



# GUIDANCE FOR THE MANAGEMENT OF GYNECOLOGIC SARCOMAS IN ONTARIO:

Recommendations Report 2017

Gynecologic Sarcoma Working Group

Sarcoma Services Steering Committee

## Introduction

Sarcomas are rare malignancies that arise in soft-tissue and bone. They affect all age groups and may arise in any part of the body. Treatment is often multi-modal and complex. Individuals with sarcoma frequently experience significant morbidity and mortality as a consequence of the disease and/or treatment (1, 2). The recognition that sarcoma care is complex and multidisciplinary has led to the adoption of evidence-based policies in Ontario that concentrate sarcoma care in designated sarcoma centres of excellence (3). This provides patients with access to highly specialized medical expertise related to pathology, imaging, surgical, radiation, and systemic therapy required to achieve the best patient outcomes.

Pelvic sarcomas may arise directly in the female reproductive organs or other pelvic tissues and involve the reproductive organs secondarily. Sarcomas arising in female reproductive organs are referred to as gynecologic sarcomas. Cancer Care Ontario data show that in the period from 2010 to 2013, approximately 90 Ontario women were diagnosed annually with a gynecologic sarcoma. This was about seven percent of all adult Ontario sarcoma cases, diagnosed during the same period (Data source: Ontario Cancer Registry (OCR). Date: July 15, 2015).

Accurate pre-operative recognition of primary and secondary pelvic sarcomas involving the female reproductive organs may help with identification of gynecologic sarcomas and is especially important because these are frequently mistaken pre-operatively for more typical gynecologic tumours, potentially leading to inappropriate or suboptimal interventions. This is especially a concern for locally advanced tumours.

## Background

Female benign pelvic tumours are frequently managed initially by general gynecologists in the community and the standard approach to investigation and management of typical uterine masses is well-established (4). However, system-level recommendations for the diagnosis and management of primary or secondary sarcomas involving female reproductive organs are lacking.

Due to the unique management challenges posed by gynecologic sarcomas in Ontario, they were not included in the [Provincial Sarcoma Services Plan](#) (5) released in 2014. However, centres that provide gynecologic sarcoma and non-gynecologic sarcoma services currently consult each other informally in order to provide care for patients with pelvic sarcomas. This includes review of cases at both sarcoma and gynecologic oncology multidisciplinary cancer conferences (MCCs) and cross service/discipline consultations. The need to formalize these processes and optimize collaboration between specialities in order to ensure access to high quality care for patients with gynecologic sarcomas was identified.

The intent of this Recommendations Report is to:

- Align Cancer Care Ontario's Sarcoma Services and Gynecologic Oncology Groups in the care of gynaecologic sarcomas.
- Provide heightened awareness of pelvic sarcomas amongst general gynecologists and pathologists, and to provide guidance on the investigation and management using the recommended care pathway for women with suspected or proven pelvic and extra-pelvic gynecologic sarcomas.

This recommendations report will provide the following:

