Case Report

Possible progression of an atypical leiomyoma to a leiomyosarcoma: a case report

Ekanayake CD[1], Herath R[2], Fernando S[3], Mahendra G[4], Balawardene J[5], De Silva A[6]

1 Department of Clinical Sciences, Faculty of Medicine, General Sir John Kotelawala Defence University, Ratmalana, Sri Lanka. +94713283668, cdekanayake2000@yahoo.co.uk

2 Department of Obstetrics & Gynaecology, Faculty of Medicine, University of Kelaniya, Sri Lanka. +94112961000, rasikaherath75@yahoo.com

3 Department of Obstetrics & Gynaecology, Faculty of Medicine, University of Kelaniya, Sri Lanka. +94112961000, sunilfer123@gmail.com

4 Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka. +94718366190, baggmahendra@yahoo.co.uk

5 Department of Clinical Sciences, Faculty of Medicine, General Sir John Kotelawala Defence University, Ratmalana, Sri Lanka. +94710219271, jaybalaya@gmail.com

6 Department of Obstetrics & Gynaecology, Faculty of Medicine, University of Kelaniya, Sri Lanka. +94112961000, basanthadesilva@gmail.com

***Corresponding Author** - Dr. CD Ekanayake Email: cdekanayake2000@yahoo.co.uk*

Keywords

smooth muscle cell tumour, atypical leiomyoma, leiomyosarcoma, smooth muscle tumour of uncertain malignant potential, case report

Introduction

The spectrum of uterine smooth muscle cell tumours (SMT) ranges from leiomyomas to leiomyosarcomas (LMS). Atypical leiomyomas (AL) are a uterine SMT of a contentious nature due to its indeterminate malignant potential. Although the risk of recurrence is low there is limited experience on management. On the other hand, it is generally believed that the uterine LMS arises de novo, rather than from a precursor lesion.[1]

Case report

A 41-year-old woman was found to have an anterior cervical fibroid (8cm diameter) for which a myomectomy was done. Microscopy showed a SMT with diffusely scattered atypical cells (Figure 1A). Maximum mitotic activity (MA) was 5 mitotic figures (MF) per 10 high-power-fields (HPF). Extensive sampling did not reveal coagulative tumour cell necrosis (CN). A diagnosis of an AL was made with a decision to follow-up.

Two and a half years later she developed heavy menstrual bleeding. Ultrasound imaging revealed another fibroid in the anterior region. She underwent a total abdominal hysterectomy with ovarian conservation. The specimen showed a well-defined myometrial nodule (7cm diameter) compressing the cervix. Microscopically this was a SMT with diffusely scatted atypical cells with bizarre nuclei, high MA (11-12 MF/10HPF) and focal CN confirming a leiomyosarcoma (Figure 1B) (FIGO stage 1B). A course of adjuvant pelvic radiotherapy was completed in line

1. Dr. CD Ekanayake is now attached to the Department of Obstetrics and Gynaecology, Faculty of Medicine, General Sir John Kotelawala Defence University, Ratmalana, Sri Lanka.

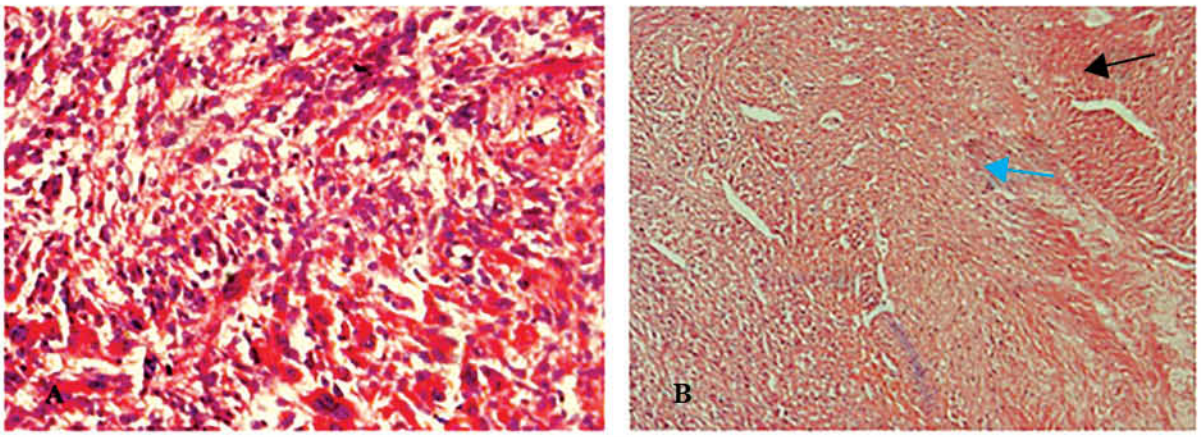

Figure 1 A , shows scattered plemorphic cells of the atypical leiomyoma, **B,** shows an area of cogaulative necrosis **(black arrow)** adjacent to viable tumor **(blue arrow)** in the LMS

with the limited evidence in literature. She is currently being followed up jointly by the gynaecology and oncology teams.

