

## **‘An extra-skeletal Ewing’s Sarcoma presenting with urinary retention- Impact of anatomical location on patient management; A case report’**

Gunathilaka K<sup>1</sup>, Jamaldeen R<sup>1</sup>, Ekanayake J<sup>2</sup>, Hettiarachchi M<sup>2</sup>, Udupihille J<sup>3</sup>, Waduge R<sup>4</sup>

<sup>1</sup>*Teaching hospital, Peradeniya*

<sup>2</sup>*Department of surgery, Faculty of Medicine, University of Peradeniya*

<sup>3</sup>*Department of Radiology, Faculty of Medicine, University of Peradeniya*

<sup>4</sup>*Department of Pathology, Faculty of Medicine, University of Peradeniya*

**Running title** - ‘An extra-skeletal Ewing’s Sarcoma presenting with urinary retention; A case report’

### **Abstract**

**Background:** Ewing’s sarcoma is one of the common malignant childhood tumours. The radiological features of pre-sacral Ewing’s sarcoma may mimic primary nerve sheath tumor or teratoma.

**Case Presentation:** A 16-year-old girl presented with acute retention of urine found to have a midline mass in the pre-sacral region of the pelvis in clinical examination and radiological scans. A preoperative biopsy was not done. The complex location and the need to excise the biopsy tract, if histology turned out to be malignant, predicated this decision. The tumor was entirely excised. The histological diagnosis was that of an Ewing’s Sarcoma, with focal infiltration of the surgical margins.

**Conclusion:** The anatomical site and size of an Ewing’s sarcoma should be considered before

preoperative biopsy is considered. A biopsy result positive for malignancy would favor adjuvant chemotherapy and radiation, but requires the biopsy tract to be excised. Primary resection is the preferred choice when the location is complex.

**Keywords:** Ewing’s Sarcoma, retroperitoneal pelvic mass, urinary retention, anatomical location, primary resection

### **Background**

Ewing’s sarcoma is one of the common malignant childhood tumour, which is named after James Ewing who described it in 1920 (1). Along with osteosarcoma, it is the second most common malignant bone tumor in children (2). ES can occur in virtually any location of the body (3). However, a pre sacral mass causing acute urinary retention is a very rare presentation (4). The radiological features of pre-sacral Ewing’s sarcoma may mimic primary nerve sheath tumor or teratoma. The management of Ewing s sarcoma most of the time is histological confirmation followed by neo adjuvant chemotherapy / radiotherapy and surgical excision. However a tailor made

‘An extra-skeletal Ewing’s Sarcoma presenting with urinary retention- Impact of anatomical location on patient management; A case report’

approach would be effective and safe depending on the different anatomical sites and sizes.

## Case Presentation

A 16-year-old girl presented with acute retention of urine. She had no constitutional symptoms like loss of weight or appetite. A digital rectal examination revealed a pre-sacral mass. A Contrast enhanced computed tomography (CECT) of the pelvis showed a well-defined, heterogeneously enhancing, midline mass in the pre-sacral region. It measured 9cm x 6.5cm x 6cm in size. It consisted of solid and cystic areas, but there were no foci of calcification. The mass was overlying the anterior surfaces of 2nd to 5th sacral vertebral bodies and was seen to communicate with the sacral spinal canal. The CECT appearance was that of a pre-sacral tumor, suggestive of a schwannoma or a

teratoma. The Magnetic Resonance Imaging (MRI) scan (Figure 1) confirmed the CECT findings, but showed no evidence of bony erosions, usually associated with a peripheral nerve sheath schwannoma.



Figure 1- MRI Scan of Abdomen and Pelvis showing the extent of the tumour within the pelvis.

