



Received: 2016.05.18
Accepted: 2016.08.16
Published: 2017.05.25

Authors' Contribution:

- A** Study Design
- B** Data Collection
- C** Statistical Analysis
- D** Data Interpretation
- E** Manuscript Preparation
- F** Literature Search
- G** Funds Collection

Retroperitoneal Ganglioneuroma Mimicking a Kidney Tumor. Case Report

Agnieszka Dąbrowska-Thing^{1BCDEF}, Wojciech Rogowski^{2BD}, Ryszard Pacho^{3CDE},
Ewa Nawrocka-Laskus^{1DEF}, Zaneta Nitek^{1DEF}

¹ Department of Diagnostic Imaging, Gruca Orthopedic and Trauma Teaching Hospital, Centre of Postgraduate Education, Otwock, Poland

² Department of Urology and Oncological Urology, Central Clinical Hospital of the Ministry of the Interior and Administration in Warsaw, Warsaw, Poland

³ 2nd Department of Clinical Radiology, Medical University of Warsaw, Warsaw, Poland

Author's address: Agnieszka Dąbrowska-Thing, Department of Diagnostic Imaging, Gruca Orthopedic and Trauma Teaching Hospital, Centre of Postgraduate Education, Otwock, Poland, e-mail: aga.dabrowska@poczta.fm

Summary

Background:

Ganglioneuroma (GN) is a rare benign tumor arising from the neural crest cells. The reported incidence of GN is one per million population. As a primary retroperitoneal tumor, it constitutes only a small percentage of 0.72 to 1.6%. GN can arise *de novo* or as a result of maturation of a neuroblastoma either spontaneously or after chemotherapy. The most common location is the posterior paraspinal mediastinum, retroperitoneum, neck and adrenal gland. However, GN can potentially occur anywhere along the peripheral autonomic ganglion sites. Most ganglioneuromas are asymptomatic and found incidentally.

Case Report:

We present a case of retroperitoneal ganglioneuroma that mimicked renal mass on imaging. The tumor was incidentally discovered during an abdominal ultrasound examination 43-year-old male patient without clinical symptoms. Complete surgical resection was subsequently performed and histopathological examination of the retroperitoneal mass revealed GN.

Conclusions:

Retroperitoneal ganglioneuroma is a rare benign tumor, generally asymptomatic, which grows slowly, and appears large when it is identified. Preoperative diagnosis can be challenging, particularly in asymptomatic case. Histopathological examination is currently the mainstay of diagnosis. In the case presented herein GN strictly adjoined to the left kidney mimicking renal mass.

MeSH Keywords:

Ganglioneuroma • Kidney Neoplasms • Magnetic Resonance Imaging • Retroperitoneal Space

PDF file:

<http://www.polradiol.com/abstract/index/idArt/899633>

Background

Ganglioneuroma (GN) is a benign tumor originating from cells of the sympathetic nervous system. It occurs very rarely, mainly in young people, and it is often diagnosed incidentally. It may develop *de novo* or as a result of maturation of neuroblastoma (spontaneous remission phenomenon), spontaneously or following chemotherapy/radiotherapy [1].

Prevalence of ganglioneuroma in a population is one per million. As a primary retroperitoneal tumor, GN constitutes 0.72% to 1.6% [2].

In numerous patients the tumor is poorly symptomatic or completely asymptomatic and may grow slowly without producing clinical symptoms. Some tumors exhibit hormonal activity, presenting a broad range of symptoms depending on the type of produced hormone.

Most common localizations include posterior mediastinum (41.5%), abdominal cavity, including adrenal glands (21%), and retroperitoneal space (37.5% [3]). However, the tumor may develop in any location where sympathetic nervous system cells are present. Scarce reports on GN demonstrate that due to nonspecific features of the tumor in imaging studies, radiological diagnostics of this lesion may be challenging.

