Castleman's disease – an unusual indication for laparoscopic adrenalectomy

Maciej Otto[1], Łukasz Wieprzowski[1], Jacek Dzwonkowski[1], Bogna Ziarkiewicz-Wróblewska[2]

[1]Department of General, Vascular and Transplantation Surgery, Medical University of Warsaw, Poland
[2]Department of Pathology, Medical University of Warsaw, Poland

Videosurgery and Other Miniinvasive Techniques 2012; 7 (1): 50-54
DOI: 10.5114/wiitm.2011.25620

Abstract

Castleman's disease is one of the rare entities which cause lymph node hyperplasia with no characteristic clinical symptoms. Pathomorphological examination usually enables diagnosis, especially in the case of a localized form. Its uncommon location in the retroperitoneal cavity is estimated at 12% in the literature. Asymptomatic nature of the disease and its untypical location in the adrenal field imitated incidentaloma of this gland in the imaging examinations. Surgical treatment, laparoscopic excision of the tumour (lymph node) and right adrenal gland from the lateral, transperitoneal approach allowed final diagnosis and offered effective therapy.

Key words: *laparoscopy, adrenalectomy, adrenal incidentaloma, Castleman's disease.*

Introduction

Laparoscopic adrenalectomy, especially from lateral, transperitoneal access, has become a reference procedure in adrenal gland disorders [1-4]. However, the size of the lesion selected for such a procedure is still under discussion [5-7]. At the same time, the definition of incidentaloma phenotype remains a continuing problem which facilitates the decision about mini-invasive surgical treatment where the diameter of ≥ 4 cm has been an invariable indication. Among adenomas, which are the most common cause of incidentaloma, other rare causes can be found where surgical treatment enables final diagnosis and plays an important role in successful therapy. Adrenal cortical carcinoma, angiosarcoma and oncocytoma are those causes we learn about only after histopathological data are obtained [8-11]. The described case of Castleman's disease is one of them. The goal of this study is to present doubts and difficulties concerning selection for laparoscopic adrenalectomy based on the case of a patient with incidentaloma, which turned out to be Castleman's disease. It is a rare cause of unclear aetiology leading to lymph node hyperplasia. It was first described in 1954 by Castleman in a series of 13 patients with limited mediastinal lymph node hyperplasia. In the literature it is also known as angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, giant benign lymphoma or lymphoid hamartoma [12-14].

Case report

Incidentaloma of the right adrenal gland with the diameter of 4-5 cm was observed in a 33-year-old patient in a USG examination performed due to a medical check-up at work. According to the patient's medical history, he did not suffer from any ailment. Physical examination did not reveal any aberrations. Routine hormone tests did not confirm

Address for correspondence
Prof. Maciej Otto MD, PhD, Department of General, Vascular and Transplantation Surgery, Medical University of Warsaw, 1a Banacha, 02-097 Warsaw, Poland, phone: +48 22 599 24 67, e-mail: maciej.otto@wum.edu.pl

endocrine activity of the tumour. Computed tomography with and without contrast confirmed a regularly shaped tumour of the right adrenal gland with the diameter of 5 cm with small foci of enhanced density (Figures 1 and 2). However, the imaging results suggested the location of the tumour just above the upper pole of the adrenal gland. Due to doubts concerning the tumour's morphology and its location, magnetic resonance (Figure 3) and positron emission tomography/computed tomography (PET/CT) (Figure 4) were additionally performed. They confirmed doubts concerning the location of the lesion. The findings determined the presence of the lesion directly beside the right adrenal gland, just above its upper pole. The patient was selected for surgical treatment with the preliminary diagnosis of incidentaloma without hormonal activity.

However, silent pheochromocytoma, a clinically silent but biologically active tumour, could not be excluded due to foci of increased density, especially as, although the methoxycatecholamines double measurements showed no aberrations, minor excess values of metanephrine were observed. That is why the patient received 10-day α-blocker (the producer's name – cardure, 4 mg) therapy as preoperative preparation. Preoperative tests revealed a slight increase in the number of leukocytes (11.98 × 10^3/μl), a little lower level of total protein, platelets at the

