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A rare case of leiomyoma of the internal anal sphincter

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ABSTRACT

INTRODUCTION: Leiomyoma is a benign tumour which derives from the smooth muscle fibres and it may occurs in every site in which this type of muscle is present. Among all benign soft tissue tumours it represents almost 3.8% and its pathogenesis remains still unknown.
PRESENTATION OF CASE: The present case is about a 62 year old woman referred to our centre complaining anal and perineal pain which increase after defecation in association with the appearance of a nodule in the perianal region fixed to the anal sphincter. A 360° tridimensional transanal ultrasound was performed and it showed an anterior nodular thickening of the internal anal sphincter. After an inconclusive preoperative biopsy and a counselling with the patient, the surgeons decided to proceed with the surgical excision. The immunohistochemical examination confirmed the preoperative suspicion of leiomyoma. At 1 year follow-up the patient had not tumour-related symptoms or fecal incontinence and any signs of local recurrence at ultrasound imaging were demonstrated.
DISCUSSION: Leiomyomas are relatively insensitive to chemotherapy whereby surgery is the treatment of choice and it should be adequate to the site and dimension of the lesion achieving a complete resection with free margins. A further close follow-up is needed too.
CONCLUSION: Nowadays there is not a gold standard technique to treat such kind of lesions and the decision of the best surgical approach should depend on the dimension and site. In fact, surgery aims to the oncological outcome trying also to minimize the possible post-operative functional complications.

1. Introduction

Leiomyoma is a benign tumour which derives from the smooth muscle fibres and it may occurs in every site in which this type of muscle is present. Among all benign soft tissue tumours it represents almost 3.8% and its pathogenesis remains still unknown [1]. One of the most frequent sites in which this tumour develops is the gastrointestinal tract with an incidence of 7% [2], and the organs commonly involved are the stomach and small bowel. Less usual sites are oesophagus, colon and anorectal localization which is fairly rare with an incidence approximately of 1 on 2000 overall rectal tumours [3,4]. Due to this exceptional presentation we report a case of leiomyoma arising from the internal anal sphincter.

2. Presentation of case

62 year old woman referred to our centre complaining anal and perineal pain which increase after defecation in association with the appearance of a nodule in the perianal region. The tumefaction had progressively growth in the previous months causing symptoms worsening. The physical examination confirmed the presence of an anterior nodule that seemed to be fixed to the sphincter. The anoscopy and vulvo-vaginoscopy did not revealed any alteration of the mucosa. A 360° tridimensional transanal ultrasound was performed and it showed an anterior nodular thickening of the internal anal sphincter. Its dimensions were 2 cm width and 2 cm length in the anal canal (Fig. 1A). After an inconclusive preoperative biopsy and a counselling with the patient, the surgeons decided to proceed with the surgical excision. A perianal linear incision and blunt dissection in the intersphincteric space was performed identifying the lesion (Fig. 2A). After its complete excision a considerable thinning of the internal anal sphincter was detected so that a side-to-side sphincteroplasty was carried out (Fig. 2B). The specimen was 2 cm × 2.8 cm (Fig. 2C) and a cross section of the lesion showed a typical fibrous capsule. The histo-pathological examination through immunohistochemistry, investigating the expression of actin, desmin, CD34 and CD117 (Fig. 3), confirmed the preoperative suspicion of leiomyoma. The hospital stay was uneventful and the patient was discharged on day three. At 1 year follow-up the patient had not tumour-related symptoms or fecal incontinence and any signs of local recurrence at ultrasound imaging were demonstrated (Fig. 1B).

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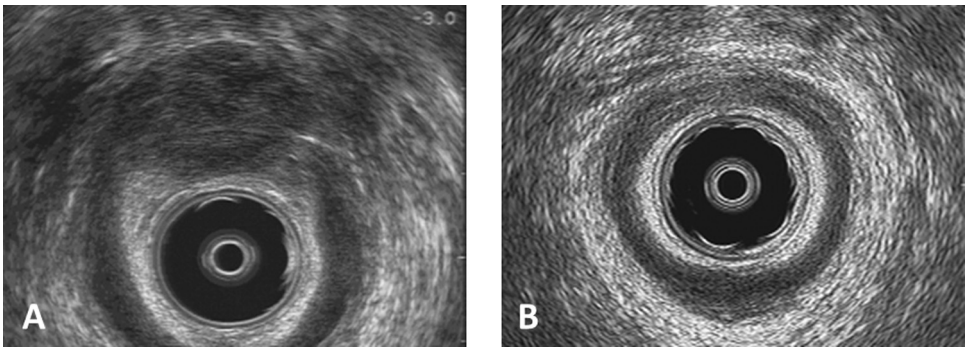

Fig. 1. Preoperative 360° transanal ultrasound (A). Postoperative 360° transanal ultrasound (B).

