Baldvinsdóttir et al., *Cogent Medicine* (2015), 2: 1115705
http://dx.doi.org/10.1080/2331205X.2015.1115705

cogent medicine

SURGERY | CASE REPORT

Solitary retroperitoneal ganglioneuroma

Bryndís Baldvinsdóttir[1]*, Jón G. Jónasson[2,3] and Rafn Hilmarsson[1]

Abstract: Retroperitoneal tumours rarely cause significant symptoms, therefore they are frequently diagnosed incidentally. Ganglioneuroma is a rare tumour that most often present in the posterior mediastinum or the retroperitoneal cavity. In the present case, a solitary ganglioneuroma is diagnosed in the retroperitoneum in an young healthy male, a rather typical presentation of this benign tumour.

Subjects: Oncologic Surgery; Urologic Surgery; Urologic Oncology

Keywords: Ganglioneuroma; Retroperitoneal tumour; Rare tumour

Received: 08 October 2015
Accepted: 28 October 2015
Published: 23 November 2015

*Corresponding author: Bryndís Baldvinsdóttir, Department of Urology, Landspítali University Hospital, Reykjavík, Iceland
E-mail: bryndisbaldvins@gmail.com

Reviewing editor:
Colin Johnson, University of Southampton, UK

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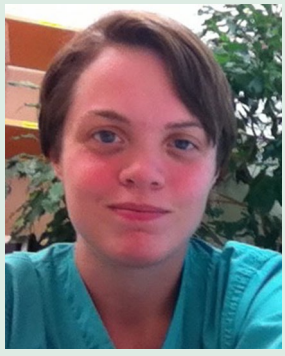
1. Introduction

Tumours located in the retroperitoneum are commonly diagnosed incidentally since they are frequently asymptomatic for a long time before giving rise to symptoms. Ganglioneuromas are rare tumours of the autonomic sympathetic nervous system, most often located in the mediastinum or the retroperitoneum. Their incidence in the general population is about one per million and they are most often sporadic (Singh et al., 2013; Vasiliadis et al., 2012; Zare, Parvin, & Ghohestani, 2008).

They are of neurogenic origin and composed of nerve fibres, ganglion cells and Schwann cells (Meng et al., 2013; Vasiliadis et al., 2012; Zare et al., 2008). They are surrounded by a pseudocapsule (Kattepura, Alexander, Kini, & Das, 2010). If available, a total surgical excision is advisable as the tumour can affect neighbouring vessels or organs. However, due to their tendency to grow around large vessels, a total excision is often very difficult. The tumours have homogenic appearance on CT and in about half of cases they have foci of calcifications (Tarantino, de Lacerda, Neto, Violante, & Vaisman, 2012). Since these tumours are most commonly benign and most often asymptomatic, observation is most often the standard management (Vasiliadis et al., 2012). This case is reported as a typical case of retroperitoneal ganglioneuroma, an exceedingly rare tumour.



Bryndís Baldvinsdóttir

ABOUT THE AUTHOR

Bryndís Baldvinsdóttir was born on 13 August 1986 in Akureyri, Iceland. She Studied medicine at the University of Iceland from 2006 to 2012 and did the internship at Landspitali University Hospital in Iceland from June 2012 to May 2013. She worked as a surgical resident at Landspitali University Hospital Iceland from June 2013 to May 2015. She is currently working at the neurosurgical department at the University Hospital in Uppsala, Sweden. Her former research work includes case reports in urology, general surgery and neurosurgery. This case report is a urological case report and is not thought to be related to another project

PUBLIC INTEREST STATEMENT

Retroperitoneal tumours rarely cause significant symptoms, therefore they are frequently diagnosed incidentally. Ganglioneuroma is a rare tumour that most often present in the posterior mediastinum or the retroperitoneal cavity. In the present case, a solitary ganglioneuroma was diagnosed in the retroperitoneum in an otherwise young healthy male, a rather typical presentation of this benign tumour. To diagnose this tumour, a laparoscopy was performed. We report this case as this tumour type is quite rare but the presentation and the case in general is quite typical and gives a good teaching value. It should be interesting for everyone interested in urology and surgery.