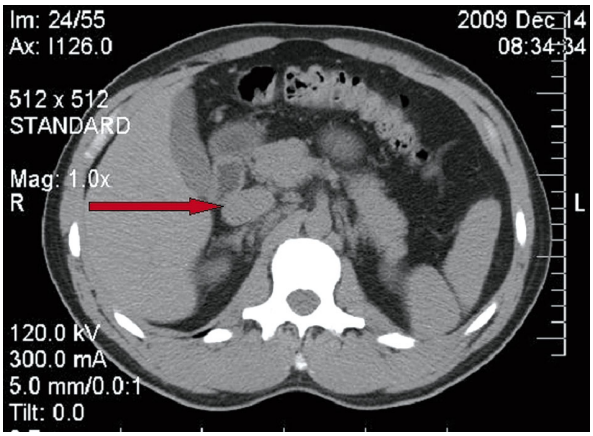

Figure 1. Computed tomography without contrast (5 cm regular tumour)

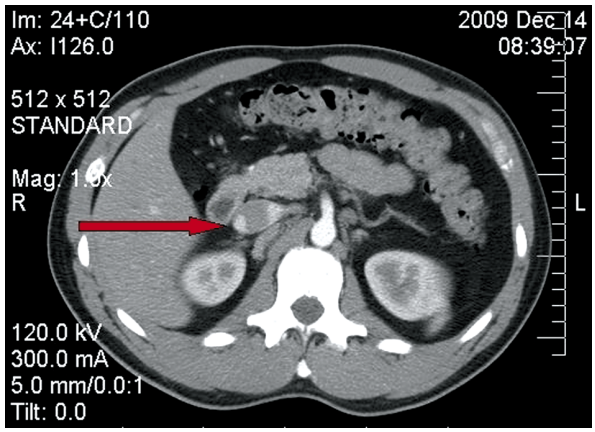

Figure 2. Computed tomography without contrast revealed small foci of enhanced density within the tumour

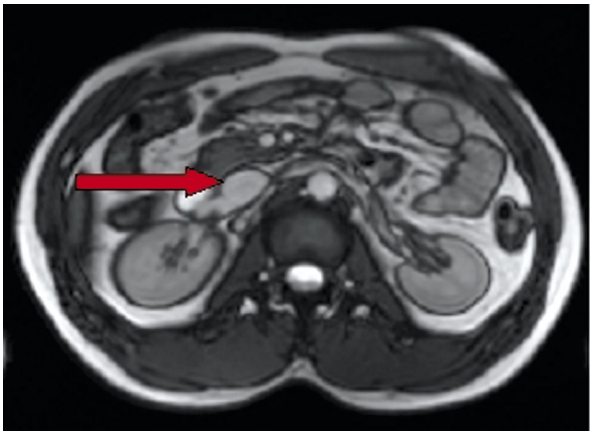

Figure 3. Magnetic resonance imaging confirmed the size of the lesion and its location in the upper pole of the right adrenal gland, although it suggested location outside the tumour

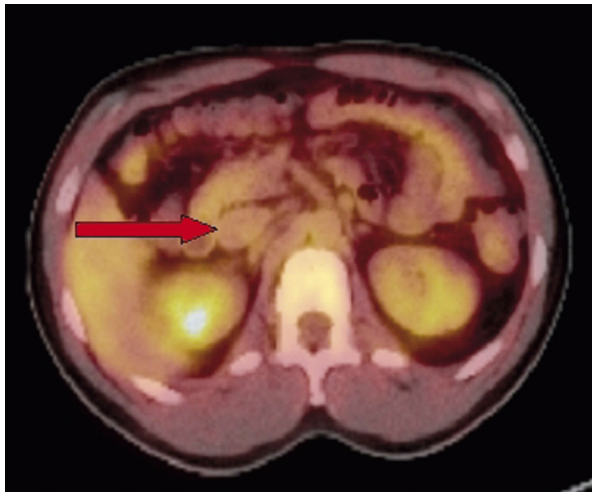

Figure 4. Positron emission tomography-computed tomography scan confirmed the lesion directly next to the right adrenal gland

