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A Case Report about a Mesenteric Desmoid Tumor Mimicking an Appendicular Abscess: A Rare Complication of a Rare Mesenteric Mass

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Abstract: Mesenteric desmoid tumor is an uncommon neoplasm. It is usually asymptomatic or diagnosed as a painful mass. Typically, it presents as a solid soft-tissue mass on cross-sectional imaging. Complications such as infection remains exceptional. Through this manuscript, we report the case of an acute appendicitis fistulized in a mesenteric desmoid tumor, mimicking an appendicular abscess. We report such an unusual case with clinicopathological correlation.

Introduction:

Desmoid tumors are an uncommon benign neoplasm. They can arise at various sites, including the extremities, trunk, and abdomen. Mesenteric location is extremely rare, usually associated with familial adenomatous polyposis. Most often, they are slow growing tumors and remains a long time asymptomatic. Acute manifestations are rare and are due to infection, rupture or compression of neighboring organs. In this manuscript, we report an acute revelation of a mesenteric desmoid tumor due to the fistulization of an acute appendicitis in a desmoid tumor and we will discuss its clinical features, imaging finding, treatment options and outcomes.

Case description

A 31-year-old man, with an unremarkable medical history, was admitted to the emergency department for the management of an acute right iliac fossa pain that started since five days. At his admission, the patient was stable and alert. His temperature was high (38.5 Celsius). His blood arterial pression (120/80 mm Hg), heart rate (90 pulses per minute) and respiratory rate (12 cycles per minute) were normal.

Physical examination found a defense at the right iliac fossa. Deep abdominal palpation found a painful mass. Thoracic, cardiovascular and neurological examination were normal.

Laboratory tests revealed high levels of C reactive protein (145mg/l) and white blood cell account (16000e/mm).

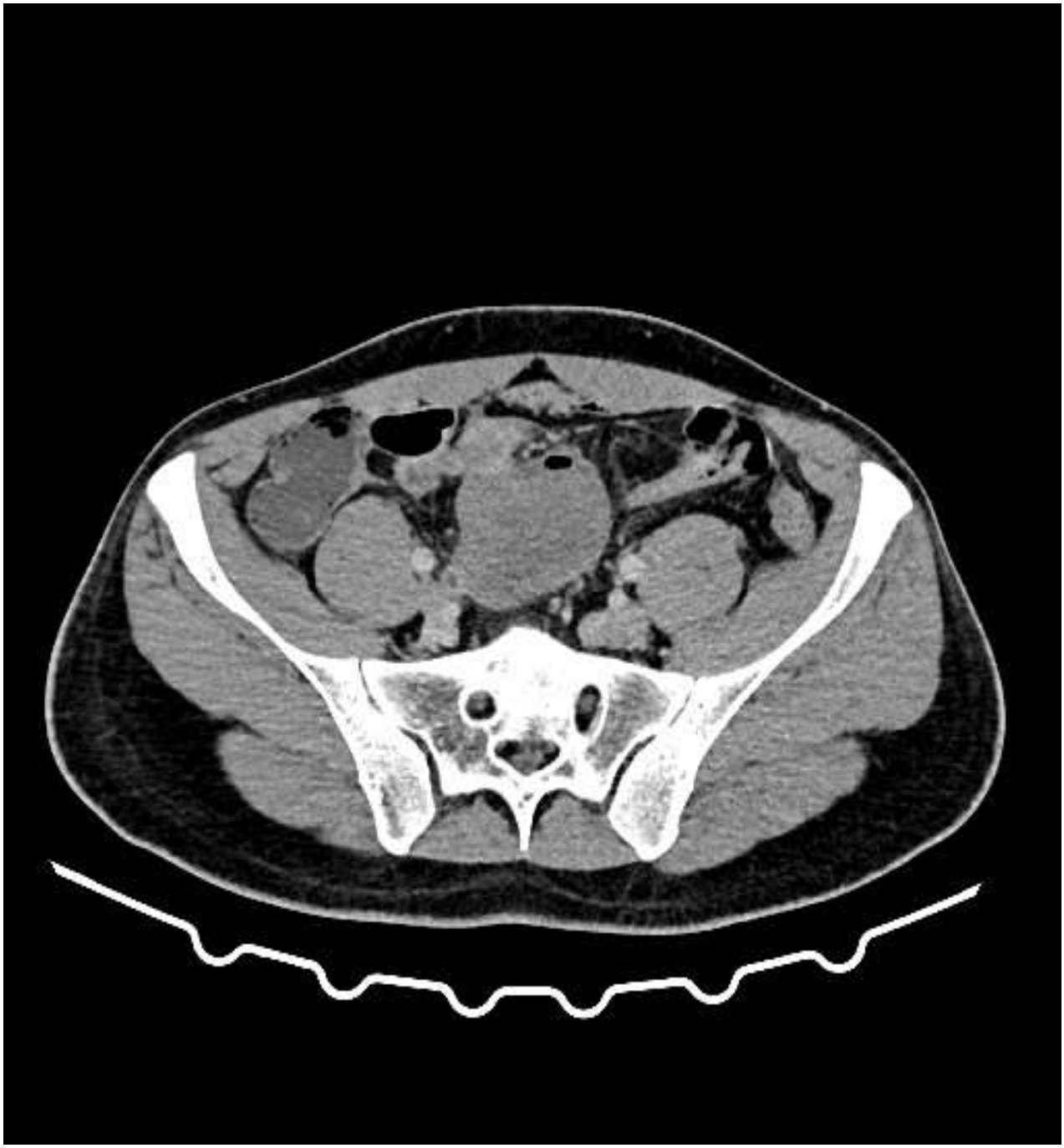
The patient first underwent an abdominal ultrasound examination, which revealed a swollen appendix communicating with a very hypoechoic round formation that was diagnosed as an abscess.

