The Natural History of Retroperitoneal-Abdominal-Pelvic Ganglioneuromas: An International Study By The Transatlantic Australasian Retroperitoneal Sarcoma Working Group (TARPSWG)

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No conflicts of interest to declare.
INTRODUCTION

• Ganglioneuromas (GN) are rare tumors derived from neural crest cells within the sympathetic plexus and adrenal glands
• Often discovered incidentally, or during work-up of non-specific symptoms caused by mass effect
• Occasionally occur in setting of hereditary syndromes (NF-1, MEN2)
INTRODUCTION

• Data on Retroperitoneal-Abdominal-Pelvic GN is limited to case reports and single-institution case series
• GN lack specific clinical symptoms/signs, laboratory findings, or diagnostic imaging characteristics
• Rates of malignant degeneration to neuroblastoma remain unclear
OBJECTIVE

• To conduct a retrospective, multi-institutional, multi-national study of the natural history and clinical management of ganglioneuroma.
GLOBAL COLLABORATION: 29 INSTITUTIONS ACROSS 5 CONTINENTS
METHODS

• Following IRB approval, retrospective chart reviews of patient diagnosed with retroperitoneal, abdominal, and pelvic GN were performed
• January 2000 – January 2020
• All ages included
• Excluded head and neck, mediastinal, and GI tract tumors

• Examined patient demographics, clinicopathological features, imaging follow up, approaches to operative & non-operative management, and patient outcomes
PATIENT DEMOGRAPHICS

- 328 patients from 29 institutions (5 continents)
- Median age at diagnosis: 37 yo (range, 4–79 yo)
- Sex: 59% female, 41% male
- Location:
  - Extra-adrenal retroperitoneum 67%
  - Adrenal glands 19%
  - Pelvis 11%
  - Peritoneal cavity 3%
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CLINICAL FEATURES

- Symptomatic presentation: 41%
- Vascular encasement: 17%
- Major nerve involvement: 8%
- Hereditary syndromes: 2.4%
  - Neurofibromatosis-1 (N=6)
  - MEN 2A (N=1)
  - Schwannomatosis (N=1)
MANAGEMENT: NON-OPERATIVE VERSUS OPERATIVE

• Reasons for non-operative management:
  • Asymptomatic
  • Indolent nature of the lesion
  • Potential risks of resection

• Median follow-up of 1.9 years (IQR 0.8 – 4.2)
## TUMOR SIZE AND SURVEILLANCE

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<tr>
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<th>Value</th>
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<tbody>
<tr>
<td>Median Tumor Diameter</td>
<td>7.2 cm (IQR 5.0 – 9.9)</td>
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<tr>
<td>Vascular encasement</td>
<td>9.4 cm (IQR 7.2 – 12.0)</td>
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</tbody>
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Progressive disease in 3 (2.3%)

Stable disease in 126 (97.7%)
OPERATIVE MANAGEMENT

- Indications: Tumor growth, symptoms/mass effect, concerns for malignancy, and diagnostic uncertainty
- 64.6% underwent resection
  - 74.5% achieving R0/R1 resection
- 84% disease free at median follow-up of 3.0 years (IQR 1.0 – 5.5)
- Recurrences: 5 (2.4%) cases
NEUROBLASTOMA TRANSFORMATION

<table>
<thead>
<tr>
<th>Neuroblastoma transformation</th>
<th>3 (0.9%)</th>
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<tr>
<td>Neuroblastoma transformation age</td>
<td>9-11 yo</td>
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Ganglioneuroma 0.9% Neuroblastoma
SUMMARY

• Most patients with ganglioneuroma present sporadically
• Less than half of patients present with symptoms
• Most GN have indolent disease courses and <1% transformed into neuroblastoma
• Non-operative management with serial imaging evaluations may be appropriate when the risks outweigh benefits of resection in confirmed benign or asymptomatic cases
• If technically feasible and safe, definitive operative management with R0/R1 resection may be recommended when tumors are symptomatic, growing or of uncertain biology
CONCLUSION

- Largest study of ganglioneuroma to date
- While retrospective in nature, we can utilize this data to learn about the natural history of this tumor type to help guide management decisions and to design future studies
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