Discussion

The classification of uterine SMT is based on the assessment MA (MF/10 HPFs), cytological atypia and CN. The diagnosis of an AL is conflicting as there is an overlap of the criteria for an AL and smooth muscle tumour of uncertain malignant potential (STUMP)[2,3,4]. As AL are thought to have a low risk of recurrence, a favourable outcome is expected even in patients treated with myomectomy[5].

Different scholars have identified and defined AL and STUMP using varying degrees of histological parameters; MA, MF per HPF and CN. Bell et al, identified 'AL with low risk of recurrence' and 'AL with limited experience' depending on the distribution of cellular atypia and MA[2]. Even the classification of Zaloudek et al, is conflicting as there is overlap between STUMP and other groups of SMTs[4]. To confuse the matters further the recent WHO classification included AL under 'leiomyomas with bizarre nuclei' which is considered a benign variant of typical leiomyoma[3].

Use of this jargon in describing a morphologically similar entity may confuse the clinician with possible implications for determining prognosis. Most AL have shown a benign behavior after total hysterectomy with no recurrence except one case with extensive pelvic and abdominal recurrences two years after hysterectomy[2].

In this patient the diffuse nuclear atypia and a maximum MA of only 5/10HPF with the absence of CN warranted a diagnosis of AL on the first presentation. Extensive sampling excluded the possibility of coexisting LMS. The other possibility is that a separate LMS distinct from the initial lesion co-existed. Although this is supported by the fact that LMS are found in 0.5% of patients undergoing hysterectomy[1], the rest of the uterus was unremarkable at time of initial surgery, which makes this unlikely.

In addition, the fact that leiomyomas are not very well demarcated, coupled with the fact that the two lesions were identified in the anterior region may raise the possibility of incomplete excision, albeit a rare possibility. This raises the possibility of local residual or recurrent disease progression from an AL to LMS and challenges the generalisability of this currently held viewpoint that ALs have a low risk profile.

Conclusion

ALs are mostly found in younger women which invariably leads to fertility conserving treatment. Though these tumors are generally thought to behave in a benign fashion, it may not really be so, and more extensive surgery or further heightened surveillance would be indicated.

Declaration of interest

The authors report no declarations of interest.

Abbreviations

AL-atypical leiomyoma, CN-coagulative necrosis, HPF-high power field, LMS- leiomyosarcoma, MA-mitotic activity, MF-mitotic figures, SMT- smooth muscle cell tumour, STUMP-smooth muscle tumour of uncertain malignant potential.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

References

1. Leibsohn S, D'Ablaing G, Mishell DR Jr, et al. 1990. Leiomyosarcoma in a series of hysterectomies performed for presumed uterine leiomyomas. Am J Obstet Gynecol, 1990; 162(4):968-974.

2. Bell SW, Kempson RL and Hendrickson MR. 1994. Problematic uterine smooth muscle neoplasms: A clinicopathologic study of 213 cases. Am J Surg Patholo, 1994; 18(6):535-558

3. Kim KR, Lax SF, Lazar AJ, Longacre TA, Malpica A,Matias-Guiu X,Nucci MR, Oliva E. Tumours of uterine corpus in WHO Classification of Tumours Female Genital Tumours. Herrington CS. Editor 5th edition 2020 IARC, Lyon, France.

4. Zaloudek CJ, Hendrickson MR , Soslow RA. 2011. Mesenchymal tumors of the uterus. In: Kurman RJ, Ellenson LH and Ronnett BM, editors, Blaustein's Pathology of the Female Genital Tract. 6th edition. New York, Springer, pp 453-527.

5. Ly A, Mills AM, McKenney JK, et al. 2013. Atypical leiomyomas of the uterus: a clinicopathologic study of 51 cases. Am J Surg Pathol. 2013; 37(5):643-649