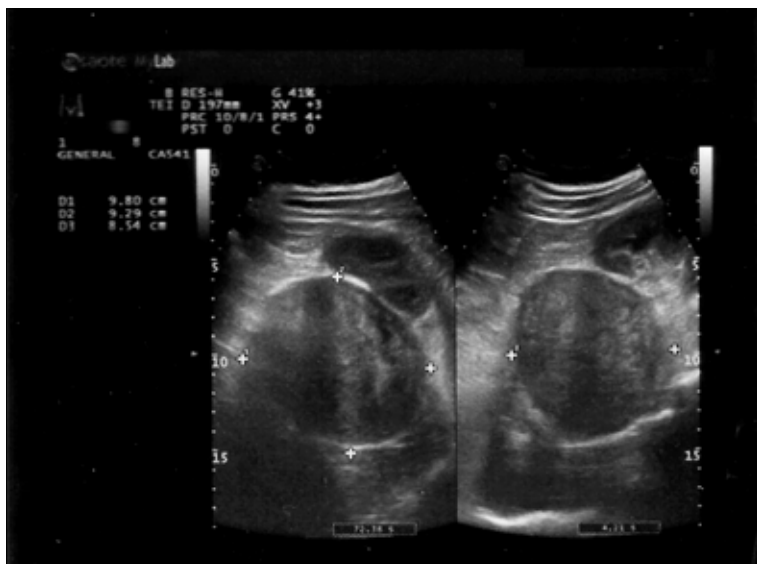


Figure 1. Sagittal abdominal ultrasonography revealed a well defined, round, hypoechoic, slightly heterogeneous solid mass, 9 cm in diameter, adjacent to the upper pole of left kidney.



Figure 2. Abdominal CT with contrast. Coronal multiplanar reconstructed CT image. Unenhanced CT scan of the abdomen shows a well defined, hypodense, homogeneous mass with density of 20HU. Unenhanced CT scan shows homogeneous.



Figure 3. CT-delayed phase acquired 10 min after contrast injection. Linear areas of mild contrast enhancement up to 30 HU were observed within the mass.

Variable image may ensue, among other things, from varying number of Schwann cells, ganglion cells, collagen fibers, fat, or myxomatous stroma composing the tumor [4].

Below, we present a case of a large tumor located in retroperitoneal space, in direct contact with the upper pole of left kidney.

Case Report

A 43-year-old male underwent ultrasound examination of the abdominal cavity, which revealed a presence of well demarcated, hypoechoic, somewhat heterogeneous solid mass, 9 cm in diameter (Figure 1). Power Doppler imaging showed no vascularization within the tumor.

The diagnostics was extended to include contrast computed tomography of the abdomen and lesser pelvis, which

confirmed the presence of a hypodense lesion in the retroperitoneal space (Figure 2).

The lesion was not contrast-enhanced in the arterial or venous phases. Discrete, linear areas of contrast enhancement were visible in the late phase, about 10 min. after administration of contrast agent (Figure 3).

Since malignancy could not be ruled out in CT images, a contrast MRI of the abdomen was also performed, revealing a solid tumor with regular margins adjacent to the upper pole of left kidney (Figures 4, 5).

Tumor signal was homogeneously low in T1-weighted images, while T2-weighted sequences showed intermediate, heterogeneous signal.

The tumor exhibited mild contrast enhancement, no diffusion restriction was noted in DWI sequences (Figure 6).



Figure 4. Coronal T1-weighted image shows a homogenous mass with homogeneously low signal intensity.