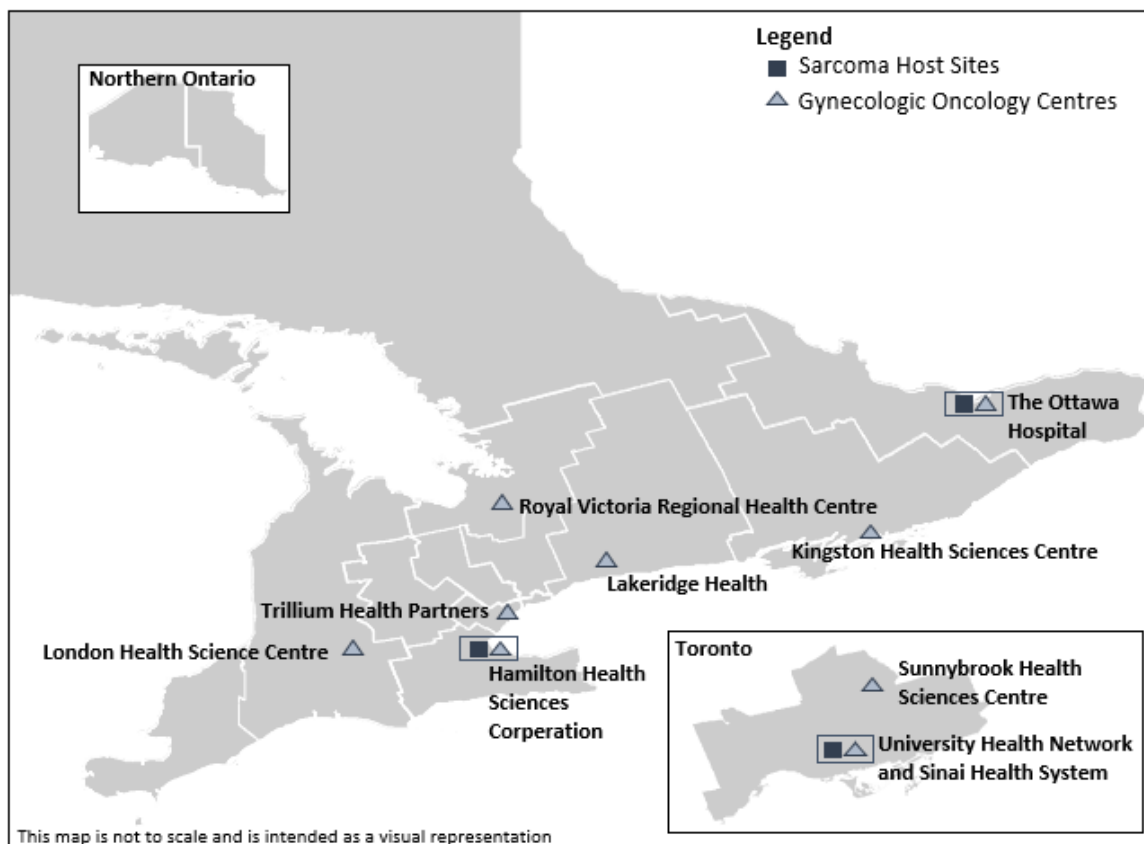
- Guidance on optimal management of a suspected pelvic sarcoma,

- Guidance on how to best direct patients who may have a suspicious pelvic sarcoma, and
- Awareness across the province regarding optimal cancer care.

The scope of this recommendations report is limited to adult patients and does not include paediatrics.

## Recommendations on the Diagnosis and Management of Gynecologic and Non-Gynecologic Pelvic Sarcomas

Since primary and secondary gynecologic sarcomas are relatively rare and the diagnosis and management is often multi-disciplinary, collaboration between Gynecologic Oncology Centres (GOCs) and Sarcoma Services Host Sites (Figure 1) is essential. GOCs are designated centres that provide surgical care, radiation therapy and systemic therapy for all invasive gynecologic oncology disease sites, and act as a hub for the management of all invasive gynecologic cancers (Appendix B). Sarcoma treatment is provided in three multi-region collaborative sarcoma programs. In each of these programs, there is a Sarcoma Services Host Site that provides a full spectrum of specialized sarcoma services (Appendix C).



**FIGURE 1:** MAP OF ONTARIO SHOWING LOCATION OF SARCOMA SERVICES HOST SITES AND GYNECOLOGIC ONCOLOGY CENTRES.<sup>1</sup>

<sup>1</sup> Please note, this map was updated in December 2020 to include Lakeridge Health as a Gynecologic Oncology Centre.

## Multidisciplinary Cancer Conference (MCC) Review

MCCs are regularly scheduled meetings where healthcare providers discuss the diagnosis and treatment of individual cancer patients and over the past 10 years, have become well established in Ontario.

The following are recommended:

1. All suspected or confirmed primary or secondary gynecologic sarcoma cases should be reviewed at an MCC, at a GOC **or** Sarcoma Services Host Site.
2. The following subset of gynecologic sarcoma cases should be presented at **both** GOC and Sarcoma Services Host Site MCCs (Table 1).

**Table 1:** Gynecologic sarcoma cases that should be presented at both Gynecologic Oncology Centre (GOC) and Sarcoma Services Host Site multidisciplinary cancer conferences (MCCs), and may benefit from secondary pathology review at a Sarcoma Services Host Site.

