Castleman’s disease – an unusual indication for laparoscopic adrenalectomy

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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.

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 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].

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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 × 10³/μl), a little lower level of total protein, platelets at the
level of 135 × 10³/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

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
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, Flandring 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.

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