Total Resection of a Giant Retroperitoneal and Mediastinal Ganglioneuroma - A Case Report

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Background: Ganglioneuromas (GNs) are extremely rare, slowly growing, benign tumors that can arise from Schwann cells, ganglion cells, neuronal or fibrous tissues (1-3). Due to their origin from the sympathetic neural crest they show neuroendocrine potential, however, most are reported to be hormonally inactive (4). Nevertheless, complete surgical removal is recommended for symptom control or for prevention of potential malignant degeneration (5).

Year	Age	Sex	Localization	DM	Procedure
2018	42	♀	thoracic	23	Open TR
2006	35	♂	retroperitoneal	22	Open TR
2017	21	♂	retroperitoneal	21.5	Open TR
2013	18	♂	Retropharyngeal	19	Open TR
2011	53	♂	retroperitoneal	19	Open TR
2019	4	♂	retroperitoneal	17.3	Open PR
2017	5	♂	mediastinal	16	Open TR
2016	42	♀	retroperitoneal	14.5	Open TR
2007	23	♀	retroperitoneal	13	Lap. TR
2013	12	♀	presacral	12	Open TR
2017	12	♀	thoracic	12	Open PR
2014	66	♀	thoracic	12	n.a.
2016	12	♀	retroperitoneal	13	Open PR

Literature review for resected thoracoabdominal tumors.
TR … total resection, PR … partial resection,
DM … maximum diameter (in cm)

Year	Entity	Age	Sex	DM	Procedure
2011	Liposarcoma	39	♂	40	Open TR
2019	Schwannoma	58	♂	20.2	Open TR
2019	GN	10	♀	8	Lap. TR