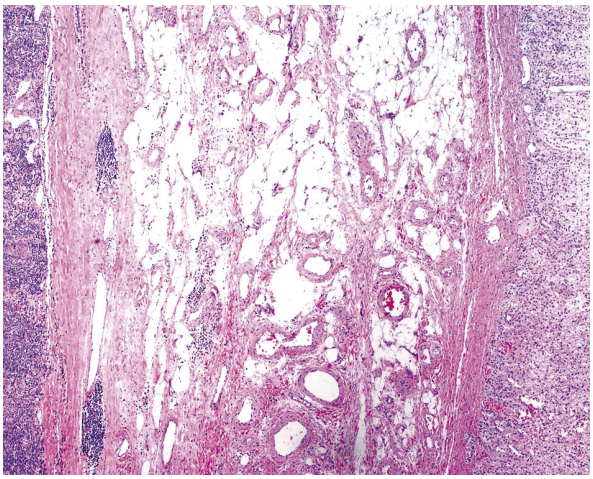

Figure 5. H&E staining. Objective magnification 4×. On the left, lymph node capsule with the narrow strip of adenoma tissue separated from the adrenal gland (right side) with fat tissue

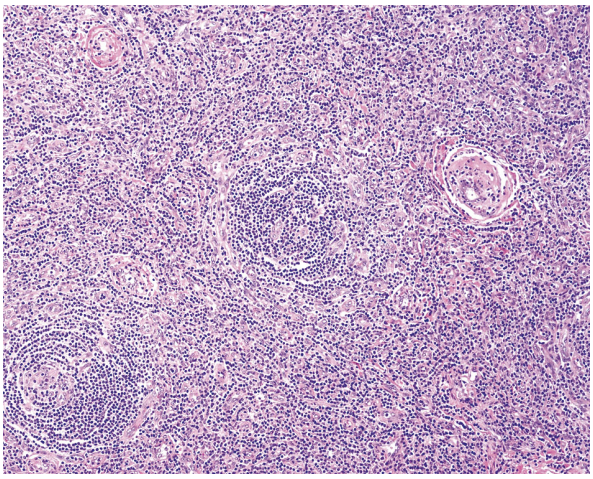

Figure 6. H&E staining. Objective magnification 10×. Small, obolescent foci of hyalinized adenoma clumps

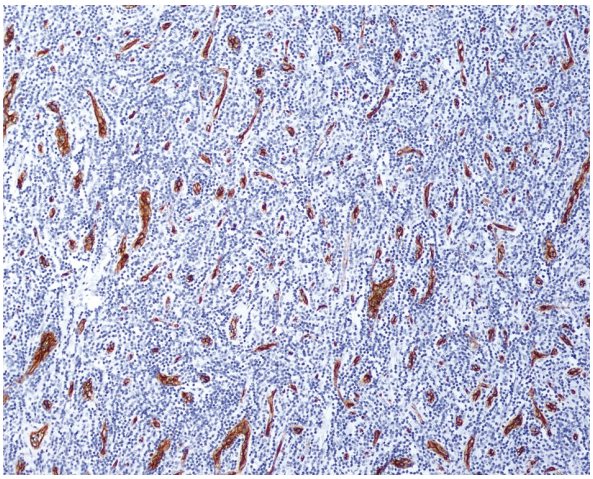

Figure 7. Immunohistochemical staining with CD34. Objective magnification 10×. Numerous small vessels

level of 135 × 10^3/µl and elevated level of creatinine (1.12 mg/dl). The rest of the test parameters were within the normal limits. On admission, heartbeat was at 84/min, blood pressure at 110/80 mmHg. The patient was operated on laparoscopically in the lateral, transperitoneal approach. A tumour hermetically joined with the liver capsule, lowering beyond the inferior caval vein, located directly above the right adrenal pole was observed intraoperatively. The right adrenal gland was slightly enlarged. Having been pushed by the tumour, it covered the adrenal vein. The tumour was spherical, irregular in the upper pole. The rest of the tumour, however, was encapsulated. The lesion beyond the inferior caval vein reached its medial edge. For the safe dissection of the tumour from the caval vein, it was necessary to remove the adrenal gland to obtain a better view. The isolation of the adrenal vein was difficult due to its "fusion" with the hard infiltration between the tumour and caval vein. The tumour was removed together with the adrenal gland. The drain was placed in the adrenal field. The postoperative course was uncomplicated. The drain was removed from the peritoneal cavity on the 1st postoperative day. Steroid substitution therapy was employed. The patient was released on the 4th postoperative day. Histological examination revealed the adrenal gland with the attached tumour sized 4 cm × 2.5 cm × 4.5 cm. Microscopically, the tumour proved to be a lymph node with hyaline-vascular variant of Castleman's disease (Figures 5-7).

