

Castelman's disease of the neck – case report

Choroba Castelmana zlokalizowana na szyi – opis przypadku

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SUMMARY:

Castelman's disease is a rare lymphoproliferative disorder most commonly diagnosed in the thorax. We present a case of Castelman disease localized in the neck, confirmed by histopathology. The patient manifested symptoms associated with compression of the brachial plexus on the right.

KEYWORDS:

Castelman's disease, head and neck, lymphoproliferative disease

STRESZCZENIE:

Choroba Castelmana jest rzadkim limfoproliferacyjnym schorzeniem najczęściej diagnozowanym w klatce piersiowej. Przedstawiamy przypadek choroby Castelmana zlokalizowanej na szyi, potwierdzonej badaniem histopatologicznym, manifestującej się dolegliwościami związanymi z uciskiem na splot barkowy po stronie prawej.

SŁOWA KLUCZOWE: choroba Castelmana, głowa i szyja, choroba limfoproliferacyjna

INTRODUCTION

Castelman's disease – angiofollicular lymph node hyperplasia, benign giant lymphoma, lymphoid hamartoma, is a rare non-neoplastic lymph node hyperplasia of unknown etiology [1, 2, 3]. This disease is most frequently diagnosed within the thorax and its course is often asymptomatic [4, 3]. We report a case of a 26-year-old female with Castleman's disease located in the neck. The course of many diseases of autoimmune or viral origin, as well as neoplastic diseases is similar to this of Castleman's disease. Similar changes in histopathological picture accompany Hodgkin's disease, IgG4-dependent disease [5, 6, 7]. *Human herpesvirus 8* (HHV-8), Epstein-Barr virus (EBV) and *Herpesvirus saimiri* [8] may play a significant role in the pathogenesis of Castleman's disease. Surgical procedure is the treatment method of choice that ensures full recovery in unicentric type cases [9]. Multicentric form may transform into

malignant lymphoid hyperplasia. This type of the disease may have aggressive course, bad prognosis. Optimum treatment in multicentric form of the disease has not been determined but there are reports describing the use of chemotherapeutic treatment with cyclophosphamide and azathioprine [1, 10].

CASE REPORT

A 26-year-old female patient was admitted to the Department due to neck tumor. The tumor had been enlarging gradually for 3 years, according to the patient. At the beginning it was asymptomatic. Over time symptoms resulting from pressure to brachial plexus nerves developed that were increasing. Glossopharyngeal nerve paresis had also developed 3 months before the patient was admitted to the Department. Moreover, the patient reported hyperhidrosis, itching and flushing in the



Fig. 1. Computed tomography in coronal plane. A tumor of posterior cervical space modelling and displacing the internal jugular vein is visible in the scan.



Fig. 3. Computed tomography in the coronary plane.



Fig. 2. Computed tomography in the sagittal plane.

face and neck. Computed tomography of the neck was performed in which a tumor of approximately 7 cm in diameter located in the posterior cervical space, which caused modeling and displacement of the internal jugular vein to the front (Fig. 1, 2, 3). The result of fine-needle aspiration biopsy was incon-

clusive and revealed the presence of reactive lymph node cells. The patient was qualified for surgical removal of the tumor under general anesthesia. A tumor in a strong and direct contact with the cervical and brachial plexus on this side was diagnosed intraoperatively. The tumor, removed totally, was 7.5x5 cm in diameter. It was encapsulated and had a yellowish hue, which was visible after cutting through the tumor. Tissue pattern of hyperplastic lymph nodes with picture corresponding to hyaline vascular type of Castleman's disease was determined in histopathological examination. The patient underwent endocrinological and immunological consultation. Diagnostics of the immune system was performed together with chest CT, and none of those revealed pathology. The patient did not require further treatment. Mobility of the upper extremity in the brachial joint recovered during three-month rehabilitation period. The patient remains under periodical laryngological follow-up. After twelve-month observation no recurrence was observed.

