraperitoneal approach, allowing the removal of the causal lesion, while avoiding peritoneal contamination by inoculation of mucous cells [6].

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Fairise A, Barbary C, Dercelle AL, Tissier S, Granger P, Marchal F, Laurent V, Régent D. Appendicular mucocele and peritoneal pseudomyxoma. J Radiol. 2008; 89:751-62.
2. Roumina Hasan, Sandeep Kumar, Anuradha Ck. Rao, Rajagopal Kadavigere. Rare presentation of pseudomyxoma retroperitonei: Stretching the limits. Mal J Med Sci. 2016;23(4).
3. Edrees WK, Hannon RJ. Pseudomyxoma extraperitonei: A rare presentation mimicking an iliacus abscess. Ulster Med J. 1999;68:38-39.
4. Kharrat Koubaa1 M, Affes N, Kharrat M, Ben Amar M, Kannous M, Kechaou MS, Jlidi R, Beirut MI. Voluminous pseudomyxoma of the psoas secondary to a mucinous cystadenoma of the appendix. 1 Service of Visceral Surgery, Habib-Bourghiba University Hospital Center, Sfax, Tunisia; 2001.
5. Gonzalez Moreno S, Shmookler BM, Sugarbaker PH. Appendicealmucocele contraindication to laparoscopic appendectomy. Surg Endosc. 1998;12: 1177-9.
6. Hasan Ediz Sikar1, Kenan Çetin1, Ersin Gündoğan1, Gökçen Alinak Gündoğan2 and Levent Kaptanoğlu1. Retroperitoneal mucinous cystadenoma of the appendix mimicking hydatid cyst: A case report. Molecular and Clinical Oncology. 2016;5: 345-347.

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