Solitary Fibrous Tumour of the Mesorectum: About a Case with Review of the Literature

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Abstract

Solitary Fibrous Tumours (SFTs) are rare entities that are most commonly seen in the pleura. However, SFTs could affect different organs and tissues. Pelvic SFTs are rarely reported in the literature and para-rectal ones are even rarer. We present here a case of a 43-year-old lady with continuous nonspecific pelvic pain and fullness. Para clinical investigations were consistent with a well-demarcated mass in the perirectal region. Despite some radiologic features that were consistent with a GIST, however the diagnosis was unsure. After successful laparoscopic resection, histopathology was consistent with SFT.

Keywords: Spindle cell tumors; Solitary fibrous tumours

Introduction

SFT can be seen in different locations throughout the human body, that could extend from the Head and neck [1] to the ischio-anal fossa [2,3]. These spindle cell tumours were reported firstly in the pleura in 1931 [4]; extrathoracic SFTs were identified later on [1,5,6]. Intrathoracic and extrathoracic SFTs could metastasize or recur. Pelvic SFTs including anorectal ones, are very rare. Preoperative diagnosis is difficult, and only in few cases paraclinical investigations have led to accurate diagnosis [7-9].

Case Presentation

A 43-year-old lady presented with six months history of pelvic pain and heaviness. Axial T2 weighted MRI showed a well demarcated left Para-rectal mass about 5 cm × 4.5 cm, 12 cm from the anal verge (Figure 1). There were no signs of necrosis or local extension. Thoracoabdominal CT-scan showed no evidence of metastasis. Endoscopic ultrasound showed an oval shaped mass developing just beneath the muscular layer of the rectum. The FNA analysis was not contributive.

After discussing the case during the Digestive Oncology meeting, we decided to perform an exploratory laparoscopy with an attempt to excise the mass. At laparoscopy, the mass was situated in the sub peritoneal space, just at the level of the cul de sac of Douglas (Figure 2). It was just below the peritoneal reflection, at the level of the upper part of the rectum, situated at the left side of the rectum. Laparoscopic excision was done successfully and sent for pathology analysis. The rectum was kept intact. On gross pathology, the tumor sized 4 x 2 x 2 cm, of white colour with some...
areas of calcifications. Histopathologically, it consisted of spindle cells, arranged arbitrarily within hypocellular and hypercellular areas (Figure 3). Vascularization was abundant with hemangiopericytoma features. Cells showed poor mitotic activity. Resection margins were free of disease. The post-operative course was uneventful and the patient is asymptomatic after one year of follow-up.

Discussion

SFTs were first described in the thoracic pleura in 1931 [4]; however many other sites have been reported in the literature. SFTs are rare, with an incidence of 0.2/100000/year [9]. Pelvic SFTs are even rarer, with only about 34 cases available on PubMed database (using the search terms “solitary fibrous tumour”, “pelvis”) [2,3,7-25]. The 1st reported case of pelvic SFT occurred in the bladder and was published in 1997 [7]. Other pelvic structures included: Seminal vesicles [1], mesorectum [19], perianal space [2,3,8] and other non-well specified pelvic areas [13-18]. Radiologic findings are non specific for GIST [18], and they are often misdiagnosed with GIST.

Accurate preoperative diagnosis was evoked in 3 reported cases, where biopsy was performed in 2 cases [7,8], and [7] F-FDG PET/CT with ultrasound guided biopsy were performed in one case [9]. Some authors have described obstructive symptoms, such as polakkuria [22], urinary retention [8] and constipation [19]. It’s Noteworthy to mention that in 3 reported cases, hypoglycaemia was the alarming symptoms and was secondary to IGF-2 secreting cells. SFTs generally show benign behaviour. A recent large retrospective study, where 110 SFT cases were reviewed, 10 year metastasis free rate was 55% [26]. Predictive signs of malignity include: necrosis, high mitotic activity, cellular atypia and large tumors. Even though, the absence of these features would not preclude metastasis or recurrence [24]. Surgery with curative intent is the best treatment option [26]. While Liver SFTs are treated with 1 cm negative margin resection [27], surgery with curative intent would be also the best treatment option for pelvic SFTs [2,3,7-25,27].

In some cases, neoadjuvant chemoradioterapy preceded surgical resection. To the best of our Knowledge, none of the previously reported anorectal cases were treated laparoscopically. In one case, excision was carried out during laparotomy, and in another one trans-sacral resection was adopted. Yan et al. [11] described recently FDG PET/CT utility in determining the metabolic activity of such tumors, and subsequently what treatment regimen should be used [9]. Doxorubicin may also be proposed as adjuvant therapy [9,25]

Conclusion

Only 3 cases of anorectal SFTs were previously reported, one in the pararectal area [19], one in the ischioanal fossa [3] and one with perianal location [2].

Biopsy is not always diagnostic in SFTs, because spindle cells are arranged haphazardly within the tumor bulk [13]. Since recurrence and metastasis are of possibility with SFTs, even in the absence of aggressive signs, thus surgical excision should be mandatory in order to establish the diagnosis, and to assess the degree of cellular atypia and mitotic index.

References

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