https://doi.org/10.56871/PED.2025.64.80.008 UDC 616.428-07-003.822-092+616-053.4-055.2

A rare case of unilateral location of cervical localization Castleman's disease in a five-year child

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ABSTRACT

Castleman's disease was first described as a separate ailment by Castleman in 1956, as a non-neoplastic lesion of the lymph nodes located in the mediastinum, histologically unique unlike any other disease. The pathogenesis of this disease remains unclear. It has now been proven that polyclonal non-neoplastic lymphocytes are the substrate. An imbalance of interleukin-6 is thought to play a significant role in the development of the disease. The presence of human herpesvirus 8 (HH8) is also significant.

A five-year-old girl's mother has brought her to the Pediatric University Consultative and Diagnostic Center with complaints of a mass on the right side of her neck. The child's medical history revealed that the mother had noticed the mass approximately two years earlier, despite her otherwise healthy condition, and that the mass had gradually enlarged. An outpatient diagnosis of cervical lymphadenopathy was made, and follow-up was recommended. Due to pain in the area of the mass, the girl's mother presented to the Pediatric University Consultative and Diagnostic Center to discuss further treatment.

Following an examination, a decision was made to perform elective surgery, which revealed a 5.5 cm diameter lesion with smooth contours. Histological examination of the removed lesion resulted in a diagnosis of Castleman disease, hyaline-vascular form. The child reported no complaints during follow-up examinations. A follow-up ultrasound of the soft tissues of the neck and abdomen revealed no abnormalities. Clinical and biochemical blood tests were unremarkable. The patient was recommended to have a clinical blood test and ultrasound of the soft tissues of the neck once a year.

This case will be of interest to otolaryngologists, surgeons, and pediatricians, as lymphadenopathy is a nonspecific and common symptom that doctors of various specialties encounter daily.

Keywords: cervical lymphadenopathy, lymphadenopathy, lymphoproliferative disease

To cite this article

Chernogaeva E.A., Pavlov P.V., Medvedeva N.A., Pendzhieva P.R. A rare case of unilateral location of cervical localization castleman's disease in a five-year child. *Pediatrician (St. Petersburg).* 2025;16(4):75–80. https://doi.org/10.56871/PED.2025.64.80.008.

Submitted: 08.09.2025 **Accepted: 26.10.2025** **Published online: 05.12.2025**

ISSN 2587-6252 (Online)

https://doi.org/10.56871/PED.2025.64.80.008 УДК 616.428-07-003.822-092+616-053.4-055.2

Редкий случай одностороннего варианта шейной локализации болезни Кастельмана у ребенка 5 лет

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АННОТАЦИЯ

Впервые болезнь Кастельмана как отдельная патология была описана Кастельманом в 1956 году как несходное гистологически ни с одним другим заболеванием неопухолевое поражение лимфоузлов, локализованных в средостении. Патогенез данного заболевания до конца неясен. В настоящее время доказано, что субстратом являются поликлональные неопухолевые лимфоциты. Большую роль в развитии заболевания отводят нарушению баланса интерлейкина-6. Также большое значение имеет наличие в организме вируса герпеса 8-го типа (*Human herpesvirus 8, HH8).*

В консультативно-диагностический центр педиатрического университета обратилась мать девочки пяти лет с жалобами на наличие у ребенка образования на боковой поверхности шеи справа. Из анамнеза известно, что образование было замечено около двух лет назад на фоне полного здоровья, и оно постепенно увеличивалось. Амбулаторно установлен диагноз «лимфаденопатия шеи», рекомендовано динамическое наблюдение, в течение которого в области образования появилась болезненность.

После выполненного обследования было принято решение о выполнении планового хирургического лечения. В результате было удалено образование диаметром 5,5 см, с ровными контурами. При гистологическом исследовании установлен диагноз: Болезнь Кастельмана, гиалиново-васкулярная форма. На повторных осмотрах жалоб ребенок не предъявлял. На контрольном ультразвуковом исследовании (УЗИ) мягкие ткани шеи и брюшной полости без патологии. Клинический и биохимический анализы крови без особенностей. Пациентке рекомендованы контроль клинического анализа крови и УЗИ мягких тканей шеи 1 раз в год.

Данный случай представляет интерес для оториноларингологов, хирургов, педиатров, так как лимфаденотопатия является неспецифическим и часто встречающимся симптомом, с которым ежедневно сталкиваются врачи разных специальностей.

Ключевые слова: шейная лимфаденопатия, лимфаденопатия, лимфопролиферативное заболевание

Как цитировать

Черногаева Е.А., Павлов П.В., Медведева Н.А., Пенджиева П.Р. Редкий случай одностороннего варианта шейной локализации болезни Кастельмана у ребенка 5 лет. *Педиатр*. 2025;16(4):75–80. https://doi.org/10.56871/PED.2025.64.80.008.