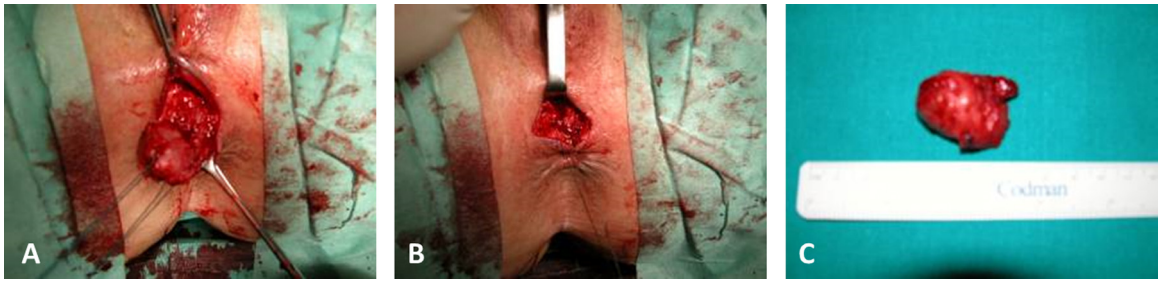

Fig. 2. Surgical excision (A). Spincteroplasty (B). Specimen (C).

3. Discussion

The first description of leiomyoma was made by Virchow in 1854 defining it as a benign tumour of mesenchimal origin arising from smooth muscle fibres. In the previous classification it was considered under the so-called gastro-intestinal stromal tumour (GIST). Nowadays the GIST belongs to the soft tissue tumours, as well as the leiomyoma, but it has an own identity based on a specific immunohistochemical pattern, whereby many tumours previously defined as leiomyoma are now classified as GIST which have even a different type of treatment [5]. The leiomyoma can develop in any site in which smooth muscle is present. It is classified as superficial or deep. The latter is further divided in somatic and retroperitoneal. The superficial variant usually affect the extremities with the same incidence in both sexes whereas the retroperitoneal generally involves the pelvic region in peri-menopausal women [6]. In the gastrointestinal tract the organs mainly involved are the stomach and small bowel, less frequent sites are oesophagus, colon and anorectal localization which is fairly uncommon [7]. In fact, Hatch et al. analysed all the cases of anorectal smooth muscle tumours described in literature from 1881 to 1996. This review included 432 cases of leiomyomas and 480 cases of leiomyosarcomas. They usually occur between 40 and 59 year, mostly in male [8]. Although these neoplasms may arise from the muscularis mucosae or the smooth muscle cells of the vessels, they usually derived from the longitudinal and circular layers of the gut. These tumours are divided by their type of growth into three variants: intraluminal, extraluminal and intramural. Intraluminal leiomyomas are usually sited in the posterior wall of the distal part of the rectum and they may be sessile or peduncolated. On the other hand, extraluminal leiomyomas generally grow from the colonic wall inside the abdomen and they often mimic a GIST [1]. They have the same estrogenic and progestinic receptors expression of the uterine leiomyomas [9,10]. Sometimes the tumours grow in both direction forming an "hour glass" [11]. The symptoms related to the presence of leiomyoma vary widely. The superficial localization is usually asymptomatic and it does not reach great dimensions because it is quickly identified because of the appearance of a nodule, while the deep variant tend commony to remain asymptomatic until it reaches considerable dimension. At that moment the symptoms may be pain, rectorrhagia, tenesmus or bowel transit alteration because of which the patient looks for medical examination. The ulceration of the overlaying mucosa is rare and described in both types, leiomyoma and leiomyosarcoma [12].

The radiological imaging, such as magnetic resonance or 360° tridimensional transrectal ultrasound, are useful to identify the mass, its precise localization, its relationship with the adjacent structures such as anal sphincter or uro-gynaecological structures and it steers the operative strategy.

Although leiomyomas may rarely have cellular atypia, they show a low mitotic cellular index, while leiomyosarcomas have a high nuclear pleomorphism and a high mitotic index. The differential diagnosis between these two tumours may be difficult [10]. Witzigmann et al. demonstrate that the prognosis of rectal leiomyosarcoma is poor, with a survival rate at 5 years of 20–25%, and that almost 80% of leiomyosarcomas have local recurrence [3]. Conversely, leiomyoma has a good prognosis without any described recurrences [13]. Histological feature of leiomyoma are the presence of spindle cells arranged in bundles and foci of dystrophic calcification are commonly present. The immunohistochemistry is fundamental to identify these tumours because they are positive for actin and desmin and negative for CD34 and CD117 (typically expressed by the GIST).

Unfortunately, it still lacks precise parameters to identify preoperatively the malignancy of such masses causing difficulties to the surgeon to choose the best operation to perform [3]. In fact, preoperative histological diagnosis is adequate only in 29% of cases and it needs an expert pathologist. The definitive diagnosis is based on the histological examination in which defined parameters are evaluated including the size of the tumour, the number of mitoses per field, the number of areas of necrosis and nuclear pleomorphism [13].

Since these tumours are relatively insensitive to chemotherapy, surgery is the first treatment and it should be adequate to the site