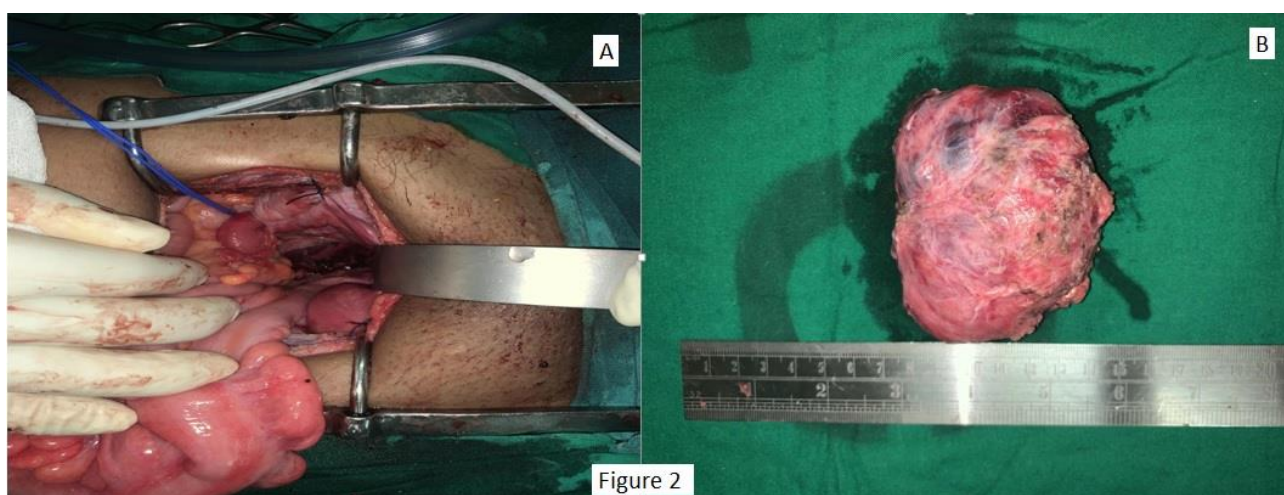


Figure 2- Intraoperative view of the tumour being transected (A) and the resected mass (B).



‘An extra-skeletal Ewing’s Sarcoma presenting with urinary retention- Impact of anatomical location on patient management; A case report’

The patient made an uneventful recover and was referred to the oncology service for further management. The histological diagnosis was that of an Ewing’s Sarcoma (Figure 3 & 4), with focal infiltration of the surgical margins.

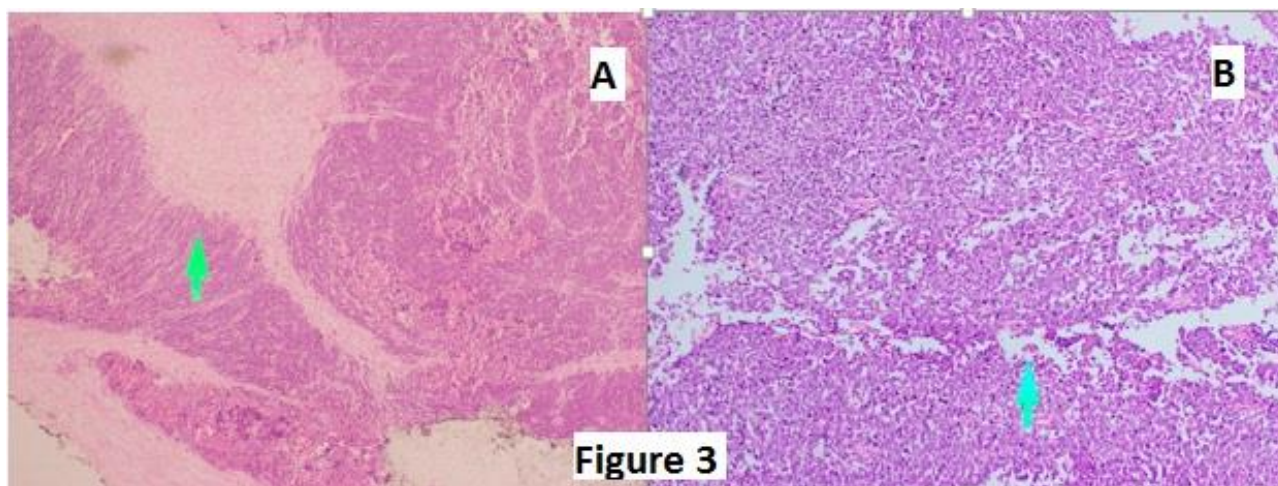


Figure 3- Histopathological slides showing neoplasm composed of sheets of small round cells separated into vague nodules by fibrous septae. Cells contain round nuclei with coarse chromatin pattern. The mitotic activity is brisk (A & B).