2019	Neuroblastoma	24	♂	7	Open TR
2019	Sarcoma	74	♀	3	Open TR

Literature review for large resected GN (>10cm diameter).
TR … total resection, PR … partial resection,
DM … maximum diameter (in cm)

References

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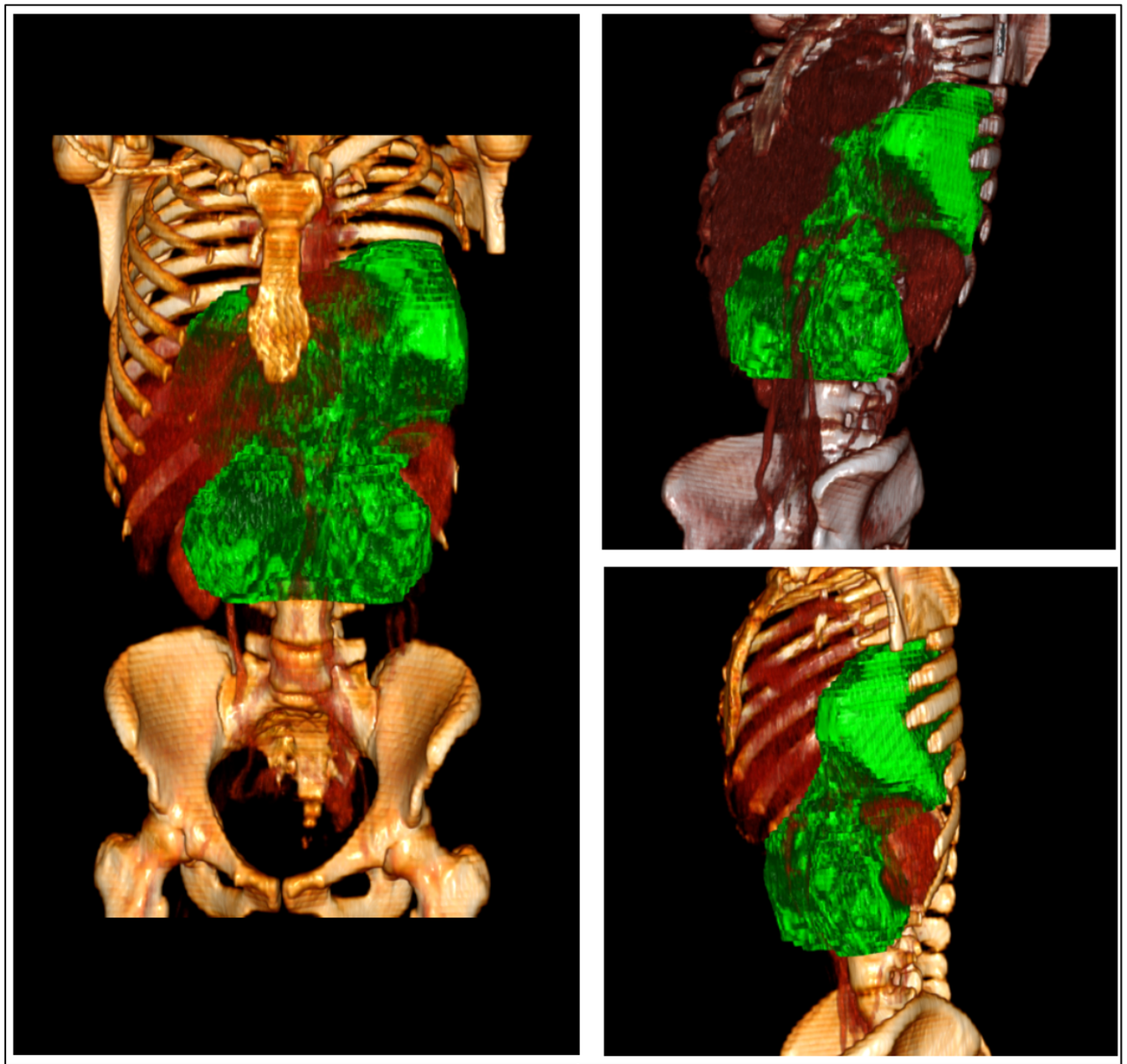
Case Report: A 30-year old female was referred to our oncologic center due to a giant retroperitoneal and mediastinal mass detected in computed tomography (CT) scans. Initial symptoms were transient nausea, diarrhea and crampy abdominal pain. There was a positive family history including 5 first- and second-degree relatives. Presurgical biopsy revealed a benign ganglioneuroma. Total resection (TR) of a 35x25x25 cm, 2550 g tumor was obtained successfully via laparotomy combined with thoracotomy and partial incision of the diaphragm. Histopathological analysis confirmed the diagnosis. Surgically challenging aspects were the bilateral tumor invasion from the retroperitoneum into the mediastinum through the aortic hiatus with the need of a bilateral 2-cavity-procedure, as well as the tumor-related displacement of abdominal aorta, the mesenteric vessels and inferior vena cava. Due to their anatomic course through the tumor mass, lumbar aortic vessels needed to be partially resected. Postoperative functioning was excellent without any sign of neurologic deficit.