An enhanced abdominal computed tomography scanner was then performed for a better assessment of the abscess and its associated complications. It showed a swollen appendix communicating with a round formation, non-enhanced after iodine contrast injection, containing gas bubbles. Thus, the final radiological diagnostic was an appendicular abscess.

The patient underwent a surgical treatment. After laparotomy, we found a swollen appendix communicating with a solid mass mimicking the abscess. An appendicectomy and biopsy of the mass were performed. Pathological analysis revealed a fibrous tumor that expressed beta-catenin with nuclear labeling and did not express CD117, PS100, AML, CD3 and Desmin.

Histological and immunohistochemical aspects suggest a desmoid tumor.

The diagnosis was then confirmed by molecular biology by demonstrating the mutation of exon 3 of the beta-catenin gene.



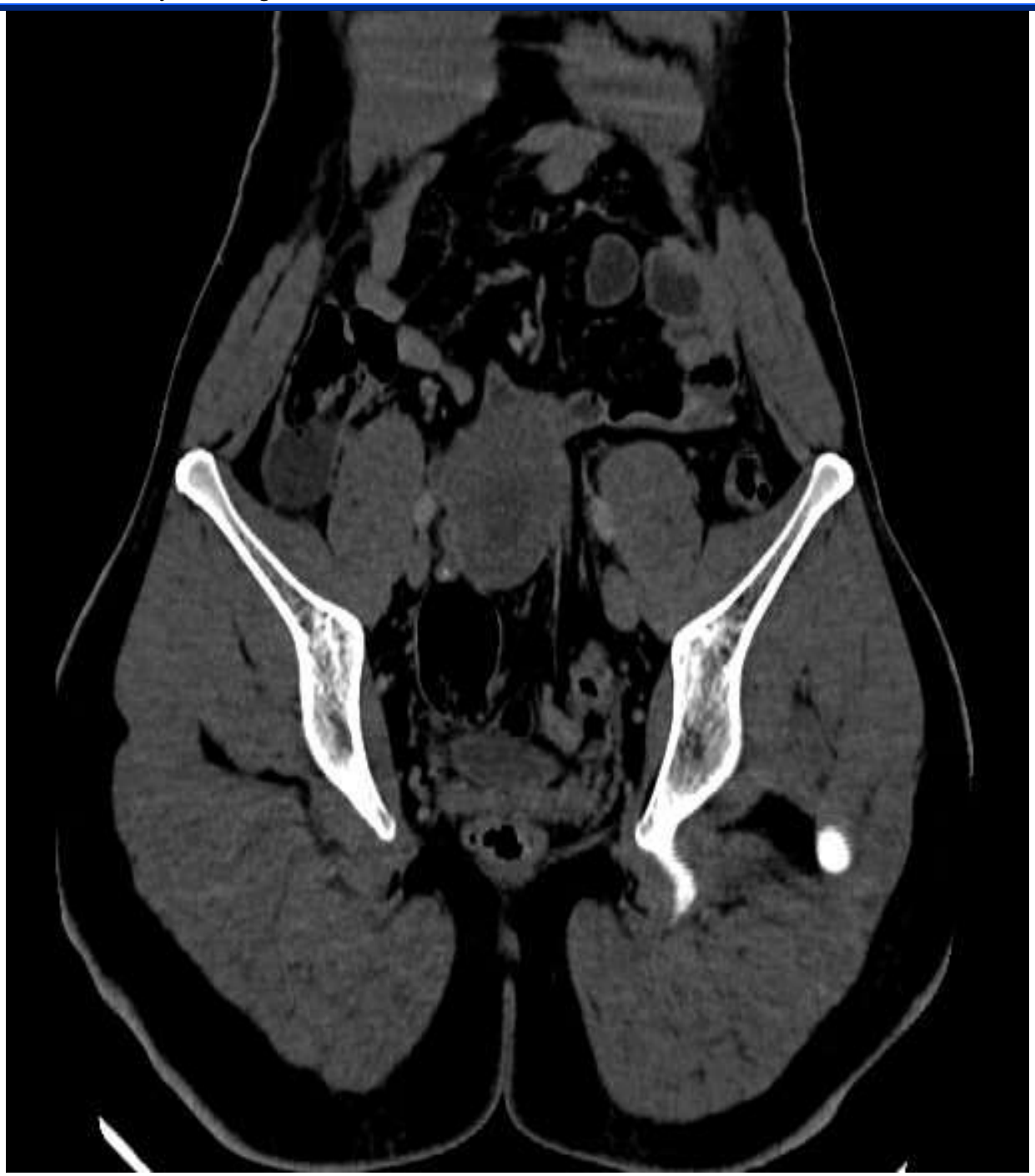


Figure 1 and 2: An injected CT scan which showed the presence of an oval mass in the hypogastric region, well limited, with regular contours, heterogeneously enhanced after injection of the contrast product. This mass was in intimate contact with the last ileal loop, as well as with the appendix which was swollen.

Discussion

Desmoid tumors are rare and represent less than 0.03% of all tumors and 3.5% of fibrous tumors [5]. It can be located either in the aponeuroses of the peripheral muscles (45% of cases), or in the muscles of the abdominal wall (45% of cases) or more rarely in the mesenteric or retroperitoneal regions (10% of cases) [6]. Indeed, it most often affects the mesentery of the small intestine [5], rarely the iliocolic mesentery, the greater omentum and the retroperitoneum [7, 6].