Рукопись получена: 08.09.2025 **Рукопись одобрена: 26.10.2025** **Опубликована online: 05.12.2025**

ISSN 2079-7850 (Print)

Castleman's disease is a condition characterized by angiofollicular hyperplasia of the lymph nodes, with an unpredictable clinical course, association with a wide spectrum of other diseases, and a high risk of transformation into lymphoma [1–3]. The disease was first described by Castleman in 1956 as a non-neoplastic lesion of the lymph nodes located in the mediastinum, which was histologically dissimilar to any other known disease. Initially, the pathology was interpreted as an ectopic thymus, teratoma, dermoid cyst, or non-Hodgkin's lymphoma. Subsequently, none of these theories were confirmed.

Later, it was established that the tumor location can vary. The most common site is the mediastinum (70% of cases), followed by the cervical lymph nodes (15–20%), and less frequently the axillary and retroperitoneal lymph nodes. According to some data, mediastinal and cervical lymph nodes are affected with equal frequency [1, 2, 4]. The rarest sites of involvement are the nasopharynx, orbit, and tongue.

The pathogenesis of this disease remains incompletely understood. It has now been established that its substrate consists of polyclonal, non-neoplastic lymphocytes. A major role in the disease development is attributed to an imbalance of interleukin-6 (IL-6), which is involved in the pathogenesis of numerous autoimmune diseases as well as in the development of systemic inflammatory and neoplastic processes. The presence of the *HHV-8* virus in the body is also of significant importance [1, 2].

A number of studies have demonstrated a direct correlation between the presence of symptoms in Castleman's disease and the level of IL-6 expression [1]. Another study established that Castleman's disease is the only condition in which marked expression of IL-6 is detected within the affected follicles of the lymph nodes [2].

Based on the clinical course, localized (unicentric) and generalized (multicentric) forms are distinguished. The unicentric form typically remains asymptomatic for a prolonged period, grows slowly, and has a favorable clinical course following radical surgical excision.

The generalized form has an unfavorable clinical course and may present with periodic fevers, weight loss, skin changes, organomegaly, elevated erythrocyte sedimentation rate (ESR), alanine aminotransferase (ALT), aspartate aminotransferase (AST), and anemia. It is often this form that is associated with the presence of *HHV-8* and HIV infection. Accordingly, the disease is classified into *HHV-8*-positive and *HHV-8*-negative forms. The *HHV-8*-positive variant is more frequently observed in immunocompromised HIV-positive patients. The combination of an *HHV-8*-positive variant and HIV infection has an unfavorable, malignant clinical course. This form is often accompanied by *POEMS* syndrome, characterized by polyneuropathy, organomegaly, endocrinopathy, and monoclonal gammopathy, as well as amyloidosis, multiple myeloma, and non-Hodgkin's lymphomas. Patients often succumb to septic complications. The disease requires treatment with cytostatic agents.

From a morphological perspective, two main tumor types are distinguished: the hyaline-vascular type (which accounts for approximately 70% of cases) and the plasma cell type. A mixed form, incorporating features of both types, is also identified. Furthermore, the possibility of transition from one form to another cannot be excluded. A fourth type, the plasmablastic variant, is classified separately and is considered clinically the most unfavorable [1–3, 5].

The management of generalized, aggressively progressing forms involves polychemotherapy, glucocorticoid therapy, antiviral therapy, immunotherapy, and monoclonal antibodies targeting CD4 (cluster of differentiation 4). For large localized tumors, radiation therapy is additionally used prior to surgical intervention [4, 6].

Due to the low prevalence of the disease, comprehensive statistical data are lacking. Standardized treatment protocols are also unavailable.

Given its non-specific clinical presentation, Castleman's disease must be differentiated from reactive, neoplastic, and IgG4-associated lymphadenopathies [2].

Diagnosis should be based on physical examination, computed tomography (CT) of the neck and chest, ultrasound of the soft tissues of the neck and abdomen, and laboratory studies. Final diagnosis relies exclusively on histopathological examination of the tumor. Fine-needle aspiration biopsy is not considered informative for diagnosing Castleman's disease [1, 2, 5].

CASE REPORT

A five-year-old girl presented in September 2024 for an outpatient consultation with a pediatrician and an oncologist at her local clinic due to complaints of a mass on the right lateral neck. The mother had noticed the mass approximately two years earlier while the child was in good health; it had been gradually increasing in size. On examination, a diagnosis of cervical lymphadenopathy was made, and follow-up observation was recommended. Due to the subsequent development of pain in the area of the mass, the mother sought evaluation at the Pediatric University Consultative and Diagnostic Center to consider surgical intervention and obtain histological verification of the lesion.

The patient was born to a primigravida mother after an uncomplicated pregnancy and delivery, with age-appropriate growth and developmental milestones. She is fully vaccinated according to the national immunization schedule, is not under any specialist's care for chronic conditions, and contracts acute respiratory viral infections (ARVI) 3–4 times per year.