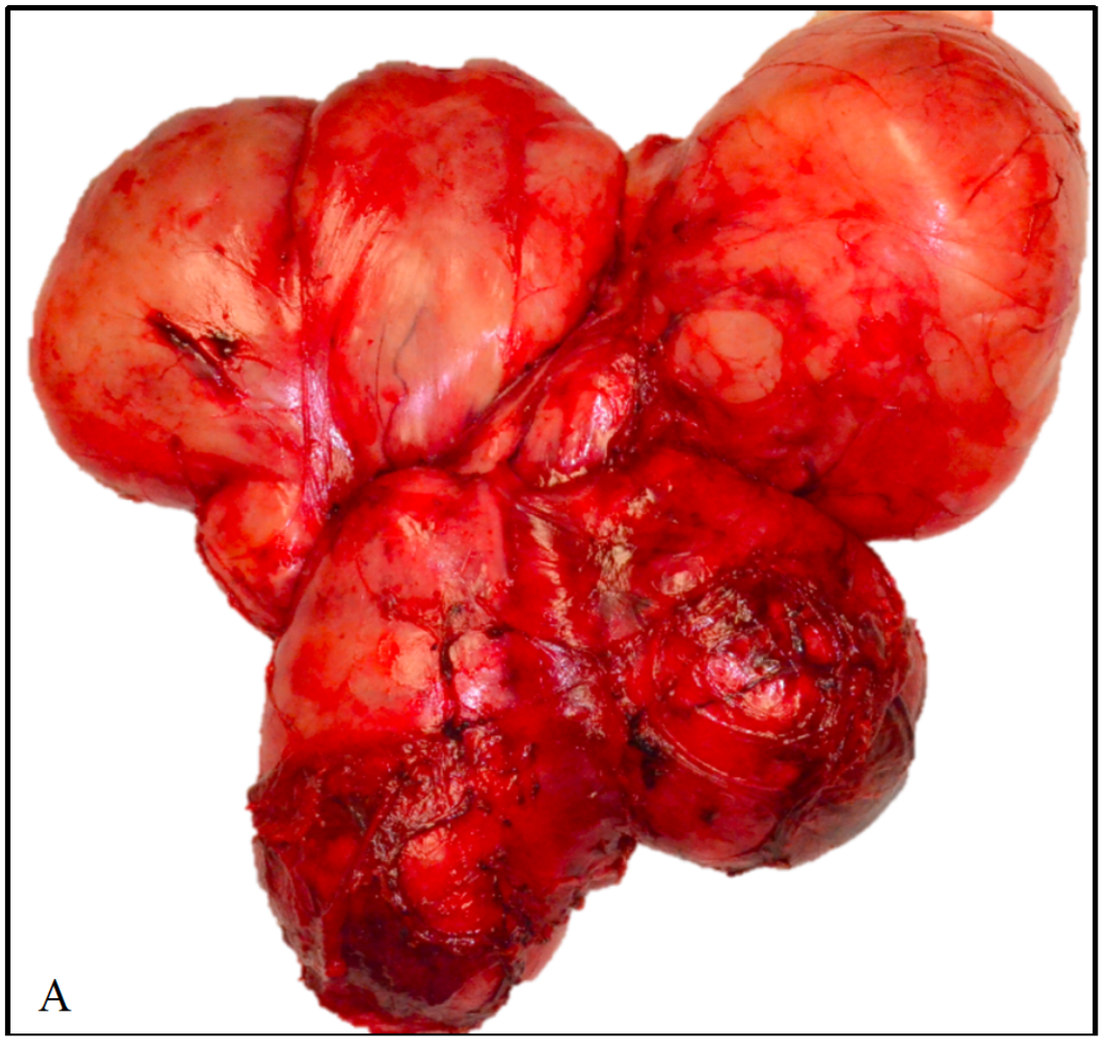


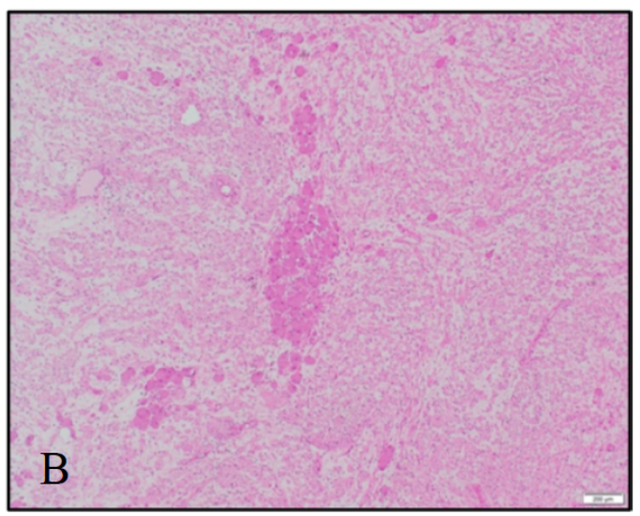
3D CT reconstructions.

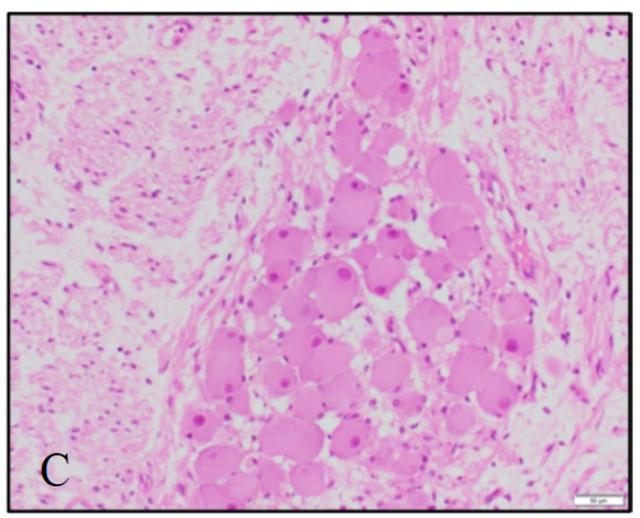
The GN (highlighted in green) expands from the retroperitoneal space through the aortic hiatus into the posterior mediastinum with major vessel enclosing.

Conclusion:

- This is the largest reportedly resected GN (35 cm) and was performed safely.
- First systematic literature review for large GN (>10 cm).
- First systematic literature review for resected thoracoabdominal tumors.







(A) Macroscopic specimen of the in toto resected GN (35 x 25 x 25 cm).

(B, C) Hematoxylin & eosin stained histopathological image of the central tumor portion with ganglion cells and stromal tissue as well as myxofibrotic and fat tissue in the transition zone can be observed.