2. Case report

A 30-year-old previously healthy man went to see his doctor complaining of a back pain which he had been suffering from for a few months following a mild trauma (a fall) to the back. Following clinical examination, a computerized tomography (CT) was ordered. The CT showed a tumour in the retroperitoneal cavity at the level of the left renal vein. The tumour extended to the aorta and was reported to have calcified spicules (see Figure 1).

Blood tests were within normal limits, including beta hCG and alfa-fetoprotein.

A blood test showed normal levels of beta-hCG and alfa-fetoprotein. Other blood tests were all within normal limits.

In order to obtain a diagnosis of this tumour, a CT-guided core needle biopsy was performed. This only showed muscle tissue without any signs of a tumour. Therefore, another CT-guided biopsy was performed. This gave the same results as the previous biopsy. In order of obtaining a reliable diagnosis of this tumour, a laparoscopy was performed. An open solid tumour biopsy was obtained (Figure 2). The pathology examination rendered a firm diagnosis of a differentiated ganglioneuroma (Figure 3). The pathology showed scattered differentiated ganglion cells as well as neurofibroma-like tissue exhibiting small foci of calcification. Due to the benign nature of the tumour, no further treatment is planned. The patient will have follow-up CT about 6 months after the diagnosis and follow-up is scheduled with control CT's in the future.

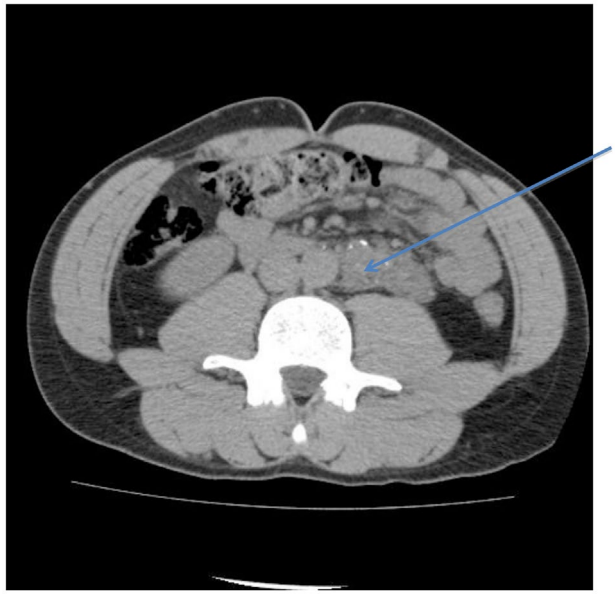


Figure 1. The computed tomography showed a tumour (arrow) adjacent to the aorta.

Note: There were calcifications in the tumour.

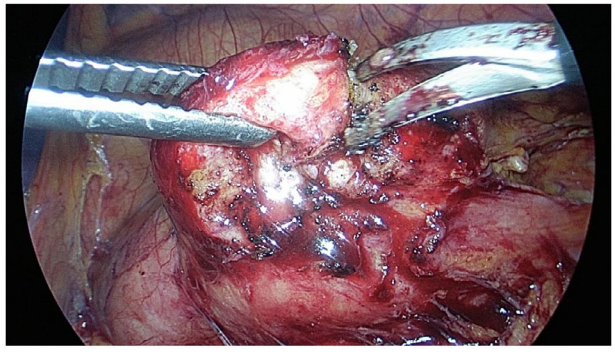


Figure 2. Picture from the laparoscopy.

Note: The tumour had a pseudocapsule and a good biopsy was obtained for histopathological examination.