Physical examination revealed a normosthenic child. Weight and height were within age-appropriate norms. The skin was of normal color and clear. Body temperature was normal. A round, well-circumscribed mass approximately 5 cm in diameter, not fixed to surrounding tissues, was palpated on the right lateral neck, posterior to the sternocleidomastoid muscle. The overlying skin appeared normal. Other cervical and head lymph nodes were not palpable. Endoscopic examination of the ENT organs revealed no visible pathology. A preliminary diagnosis of a right lateral neck mass was made. Further investigations were planned: complete blood count, biochemical blood analysis, virological testing for major herpesvirus groups, ultrasound (US) of the neck soft tissues, computed tomography (CT) of the neck soft tissues and chest, abdominal ultrasound, and throat and nasal swabs for microbiological culture. The child underwent consultations with specialists in infectious diseases, oncology, phthisiology, surgery, hematology, and dentistry. The results of the investigations were as follows: the complete blood count revealed leukocytosis (15×10^9/L) with a normal erythrocyte sedimentation rate (ESR). C-reactive protein and interleukin-6 levels were within normal limits. Serological tests for Epstein-Barr virus, human herpesvirus 8 (*HHV-8*), and HIV were negative. Ultrasound and CT of the neck soft tissues identified a solitary mass in the region of the right posterior cervical lymph nodes, measuring 5.5 cm in diameter, with smooth borders, no contrast enhancement, and no infiltration of adjacent tissues. The remaining test results were unremarkable.

To excise the mass and establish a definitive histological diagnosis, elective surgery was scheduled. Following the consultation, the patient was admitted to the hospital of the Pediatric Medical University. Under general anesthesia, the mass was surgically removed. Intraoperative findings revealed a well-circumscribed mass, 5.5 cm in diameter, which was easily dissected from the surrounding tissue and excised in toto. The adjacent soft tissues appeared normal. The surgical wound was closed in layers. The procedure was completed without any intraoperative complications.

The postoperative period was uncomplicated. The child received a course of antibiotic and symptomatic therapy. The patient was discharged on the seventh postoperative day for continued outpatient care.

Histopathological examination revealed a spherical, bluish-red mass with dark red areas and a soft, elastic consistency. The cut surface appeared yellowish with small, dark red foci. Microscopically, the tissue consisted of a lymph node with altered follicular architecture. Numerous follicles were small and, in areas, closely spaced, containing hypocellular germinal centers. Within both the follicles and interfollicular zones, vessels of varying caliber exhibited wall thickening due to hyaline deposition. Mantle zone small lymphocytes were arranged in regular, concentric layers around pale centers. One or several small blood vessels, predominantly capillaries and small venules, were seen penetrating the center of some follicles, creating a characteristic “lollipop” appearance. The interfollicular zones were hypervascular. Endothelial cells of these vessels were markedly enlarged, featuring large, round to slightly elongated nuclei and scant eosinophilic cytoplasm. The interfollicular stroma contained scattered lymphocytes, macrophages, and plasma cells, some arranged in small clusters. There was prominent congestion of vessels of various calibers, accompanied by numerous small and large focal hemorrhages. The final histopathological diagnosis was Castleman disease, hyaline-vascular type.

Follow-up examinations at 3, 6, and 9 months postoperatively revealed no complaints. A thin scar was noted at the site of the excised lymph node. Control ultrasound of the neck soft tissues and abdomen showed no abnormalities. Complete blood count and biochemical analysis were within normal limits. Annual follow-up with complete blood count and neck ultrasound was recommended.

This case is notable for the patient’s young age (the reported mean age at diagnosis is 40 years) and the presence of symptomatic disease. Typically, Castleman’s disease is either asymptomatic or manifests with non-specific symptoms such as asthenia, weight loss, and low-grade fever. In the present case, however, the patient presented with a distinct pain syndrome. The correct therapeutic approach led to the patient’s full recovery. It is important to emphasize that fine-needle aspiration biopsy lacks diagnostic utility in Castleman’s disease.

This case highlights a diagnostic challenge relevant to otorhinolaryngologists, surgeons, and pediatricians. Patients with Castleman’s disease often undergo prolonged, inconclusive evaluations by various specialists for lymphadenopathy of unknown origin, a common scenario that leads to diagnostic delay and difficulty.

ADDITIONAL INFORMATION

Author contribution. Thereby, all authors made a substantial contribution to the conception of the study, acquisition, analysis, interpretation of data for the work, drafting and revising the article, final approval of the version to be published and agree to be accountable for all aspects of the study.

Competing interests. The authors declare that they have no competing interests.

Funding source. This study was not supported by any external sources of funding.

Consent for publication. Written consent was obtained from legal representatives of the patient for publication of relevant medical information within the manuscript.

ДОПОЛНИТЕЛЬНАЯ ИНФОРМАЦИЯ

Вклад авторов. Все авторы внесли существенный вклад в разработку концепции, проведение исследования и подготовку статьи, прочли и одобрили финальную версию перед публикацией.

Конфликт интересов. Авторы декларируют отсутствие явных и потенциальных конфликтов интересов, связанных с публикацией настоящей статьи.

Источник финансирования. Авторы заявляют об отсутствии внешнего финансирования при проведении исследования.

Информированное согласие на публикацию. Авторы получили письменное согласие законных представителей пациента на публикацию медицинских данных.

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