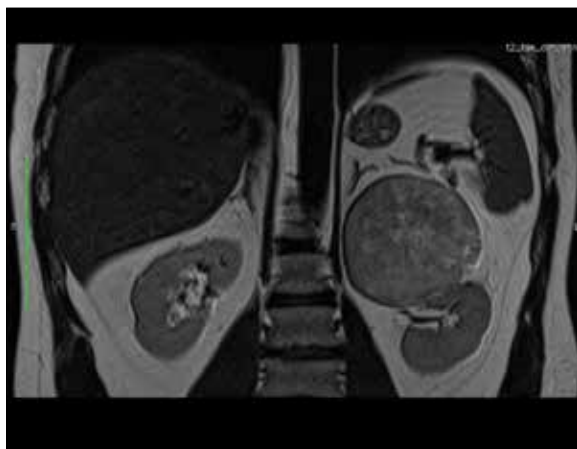


Figure 5. Coronal T2-weighted image shows heterogeneously intermediate signal in the lesion.

Image was still ambiguous and the differential diagnosis included mild, slowly growing, solid tumor, an atypical cancer, and oncocytoma (without a typical central scar).

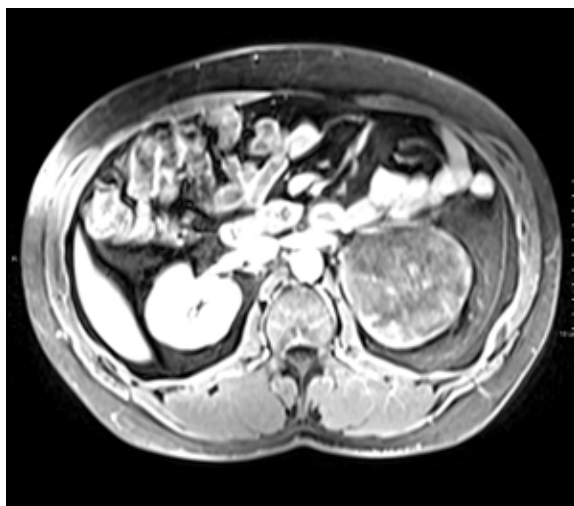


Figure 6. T1-weighted image, dynamic sequences with contrast enhancement. Interspersed areas with weak contrast enhancement were visualized within the mass.

No enlarged or pathological lymph nodes were visualized in any of the conducted imaging studies. The remaining abdominal structures were not pathologically changed. Routine blood tests showed no deviations from normal values. Erythrocytes were identified in the urine.

Patient was referred for removal of the lesion and was admitted to the urology unit. It was revealed during tumor resection (operator: Wojciech Rogowski, MD) that the lesion was not connected to the kidney, but only adjacent to it and covered by a common capsule. Adrenals were unchanged. Tumor of the retroperitoneal tumor was removed as a whole and sent for histopathological examination (Figure 7). Histopathological diagnosis of the tumor was ganglioneuroma and the result was confirmed with immunohistochemistry.

The tumor was composed of Schwann cells and mature ganglion cells. There were no features of nuclear atypia or necrotic foci.

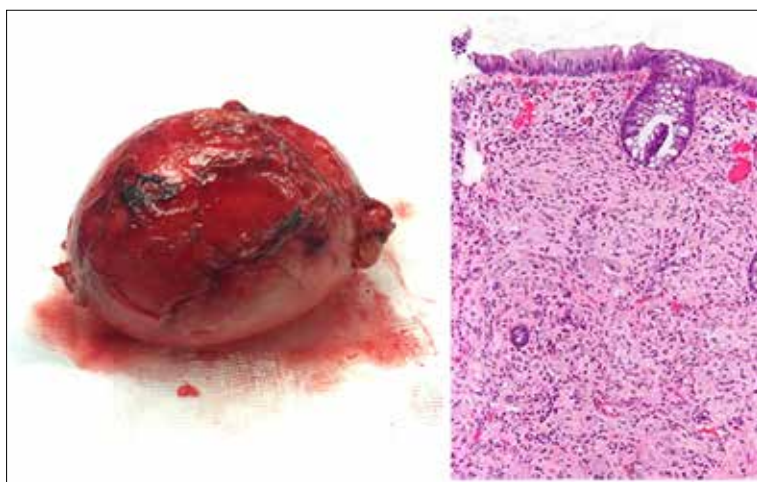


Figure 7. Macroscopic picture of the specimen.