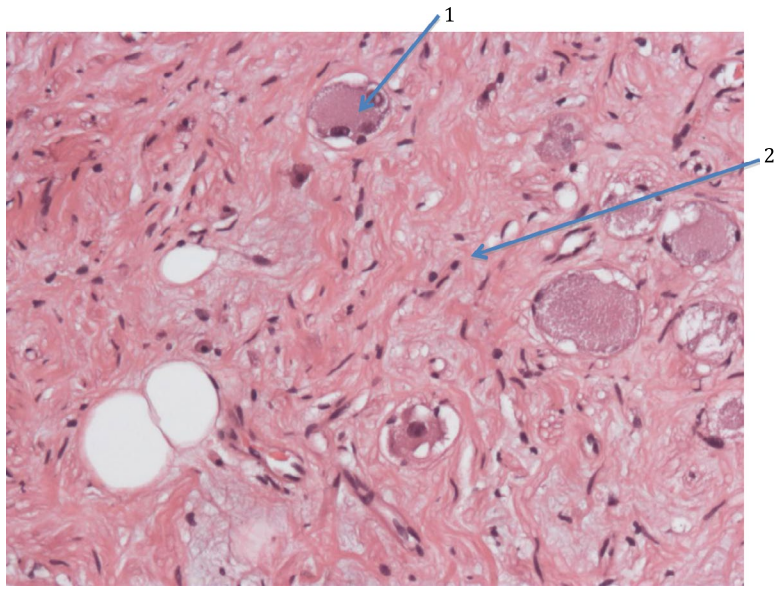


Figure 3. The histopathological examination showed a differentiated ganglioneuroma.

Notes: Arrow 1 showing ganglion cells. In between, uniform spindle cells predominated (arrow 2).

3. Discussion

Retroperitoneal tumours are most often diagnosed as incidental findings, due to anatomic location and late presentation of symptoms. Sarcomas are about one-third of all retroperitoneal tumours. Other types of retroperitoneal tumours include e.g. lipomas, liposarcoma, lymphoma, other rare mesenchymal sarcomas, renal angiomyolipomas, benign neurogenic tumours, metastatic testicular carcinoma, etc. (Strauss, Hayes, & Thomas, 2011).

In the case presented the most likely differential diagnoses, clinically, was liposarcoma, teratoma or metastatic testicular cancer. Two core needle biopsies didn't provide a diagnosis. Therefore, a diagnostic laparoscopy was undertaken following which a differentiated ganglioneuroma was firmly diagnosed on an open biopsy obtained at operation. In the present case, the tumour was located in the retroperitoneum, adjacent to the aorta and actually surrounded the aorta.

This kind of behaviour is characteristic of ganglioneuromas (Cocieru & Saldinger, 2011). These tumours are benign and classified in the group of peripheral nerve sheath tumours, a group of tumours which is diverse and not always with a well-documented behaviour. Therefore, a regular follow-up is necessary in all patients with ganglioneuroma (Cocieru & Saldinger, 2011).

The obvious benefit with needle biopsy is the fact you can get a diagnosis without the patient undergoing a surgery. The risk with a biopsy in this particular case lies in the chance of hitting the aorta while obtaining the specimen. The benefit of laparoscopic surgery is that it's a good chance of getting a good specimen that's likely to give a definitive diagnosis. The risk, as with all surgeries, is of that of damaging adjacent structures, i.e. nerves, veins and arteries, and the risk of too much blood loss, infection and the risks that follow general anaesthesia.

The presented case is interesting due to the rarity of this tumour type, it emphasizes the typical delay in obtaining a correct diagnosis and the incidental finding in an otherwise healthy young man. All of this is what can be expected of a differentiated ganglioneuroma originating in the retroperitoneum. What we can learn from this case is to keep this tumour type in mind when patients present with atypical back pain. It also underlines how good well performed laparoscopy is as a diagnostic tool.

Funding
The authors received no direct funding for this research.

Competing interests
The authors declare no competing interest.

Author details
Bryndís Baldvinsdóttir[1]
E-mail: bryndisbaldvins@gmail.com
Jón G. Jónasson[2,3]
E-mail: jongj@landspitali.is
Rafn Hilmarsson[1]
E-mail: rafnhi@landspitali.is
[1] Department of Urology, Landspítali University Hospital, Reykjavík, Iceland.
[2] Department of Pathology, Landspítali University Hospital, Reykjavík, Iceland.
[3] Faculty of Medicine, University of Iceland, Reykjavík, Iceland.

Citation information
Cite this article as: Solitary retroperitoneal ganglioneuroma, Bryndís Baldvinsdóttir, Jón G. Jónasson & Rafn Hilmarsson, *Cogent Medicine* (2015), 2: 1115705.

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***Cogent Medicine* (ISSN: 2331-205X) is published by Cogent OA, part of Taylor & Francis Group.**