Removal of a large bizarre uterine leiomyoma by operative hysteroscopy. Case report and review of the literature

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Summary

Background: Atypical leiomyomas are relatively uncommon in the general practice of gynaecology. We present a case of a large uterine bizarre leiomyoma removed by operative hysteroscopy and review of the literature. *Case:* The patient, a 49-year-old, gravida 3, para 3, perimenopausal Greek woman presented to our Department because of dysmenorrhea and abnormal vaginal bleeding. She underwent hysteroscopy in which a large submucosal leiomyoma was detected and entirely removed in one session. The histopathology revealed bizarre uterine leiomyoma. *Discussion:* There is no evidence to indicate that hysterectomy is necessary, if the diagnosis of atypical leiomyoma has been firmly established.

Key words: Atypical leiomyoma; Hysteroscopy; Bizarre leiomyoma.

Introduction

In the current World Health Organization (WHO) classification, leiomyoma with bizarre nuclei (LBN) is grouped under the heading 'atypical leiomyoma' [1]. An atypical leiomyoma by definition is a smooth muscle tumor that shows cellular atypia and may sometimes have increased mitotic rates. Once regarded as in-situ leiomyosarcoma, [2] this tumor is now generally accepted as a clinically benign variant of leiomyoma, despite microscopic features of malignancy [3, 4]. In the earlier edition of the WHO classification, LBN was defined as 'a leiomyoma containing giant cells with pleomorphic nuclei with little or no mitotic activity' [5]. Symplastic leiomyoma, bizarre leiomyoma, and pleomorphic leiomyoma are older synonyms for atypical leiomyoma. We present a case of an atypical uterine leiomyoma which was diagnosed and removed by operative hysteroscopy in our department, together with a review of the literature.

Case Report

The patient, a 49-year-old, gravida 3, para 3 perimenopausal Greek woman presented with a complaint of dysmenorrhea and episodes of menometrorrhagia during the last six months. Her past medical and surgical histories were unremarkable. For the last three months the patient had received gonadotropin-releasing hormone agonists. The gynaecologic examination was normal and Papanicolaou examination of cervical, vaginal smears was also normal. Transvaginal ultrasound (TVS) revealed a large submucosal leiomyoma at the fundus of the uterine cavity. The endometrium had also a width of 16.8 mm, with multiple blood vessels at the Doppler examination.

Hysteroscopy was then performed using saline infusion as a distension medium. A large, submucosal uterine leiomyoma 4 x 5 cm, that filled up all the uterine cavity, was detected. Myomectomy by operative hysteroscopic resection and uterine curettage under general anaesthesia were undertaken. The mass was entirely removed and the specimens were sent for histological examination. The tumour had a solid, soft, multinodular, yellow-tan surface, but without any other signs to the naked eye that may have helped in the differential diagnosis from the usual type of leiomyoma.

Histologic evaluation of the specimens revealed an atypical uterine leiomyoma. Large cells with eosinophilic cytoplasm and bizarrely shaped, multilobated, hyperchromatic nuclei, that characterize this tumor, were present. There was no mitotic activity or necrosis. Follow-up six months after surgery with TVS showed no evidence of recurrence.

Discussion

The aetiology and pathogenesis of atypical leiomyomas remain unclear. The question why bizarre cells develop in these leiomyomas has not yet been answered [6]. A few cases have occurred in patients receiving synthetic progestins [7, 8], but a causal relationship has not been proven. Symplastic-type giant cells often have been attributed to degeneration, a theory supported by the cytologic features of some of the bizarre cells and by at least one ultrastructural study [9]. However, the cause for such degeneration is not apparent. There is also no known relationship of bizarre cells in leiomyomas to preoperative – as in our case – treatment with gonadotropin-releasing hormone agonists, such as leuprolide acetate [10].

The main symptoms and clinical findings in cases of atypical leiomyomas are irregular vaginal bleeding, pain and pelvic tumour [10]. Most LBNs are less than 5.5 cm in diameter [6]. This contrasts with the relatively large size of most leiomyosarcomas in which the mean maximum dimension is about 10 cm [11, 12]. In our case the patient presented because of menometrorrhagia and dysmenorrhea and the tumour was less than 5 cm in maximum diameter.