Most mesenteric desmoid tumors are sporadic, 10% of them are part of the Gardner syndrome [6], associated with familial polyposis coli by mutation of the APC gene [8]. In this case, these tumors are more aggressive, responsible for 11% of cases of death [2, 5]. Its etiopathogenesis is not clear. It could result from the sum of three factors: an initiation of cell proliferation, possibly following a trauma [9, 10]; a promoting effect of sex steroids: supported by their predominance in genitally active women, the possibility of aggravation during pregnancies and spontaneous regression at menopause, as well as the frequent presence of hormone receptors explaining its possible hormone-sensitivity [4, 9, 11]; a particular genetic terrain marked by a disorder of the regulation of fibroblastic growth [9].

They are more frequent in young adults, with a peak in frequency at the age of 30, and affect both men and women [2, 6]. Their most frequent mode of revelation is a painless abdominal solid mass when their size is small. [5]. Indeed, they often remain asymptomatic until they reach a significant size [2, 8], leading to complications such as: occlusion, ischemia or intestinal perforation, hydronephrosis, digestive fistula, deep vein thrombosis, neurological compression and alteration of the ileal function [5, 12].

The ultrasound characteristics of the tumor are aspecific depending on its collagen and fibroblast content and its vascularization [7]. CT is the first-line imaging tool, it allows to characterize the tumor, to guide biopsies [2, 7] and to establish poor prognostic criteria such as: diameter < 10 cm, multiple mesenteric locations, extensive involvement of the small intestine or the superior mesenteric artery, bilateral hydronephrosis [5]. Typically, it presents as a well-circumscribed mass attached to the small intestine, sometimes ill-defined and irregular testifying to its infiltrative nature. Its appearance is hyperdense compared to the muscle on the non-enhanced CT scan with variable and heterogeneous contrast enhancement on the injected CT scan. MRI is especially recommended in the follow-up [2, 7], since its better resolution allows to differentiate post-therapeutic changes, from a recurrence and to predict tumor growth [5]. Its appearance is often heterogeneous, with an intermediate or hypo global signal in T1 and a mixed signal in T2 [13]. As for colonoscopy, it should be performed as a matter of principle in order to eliminate the rare but possible association with colonic polyposis [9].

Histologically: the lesion is poorly limited and infiltrates the adjacent tissues. The proliferation is made of elongated, spindle-shaped cells, small in size, with a small nucleus, without atypia. It binds Vimentin and Muscle Actin [14].

Wide surgical excision is the first-line treatment of mesenteric desmoid tumors [2]. However, these tumors pose the problem of resectability on the one hand, by their intra-mesenteric localization, notably their contact with the mesenteric vessels [5], and on the other hand by the infiltration of the neighboring organs [14]. Consequently, the majority of cases require resection of the affected intestinal segment [2], especially since the high risk of recurrence in these locations (50% to 80% of cases) [5], depends above all on the quality of the resection margins (27% recurrence if the margin is histologically healthy, compared with 54% if the margin is invaded).

The frequency of local relapse and the difficulty of managing advanced forms have led to the initiation of adjuvant treatments [15]: radiotherapy is indicated in cases of incomplete resection, inoperable tumor or relapse. However, its digestive toxicity, and its potentially carcinogenic effect make its use delicate [5, 11, 14]; anti-hormonal treatment (Tamoxifen 20mg/day) alone or in combination with a non-steroidal anti-inflammatory (Indometacin: 75mg/day) allows long stabilizations and an objective response in more than 50% of cases. In case of failure or relapse, a second line of hormonal therapy (toremifene, aromatase inhibitors, and gonadotropin derivatives) may be effective; multidrug therapies are readily proposed to rapidly progressive, or resistant tumors [11]. In fact, the combination of low doses of methotrexate and vinblastine is an effective treatment in 70% of cases [5].

The prognosis is good after complete excision. However, in the event of local recurrence, iterative surgical treatment is associated with significant morbidity and mortality. Indeed, the strong infiltration of the abdominal viscera represents the ultimate cause of death after several years [7].

Conclusion

To conclude, we report the first case of an infected mesenteric desmoid appearing as an appendicular abscess on both ultrasound and CT, and illustrate this rare and perhaps misleading mimicry.

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