- Histologies that are uncommonly seen in gynecologic sarcomas and more frequently seen in soft tissue sarcomas, such as, but not limited to:
  - Epithelioid sarcomas, Ewing's family tumours, gastrointestinal stromal tumours (GISTs), and angiosarcomas,
  - Where the differential of a soft tissue type sarcoma is suspected or remains after appropriate work-up,
  - Gynecologic tumours with rhabdosarcoma component:
    - Pure rhabdomyosarcomas
    - Gynecological sarcomas with rhabdomyosarcomatous differentiation
    - Gynecological mixed epithelial-mesenchymal tumours (adenosarcoma, carcinosarcoma) with rhabdomyosarcomatous differentiation
  - Cases requested by a pathologist with a specialty or special interest in gynecologic pathology to be reviewed by a pathologist with a specialty or special interest in sarcoma.
- Locally extensive tumours where neoadjuvant therapy (radiation or chemotherapy) may lead to less morbid surgery
- Locally advanced tumours of borderline resectability that might benefit from initial radiation or chemotherapy
- Complex cases that involve multiple organs or pelvic sidewall
- Cases that are being considered for salvage treatment for recurrence that may benefit from genetic profiling, and receive chemotherapy and/or radiotherapy pre-operatively
- Metastatic cases for systemic chemotherapy and for consideration for clinical trials

## Referral of Gynecologic Sarcomas

### *Imaging Recommendations*

Pre-operative diagnosis of any pelvic sarcoma is optimal, but frequently does not occur. This can be a result of a low index of suspicion coupled with the use of pre-operative imaging modalities that provide inadequate information about the true nature of the tumour being imaged.

- Adequately imaged suspected fibroid cases do not require further imaging since imaging is not able to distinguish between fibroids and organ confined leiomyosarcoma (LMS) or endometrial stromal sarcomas

(ESS). These cases are frequently identified post-operatively. All completely resected uterine sarcomas should be referred to a GOC for pathology review, followed by a review by a gynecologic oncologist.

Isolated pelvic masses should not be operated on without imaging as outlined below:

- All pelvic masses with the following suspicious features on ultrasound should be followed up with magnetic resonance imaging (MRI):
  - The ultrasound does not show the contour of the mass (e.g., very large mass)
  - The patient has compressive symptoms (e.g., leg swelling, neuropathic pain)
  - There is suggestion of sidewall involvement on ultrasound
- If extrauterine extension or pelvic side wall involvement is confirmed, the case should be referred to a soft tissue sarcoma or gynecologic oncology surgeon without further intervention.

#### *Surgical Recommendations*

Patients with pre-operative abnormal bleeding should have an endometrial biopsy as some sarcomas may be diagnosed this way.

Incisional and excisional procedures should be avoided if pre-operative or intra-operative findings suggest an atypical diagnosis. These patients should be referred for diagnostic assessment at a specialist centre. Patients with suspicious or confirmed cases of gynecologic sarcomas should have surgery at GOCs or Sarcoma Services Host and Partner Sites (usually for complex cases that involve multiple organs or pelvic sidewall or pelvic sarcomas with secondary involvement of female reproductive organs).

#### *Pathology Recommendations*

All suspected gynecologic sarcoma cases, regardless of topography local extent or completeness of excision, should have the pathology reviewed by pathologists located at a GOC.

Pelvic sarcomas with secondary involvement of female reproductive organs should have the pathology reviewed by pathologists located at a Sarcoma Services Host Site. Cases that may benefit from secondary pathology review at a Sarcoma Services Host Site are enumerated in Table 1.

#### *Adjuvant Treatment Recommendations*

There is no strong evidence to support or not support the use of adjuvant therapy for gynecologic sarcomas. Thus decisions regarding treatment should be determined with the patient on a case by case basis after secondary pathology review by a pathologist with a specialty or special interest in sarcoma and/or gynecologic pathology and discussion at MCC.

#### **Additional Recommendations**

- GOCs and Sarcoma Services Host Sites should be aware of clinical trials available for gynecologic sarcoma patients. Consideration for participation in clinical trials is strongly recommended for each patient at each stage of therapy.
- Early involvement of palliative care is recommended for gynecologic sarcoma patients requiring ongoing symptom management or those anticipated to require end of life care.
- Gynecologic sarcoma patients can experience severe pain, edema, and other symptoms that can impact on their quality of life. Timely symptom management and referral to a palliative care team should be considered.