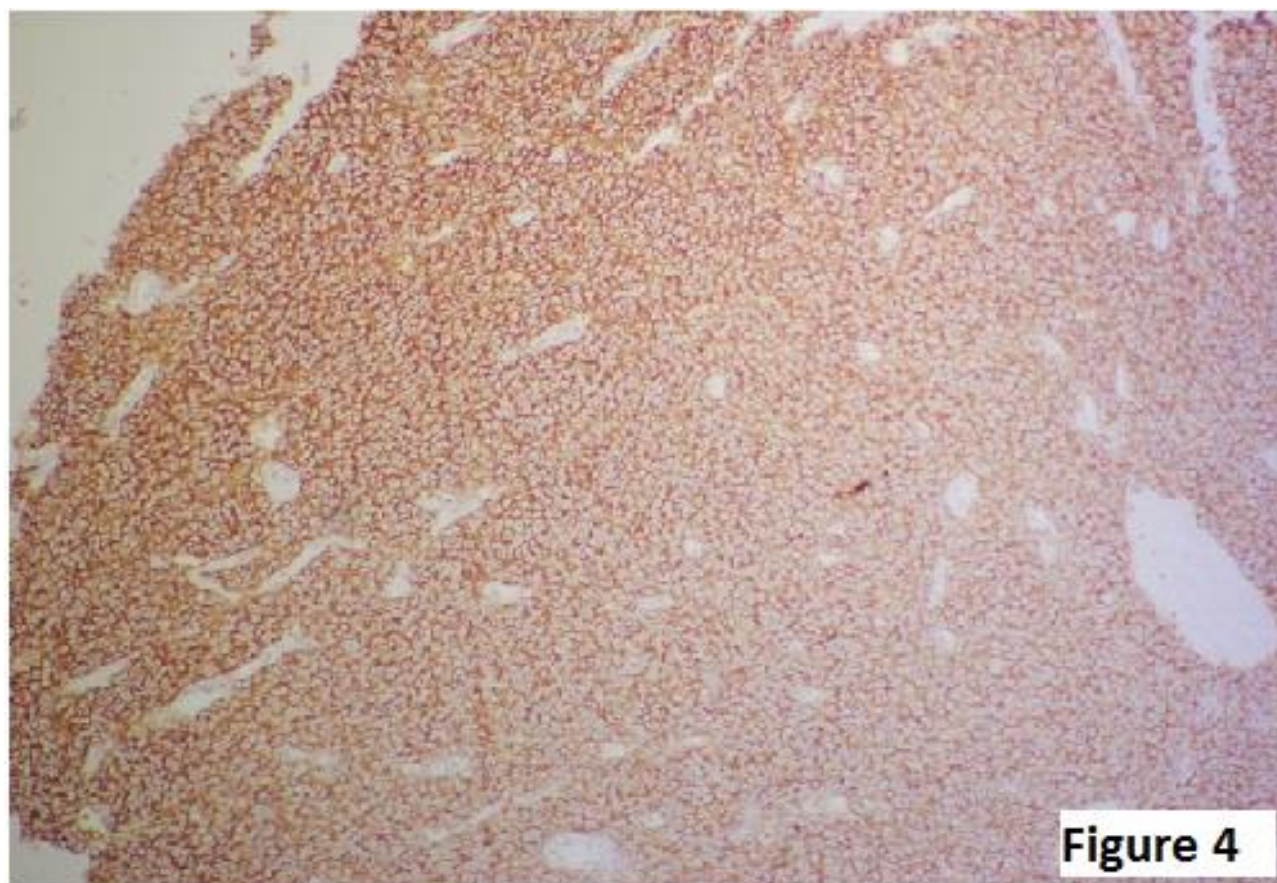


Figure 4 – The immune stain slide that showing diffuse cytoplasmic positivity of tumour cells.