DISCUSSION

Castleman's disease is a rare disorder that was described for the first time in 1954 by Castelman et al. in 13 patients with localized mediastinal lymph node enlargement [11]. Approximately 50% of the cases are located in thorax along the tracheobronchial tree, in mediastinum or the hilum of the lung. It can also be located outside thorax, e.g. in the neck, axillary or retroperitoneal lymph nodes, skeletal muscles, orbits, peripheral

nerve system, lungs, salivary glands and pancreas [4, 11]. The etiology of the disease is unknown. Theories that explain the development of disorders that accompany Castelman's disease include chronic inflammatory processes, autoimmunological processes, immunodeficiency, *Epstein-Barr* virus infection, *Toxoplasma gondii* (protozoan) infection or *Mycobacterium tuberculosis* (bacterium) infection [1]. Relationship between Kaposi's sarcoma and Castleman's disease was proven in patients with Human Immunodeficiency Virus (HIV) infection [8,12]. Other accompanying disorders include e.g. hemolytic anemia, multiple myeloma and pemphigus [1]. Follicular dendritic cell dysplasia, cytokine dependence (especially interleukin-6 dependence) and oversecretion of adhesive molecules [2,8,13] are also considered in the etiology of the disease. Incorrect reaction to chronic inflammatory processes may be the cause that leads to the development of discussed pathology [19]. Patients usually remain asymptomatic or symptoms are non-specific. Castleman's disease can be accompanied by various immunological disorders, body weight loss, skin itching, anemia, and hypergammaglobulinemia. Also peripheral edema and, in 20% cases, peripheral neuropathy, may develop [6]. A few classifications of Castleman's disease were proposed. Clinically, two types of the disease are distinguished: unicentric and multicentric [14]. The unicentric type usually develops in young patients. The lesions are unifocal. The multicentric form develops more frequently in older patients and poses a greater risk for the development or coexistence of neoplasms, also Kaposi's sarcoma, non-Hodgkin lymphoma or Hodgkin lymphoma; it can be accompanied by hepatosplenomegaly as well by *Herpes* (HHV-8) virus infection [1,15]. The second classification was introduced according to the occurrence of the disease in HIV-positive and HIV-negative patients [16,17]. Higher IL-6 interleukin levels are also observed in these patients [1, 18]. Three histological disease types were distinguished: hyaline vascular type, plasma-cell type, and the mixed type [11, 19]. Hyaline vascular type is the most frequent one (incidence is estimated at approximately 91%), the other cases are comprised by plasma-cell and mixed type (approximately 9%) [11].

Flendring and Schilling described 2 histopathological types, and Kelle et al. described the mixed type in 1972 [1, 7, 20].

Preoperative diagnosis is difficult as there are no specific symptoms or radiographical features. Diagnostic imaging procedures that should be performed in the diagnostics of this disease include CT, MRI, in some cases also PET. Aspiration biopsy does not provide satisfactory results and is connected with high hemorrhage risk because of significant hypervascularity component [21]. Final diagnosis is usually made after surgical removal of the tumor. Our differential diagnosis included liposarcoma, atypical lipoma, neurogenic tumors, vascular tumors and teratomas. Histopathological changes similar to Castleman's disease are described for rheumatoid arthritis, Sjögren syndrome, phenytoin hypersensitivity, iatrogenic immunosuppression, immunodeficiency, as well as for Hodgkin's lymphoma and IgG4-dependent disease [1]. If total tumor resection is impossible, partial tumor removal or radiotherapy may be helpful in preventing the inflammatory process from becoming multicentric [5, 22]. Patients with multiple structures involved should be disqualified from surgical treatment and should be considered candidates for a therapy that combines glucocorticosteroids, chemotherapy and biological treatment [21]. Surgical resection enables recovery and prevents from the development of lymphoma in 90-95% of cases. Optimum treatment method has not been determined up to date [1, 10, 22].

CONCLUSIONS

Ambiguous diagnostic imaging and fine-needle aspiration biopsy results motivate head and neck surgeons to remove the tumor totally and make final diagnosis after they receive histopathological test results

Diseases of autoimmune origin, including Castleman's disease, should be taken into consideration in the diagnostics of neck tumors

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