## Quality Assurance

In order to measure system performance, quality metrics will be put into place. A number of measures are now being developed to assess all sarcoma services and a subset to assess gynecologic sarcomas:

- Wait times
- Percent of cases reviewed at MCC
- Percent of cases with secondary pathology review
- Recurrence rate of disease

In addition, the following indicators are recommended for this patient population:

- Incidence of gynecologic sarcomas in Ontario and stage of disease at time of diagnosis
- The percentage of gynecologic sarcoma patients that receive care at a GOC or Sarcoma Services Host Site

## Conclusions

In conclusion, this recommendations report outlines the key components that should be instituted to increase collaboration between sarcoma service providers and gynecologic oncology centres. Hopefully, it will also provide heightened awareness of pelvic sarcomas amongst general gynecologists and pathologists, and provide guidance on the investigation and management of women with suspected or proven pelvic and extra-pelvic gynecologic sarcomas. This report will also help to inform the provincial approach for planning, funding and performance management for this patient population.

Future work:

- Further exploration of the role of adjuvant treatment and other new treatments for these patients
- Guidance on treatment of patients with metastatic disease for all sarcoma subtypes is needed;
- Quality of life, survivorship, and fertility/sexual health support should be available to this patient population; and
- The role of molecular profiling for this group should be defined.

## References

1. MacNeill AJ, Miceli R, Strauss DC, Bonvalot S, Hohenberger P, Van Coevorden F, Rutkowski P, Callegaro D, Hayes AJ, Honoré C, Fairweather M, Cannell A, Jakob J, Haas RL, Szacht M, Fiore M, Casali PG, Pollock RE, Raut CP, Gronchi A, Swallow CJ. Post-relapse outcomes after primary extended resection of retroperitoneal sarcoma: A report from the Trans-Atlantic RPS Working Group. *Cancer*, 2017; doi: 10.1002/cncr.30572.
2. Davis AM, O'Sullivan B, Turcotte R, Bell R, Catton C, Chabot P, Wunder J, Hammond A, Benk V, Kandel R, Goddard K, Freeman C, Sadura A, Zee B, Day A, Tu D, Pater J. Late radiation morbidity following randomization to preoperative versus postoperative radiotherapy in extremity soft tissue sarcoma. Canadian Sarcoma Group, NCI Canada Clinical Trial Group Randomized Trial. *Radiother Oncol*, 2005;75(1):48-53.
3. Sarcoma Expert Panel (2009). *Adult Sarcoma Management in Ontario – Recommendations Report*. Available at <http://www.cancercare.on.ca/common/pages/UserFile.aspx?fileId=121319>.
4. Donnez J, Dolmans MM. Uterine fibroid management: from the present to the future. *Hum Reprod Update*, 2016;22(6):665-686.
5. Sarcoma Services Steering Committee (2015). *Provincial Sarcoma Services Plan, Version 2.0*. Available at [www.cancercare.on.ca/sarcomacare](http://www.cancercare.on.ca/sarcomacare).

## Appendices

### Appendix A: Gynecologic Sarcoma Codes

Gynecologic sarcoma cases were identified in OCR through the following gynecologic topography codes (anatomical location).

Topography Code	Topography Description
C519	Vulva NOS
C529	Vagina NOS
C530	Endocervix
C510	Labium majus
C572	Round ligament
C549	Corpus uteri
C559	Uterus NOS
C569	Ovary
C571	Broad ligament
C574	Uterine adnexa
C578	Overlapping lesion of female genital organs
C579	Female genital tract NOS
C538	Overlapping lesion of cervix uteri
C540	Isthmus uteri
C548	Overlapping region of the corpus uteri