Discussion

Castleman's disease is a rare diagnosis in departments all over the world. This proves that it remains detectable at relatively low levels, which is connected with few symptoms and little knowledge about it. In the literature small series of patients are described and optimal treatment is not definitely determined. There are a few theories attempting to explain the causes of morphological changes in the patients' lymph nodes. Some of the concepts include viral

infection, immunological factor, resistance disorder and chronic inflammation [13, 15]. The multitude of ideas suggests a disease with a complex aetiology and each of the concepts may be correct to some extent. The role played by interleukin 6 in the pathomechanism of hypertrophic lymph nodes (both types – localized and multicentric), which stimulates B lymphocyte proliferation and maturation, is emphasized. Its presence within the circulation causes constitutional symptoms of the disease such as fever, weakening and anaemia [15]. Clinically, localized and multicentric types of the disease can be distinguished [15]. The localized variant is characteristic of young people. Lesions which are unifocal occur mainly in the mediastinum [16]. The multicentric type of Castleman's disease, in contrast, is diagnosed in older patients and its course is more severe. In 1969, Flendring and Schilling identified and suggested the division of the disease into two histological variants: hyaline vascular and plasma cell type [17]. A few years later, Kellner *et al.* also described a mixed type of the disease [16]. The histological picture, however, does not always allow for unequivocal diagnosis since similar lesions in the lymph nodes can be observed in Sjögren's syndrome, rheumatoid arthritis, cancer spreading via the lymphatic system, iatrogenic immunosuppression, and congenital or acquired immunodeficiency [13, 18, 19]. The hyaline vascular variant of Castleman's disease is thought to be benign and asymptomatic. Treatment consists of radical surgery to remove the altered lymph node, and the 5-year survival rate reaches 100%. Altered lymph nodes are usually located in the anterior mediastinum (70%), and outside the chest, lesions occur in the cervical lymph nodes (14%), mesenteric and retroperitoneal lymph nodes (12%), axillary lymph nodes (4%), the central nervous system, eye sockets, pelvis and skeletal muscles [13]. The multicentric form of Castleman's disease is currently believed to be a potentially malignant lymphatic hyperplasia associated with POEMS: polyneuropathy, organomegaly, endocrinopathy, monoclonal protein and skin lesions [20, 21]. This type is associated with various immunological disorders, systemic symptoms, aggressive course and poor prognosis. The mortality rate is around 50% and the survival rate is about 26 months. Treatment is intended to be combined with chemotherapy, steroid therapy and radiotherapy, but the optimal treatment is not entirely determined [20, 22]. The majority of cases are observed in the 5th decade of life and women are prevalent among those diagnosed. The present study proves that despite access to a modern, specialist diagnostic base, sometimes it is difficult to determine the character and location of a tumour [3, 9, 11, 23]. On the other hand, it shows that a lateral, transperitoneal approach to laparoscopic adrenalectomy is universal in the case of adrenal lesions. It allows a suitable location for trocars entry, dependent on anatomical conditions, and as a result provides an extensive and safe surgical field. These possibilities confirm that the method is most exemplary in the treatment [1, 5, 7]. The proven advantages and safety of the videoscopic surgery concurred with the constant development and introduction of new technical elements, which made the surgery more attractive, but most importantly less traumatic. Single, percutaneous incision LA (SILS) is one of them [24-26]. However, despite positive results, the technique can be used in the case of so-called small tumours and cases with high probability of initial and final diagnosis consistency.

Conclusions

Castleman's disease is a rare cause of lymph node hypertrophy, which is why it is often overlooked when diseases with such symptoms are diagnosed. It is necessary to remember about the possibility of its occurrence and take it into consideration when eliminating the most common causes of lymph node enlargement. The proper cooperation between the clinician and pathologist allows for early diagnosis and suitable therapy. It also proves that this cause should be considered when possible causes of adrenal incidentalomas are differentiated.