Revised manuscript accepted for publication February 10, 2011

Fig. 1

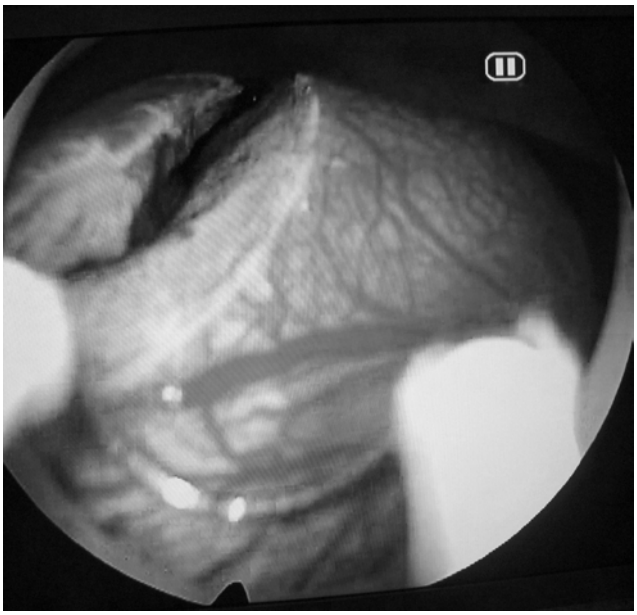

Fig. 2

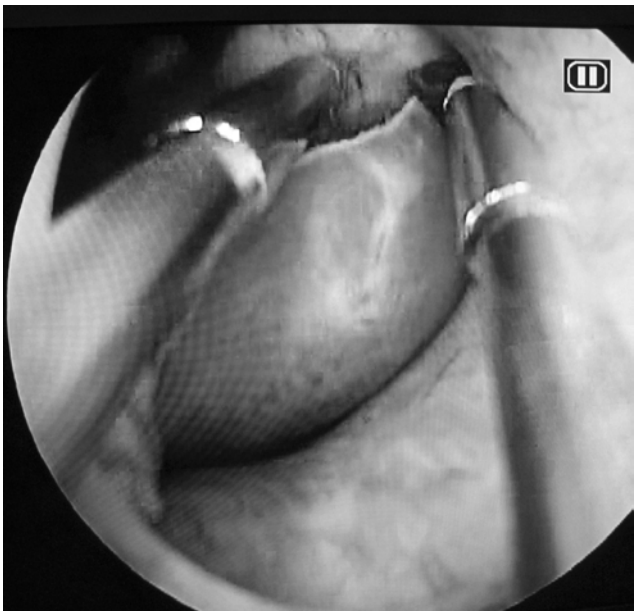

Fig. 3

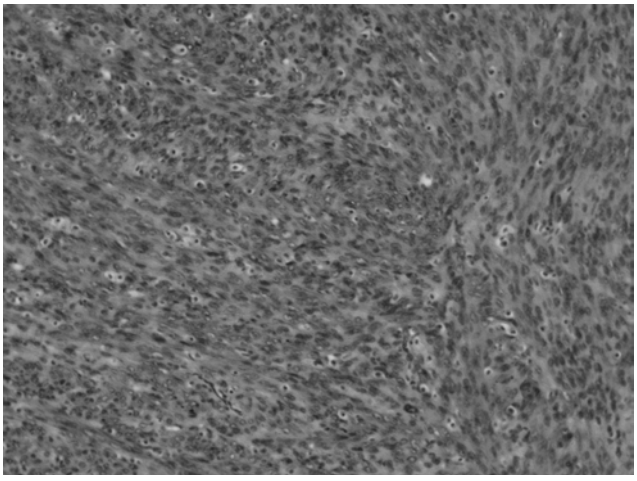

Figure 1. — Hysteroscopic view of a large submucosal leiomyoma with multiple branching vessels.

Figure 2. — The same leiomyoma during the process of hysteroscopic removal.

Figure 3. — Histological section of uterine tumour showing morphology consisting of atypical (bizarre) leiomyoma (haematoxylin – eosin x 25).

The gross appearance of atypical leiomyomas usually resembles that of typical leiomyomas, but yellow to tan areas, softening, and cysts are seen in a minority of cases. The cardinal microscopic feature is the presence of large cells with eosinophilic cytoplasm and bizarrely shaped, multilobated or multinucleated, hyperchromatic, often 'smudged' nuclei, but an absence of other worrisome histological features. The nuclei often contain cytoplasmic pseudoinclusions. In our specimens these large bizarre, pleomorphic tumour cells with atypical nuclei and eosinophilic cytoplasm were present.

LBNs are reported to be clinically benign. The major criteria of malignancy are mitotic index, nuclear atypia and necrosis [13, 14]. Mitotic activity is low compared to leiomyosarcomas. Downes and Hart studied 24 LBNs and found 0-7 mitotic figures (MFs) per 10 high-power fields (HPFs) using the highest count method (mean 1.6 MFs/10HPFs), or 0-2.8 MFs/10HPFs using the average count method (mean 0.8 MFs/10HPFs). Although one of the tumors had 7 MFs/10HPFs, none of them recurred after a mean follow-up of 11.2 years [6]. In our case there was no mitotic activity or necrosis and no evidence of recurrence has been detected during this six-month follow-up period.

When an atypical leiomyoma is discovered in a hysterectomy specimen, no further treatment is required. Despite its good reputation, cases of recurrence following conservative surgical treatment for atypical leiomyoma with myomectomy have been reported. With an increasing number of patients being treated by myomectomy, however, the frequency of LBNs in myomectomy specimens will undoubtedly increase. Although the number of reported cases in which myomectomy only was performed and long-term follow-up remains small, there is currently no evidence to support that hysterectomy is necessary, once the diagnosis of LBN has been firmly established. However a much more cautious approach must be adopted in the management of atypical

leiomyoma following myomectomy, especially when the atypia is diffuse or the tumour is large, by recommending follow-up with non-invasive imaging in order to exclude regrowth in the first several years.

Hysteroscopic diagnosis and treatment of atypical submucosal uterine leiomyomas is a safe minimally invasive surgery method. Hysteroscopy does not increase the risk of microscopic intraperitoneal spread even in cases with malignancy compared with diagnosis by curettage or endometrial biopsy. It is a method of diagnosis and treatment under direct view, with a high therapeutic outcome and a mean hospitalisation time of less than eight hours.

Conclusions

Atypical uterine leiomyoma is a challenging diagnosis. Careful evaluation of the microscopic features will assist the pathologist in determining the benign nature of the neoplasm. There is no evidence to indicate that additional therapy is needed once the diagnosis of atypical leiomyoma in myomectomy specimen has been firmly established.

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