### Appendix B: List of Gynecologic Oncology Centres (GOCs)

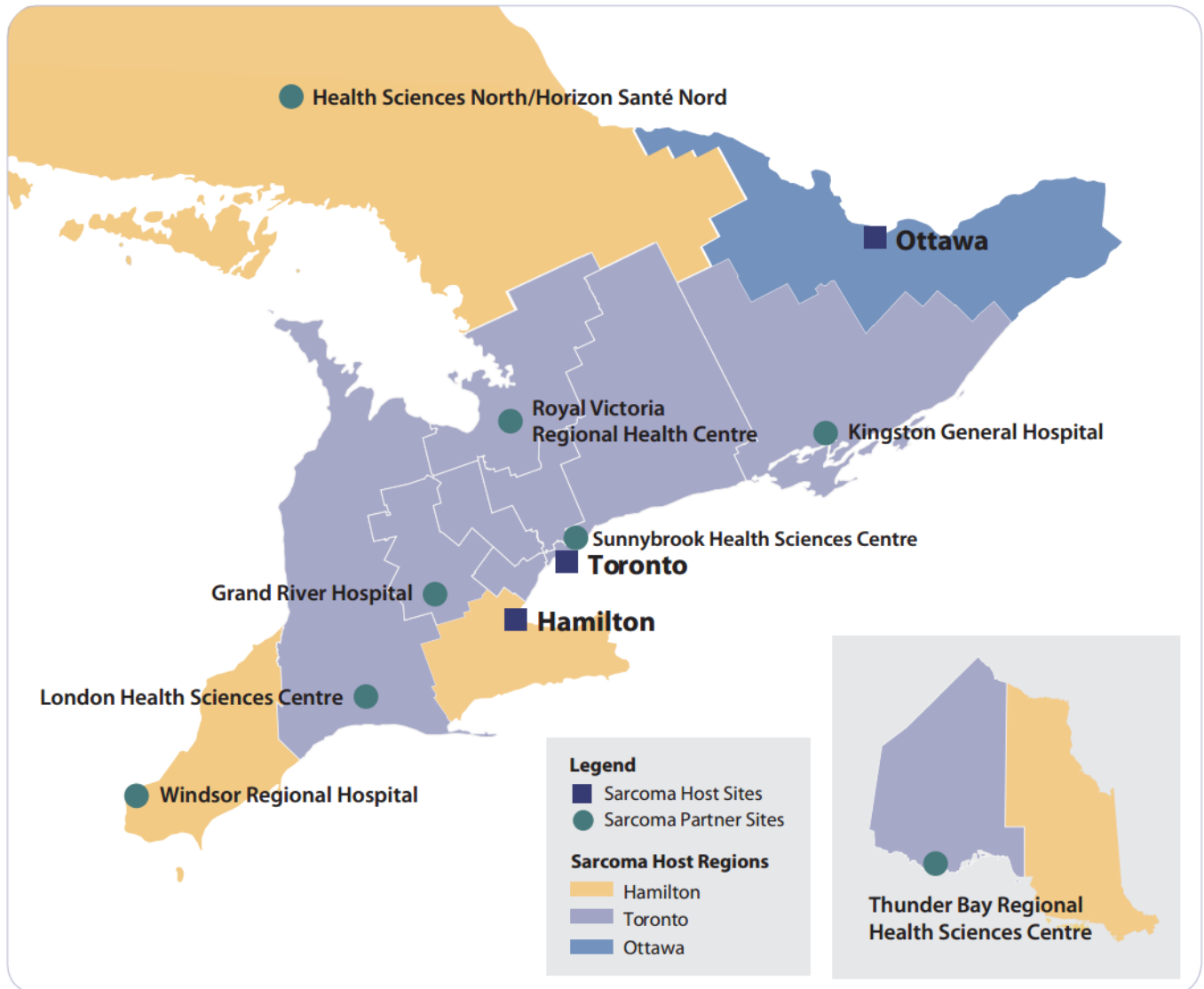
- University Health Network
- Hamilton Health Sciences Centre
- The Ottawa Hospital
- Sunnybrook Health Sciences Centre
- London Health Sciences Centre
- Trillium Health Partners
- Kingston Health Sciences Centre
- Royal Victoria Regional Health Centre
- Sinai Health System

For more information regarding Gynecologic Oncology Centres, please visit:

[http://www.csqi.on.ca/by\\_patient\\_journey/treatment/gynecologic\\_oncology\\_centres/](http://www.csqi.on.ca/by_patient_journey/treatment/gynecologic_oncology_centres/)



## Appendix C: Location of Sarcoma Services Host and Partner Sites



For more information regarding Sarcoma Services Host and Partner Sites, please visit:

[www.cancercare.on.ca/sarcomacare](http://www.cancercare.on.ca/sarcomacare).

## Appendix D: Gynecologic Sarcoma Working Group Members

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