References

1. Smith CD, Weber CJ, Amerson JR. Laparoscopic adrenalectomy: new gold standard. World J Surg 1999; 23: 389-96.
2. Ogilvie JB, Duh QY. New approaches to the minimally invasive treatment of adrenal lesions. Cancer J 2005; 11: 64-72.
3. Otto M, Szostek G, Nazarewski S, et al. Laparoscopic transperitoneal lateral adrenalectomy – method and intraoperative difficulties. Videosurgery 2002; 7: 15-20.
4. Kasperlik-Załuska AA, Otto M, Cichocki A, et al. 1,161 patients with adrenal incidentalomas: indications for surgery. Langenbecks Arch Surg 2008; 393: 121-6.
5. Parnaby CN, Chong PS, Chisholm L, et al. The role of laparoscopic adrenalectomy for adrenal tumors of 6 cm or greater. Surg Endosc 2008; 22: 617-21.

6. Gumbs AA, Gagner M. Laparoscopic adrenalectomy. Best Pract Res Clin Endocrinol Metab 2006; 20: 483-99.
7. Tsuru N, Suzuki K, Ushiyama T, Ozono S. Laparoscopic adrenalectomy for large adrenal tumors. J Endourol 2005; 19: 537-40.
8. Kasperlik-Załuska AA, Otto M, Cichocki A, et al. Incidentally discovered adrenal tumors: a lesson from observation of 1,444 patients. Horm Metab Res 2008; 40: 338-41.
9. Kebebew E, Siperstein AE, Clark OH, Duh QY. Results of laparoscopic adrenalectomy for suspected and unsuspected malignant adrenal neoplasms. Arch Surg 2002; 137: 948-53.
10. Lombardi CP, Raffaelli M, De Crea C, Bellantone R. Role of laparoscopy in the management of adrenal malignancies. J Surg Oncol 2006; 94: 128-31.
11. Otto M, Dzwonkowski J, Jędrasik M, et al. Trudności kwalifikacyjne i operacyjne adrenalektomii laparoskopowej. Pol Przegl Chir 2010; 82: 64-78.
12. Castleman B, Iverson L, Menendez VP. Localized mediastinal lymphnode hyperplasia resembling thymoma. Cancer 1956; 9: 822-30.
13. Frizzera G. Castleman's disease: more questions than answers. Hum Pathol 1985; 16: 202-5.
14. Wilczak M, Kampioni M, Szmeja J. Castleman's disease – a case report. Videosurgery and Other Miniinvasive Techniques 2011; 6: 27-32.
15. Isaacson PG. Castleman's disease. Histopathology 1989; 14: 429-32.
16. Keller AR, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. Cancer 1972; 29: 670-83.
17. Flendrig JA, Schillings PHM. Benign giant lymphoma: the clinical signs and symptoms. Folia Medica Neerlandica 1969; 12: 119-20.
18. Yoshizaki K, Matsuda T, Nishimoto N, et al. Pathogenic significance of interleukin-6 in Castleman's disease. Blood 1989; 74: 1360-7.
19. Francis ND, Hollowood K, Gabriel R. Angiofollicular lymph node hyperplasia. J Clin Pathol 1988; 41: 353-4.
20. Peterson BA, Frizzera G. Multicentric Castleman's disease. Semin Oncol 1993; 20: 636-47.
21. Wang LY, Jiang TA, Teng XD, et al. Multicentric hyaline-vascular Castleman's disease in the retroperitoneum. Hepatobiliary Panreat Dis Int 2009; 8: 554-7.
22. Kisch CF, Webb EM, Webb WR. Multicentric Castleman's disease and POMES syndrome: CT findings. J Thorac Imaging 1997; 12: 75-7.
23. Myśliwiec P, Dadan J, Łukaszewicz J, Sulik M. Two tumors of the right adrenal gland treated videoscopically by the posterior retroperitoneal approach. Videosurgery and Other Miniinvasive Techniques 2009; 4: 126-30.
24. Łosin M, Czauderna P, Gołębiewski A, Stefanowicz J. Single incision laparoscopic adrenalectomy – initial experience. Videosurgery and Other Miniinvasive Techniques 2010; 5: 104-6.
25. Budzyński A, Pędziwiatr M, Natłok M, et al. Preliminary experience with transperitoneal single incision laparoscopic surgery adrenalectomy. Videosurgery and Other Miniinvasive Techniques 2010; 5: 87-92.
26. Cywiński J, Kuzdak K, Kołomecki K. One-incision approach (SILS) for retroperitoneal videoscopic adrenalectomy. Videosurgery and Other Miniinvasive Techniques 2010; 5: 70-1.