‘An extra-skeletal Ewing’s Sarcoma presenting with urinary retention- Impact of anatomical location on patient management; A case report’

She was commenced on chemotherapy followed by radiotherapy. A repeat CT confirmed complete excision of mass.

## **Conclusion**

The CECT and MRI images in our patient were strongly suggestive of a benign schwannoma or teratoma and the presentation with urinary retention had made schwannoma the most likely possibility. However, the radiology report had stated that a malignant tumor could not be excluded. The decision to proceed directly to laparotomy had been strongly influenced by the highly suggestive radiological findings. Pre-operative ultrasound guided biopsy, via a trans-rectal or percutaneous route, was considered. Due to the position of the tumor, a percutaneous biopsy via the posterior abdominal wall was considered the safest option. However, if the histology had proved the tumor to be malignant, comprehensive resection would have required excision of the entire biopsy tract as well. This would have been a significant surgical challenge, regardless of the approach used to biopsy the tumor. In this instance, diagnosis of ES was only made post-operatively.

A pre-surgical histological diagnosis may have changed management, to include radiotherapy and neo-adjuvant chemotherapy (5). This may have provided a better outcome with regard to local recurrence and survival. However, we believe the benefit to our patient would have been marginal, as the tumor was successfully resected in its entirety.

The anatomical site and size of an Ewing’s sarcoma should be considered before preoperative biopsy is considered. A biopsy result positive for malignancy would favor adjuvant chemotherapy and radiation, but requires the biopsy tract to be excised. Primary resection is the preferred choice when the location is complex.

## **List of Abbreviations**

ES – Ewing’s sarcoma

CECT - A Contrast enhanced computed tomography

MRI- The Magnetic Resonance Imaging

## **Acknowledgements**

We would like to express our sincere gratitude to the patient and her parents for giving the consent to present this case in this paper. We would like to appreciate Prof. H.D.P. De Silva for guiding us in fine adjustments in language and editing the manuscript.

## **Competing Interests**

The authors declare that there are no competing interests.

## **Funding**

No funding sources were used for the case report preparation.

‘An extra-skeletal Ewing’s Sarcoma presenting with urinary retention- Impact of anatomical location on patient management; A case report’

## Corresponding Author

Kasun Gunathilaka,  
Professorial Surgical Unit,  
Teaching Hospital,  
Peradeniya

mrkm.gunathilaka@gmail.com  
071 7233026

CC BY 4.0

This is an Open Access article distributed under the terms of the Creative Commons Attribution 4.0 license (unless stated otherwise) which permits unrestricted use, distribution and reproduction in any medium, provided the original work is properly cited. Copyright is retained by the author(s).



## References

1. Choi EY, Gardner JM, Lucas DR, McHugh JB, Patel RM. Ewing sarcoma. *Semin Diagn Pathol.* 2014 ; doi: 10.1053/j.semdp.2014.01.002.
2. Ottaviani G, Jaffe N. The epidemiology of osteosarcoma. *Cancer Treat Res.* 2009; doi: 10.1007/978-1-4419-0284-9\_1.
3. Balamuth NJ, Womer RB. Ewing's sarcoma. *Lancet Oncol.* 2010 ;doi: 10.1016/S1470-2045(09)70286-4.
4. Ray P, Girach J, Sanghrajka AP. Ewing's sarcoma of the pelvis: an unusual, but not to be missed, cause of an irritable hip. *BMJ Case Rep.* 2016 ; doi: 10.1136/bcr-2015-213782.
5. Dunst J, Sauer R. Therapie des Ewing-Sarkoms *Strahlenther Onkol.* 1993 Dec;169:695-708.
6. Ahmed SK, Robinson SI, Arndt CAS, Petersen IA, Haddock MG, Rose PS, Issa Laack NN. Pelvis Ewing sarcoma: Local control and survival in the modern era. *Pediatr Blood Cancer.* 2017 ; doi: 10.1002/pbc.26504.