Discussion

Retroperitoneal space is a potential space for growth of both malignant and benign lesions. Tumors located in this area may not present clinical symptoms for a long time regardless of their histological origin and the diagnosis is often incidental.

Primary tumors of the retroperitoneal space are rare. They are malignant in the majority of cases – about 80% [5]. First, we should mention sarcomas comprising up to 90% of all malignant tumors in this area. They are formed outside the retroperitoneal organs and their classification depends on the histological type. The most common include liposarcomas, malignant fibrous histiocytoma, and leiomyosarcoma. Most common benign tumors include lesions of neural origin (schwannoma, neurofibroma), paragangliomas, fibromas, angiomyolipomas of renal origin, and lipomas.

Ganglioneuroma is a rare, benign tumor originating from the cells of the sympathetic trunk. It may grow nearly anywhere along sympathetic ganglia, or in the adrenal medulla. This kind of tumor was first described by Loretz in 1870 [6]. Literature reports on GN published since then provided more information regarding characteristics of this tumor.

The reported case is a classical example of incidentally found pararenal tumor, where extended diagnostic imaging failed to yield unequivocal diagnosis. There are numerous reports in the literature of asymptomatic renal and pararenal tumors incidentally detected during routine imaging studies. Patient did not report any symptoms, which according to Papavramidis corresponds to about 40% of asymptomatic GNs [7].

A large tumor located in the retroperitoneal space in close proximity to the left kidney was detected during ultrasound examination. Ultrasonographic image of the lesion, i.e. well demarcated, hypoechogenic with heterogeneous echogenicity, might have corresponded to GN [8,9]. Available literature contains only 2 cases where GN mimicked a renal tumor in an adult patient [10,11]. Due to the incidental nature of this phenomenon, pararenal GN was not considered in the differential diagnosis. However, we did take into account more common lesions, such as complex cyst or a benign tumor of renal origin.

Contrast enhanced CT examination confirmed the presence of a hypodense tumor with density of 20 HU and exhibiting late phase contrast enhancement. These characteristics were in agreement with the results of studies conducted by Guan, who compared pictures of GN in computed tomography [12]. In the mentioned study lesions were described as hypodense, homogeneous or heterogeneous, with densities ranging between 20 and 40 HU and pronounced late phase contrast enhancement in a range of 10–20 HU. Magnetic resonance is a useful diagnostic tool for identification of the character of a lesion. Publications regarding GN report varying signal in MR examinations. An analysis conducted by Zhang [4] provided better understanding of the relationship between histologic structure of these tumors and their MR picture. As it turned out, variable character of the signal resulted from different proportions of, i.e. Schwann's cells, ganglion cells, collagen fibers, fat tissue, or myxomatous stroma within the tumor [4]. The most commonly described features of GN in MRI include: well defined borders, encapsulation, homogeneously low or intermediate signal in T1-weighted and heterogeneously intermediate or high signal in T2-weighted images. This kind of tumor most often exhibits slight, heterogeneous contrast enhancement.

Radiological diagnosis of retroperitoneal GN is difficult, which is due to i.a. variable histological structure of the tumor. In the presented case mimicking of a renal tumor by GN posed an additional challenge. According to the available literature, ganglioneuromas is most often diagnosed on the basis of histopathological examination following tumor excision [1].

Resection is the best form of treatment of such lesions. Progression, late recurrences, or even malignant transformation of the tumor were observed in the past; thus, long-term radiological follow-up is recommended.

Conclusions

Ganglioneuroma of the retroperitoneal space is a rare, benign tumor, which may be difficult to differentiate from other lesions occurring in this localization. Radiological features characteristic for GN are often insufficient to establish unequivocal diagnosis. However, as shown by the presented case of a pararenal lesion, differential diagnosis of retroperitoneal tumors should include a ganglioneuroma.

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