

DOI: 10.14744/ejma.2022.76486 EJMA 2022;2(1):20–22

## **Case Report**



# Castleman's Disease with a Rare Paraduodenal Localization Treated by Laparoscopy: Case Report and Literature Review

🔟 Emrah Akin,¹ 🕩 A.Tarik Harmantepe,² 🕩 Bilge Elcin,³ 🕩 Ugur Can Dülger,² 🕩 Metin Senol,⁴ 🕩 Fatih Altintoprak¹

#### **Abstract**

Castleman disease is a rare proliferation disorder of the lymphatic system. Castleman Disease occurs less frequently in the mesenteric-paraduodenal localization. Surgical complete resection is recommended for the diagnosis and treatment of symptomatic paraduodenal masses. The number of reported cases of laparoscopic resection of Castleman's Disease located in the paraduodenal region is quite low in the literature. This article aims to present a rare case with characteristic histological and radiological findings treated with a laparoscopic approach.

**Keywords:** Castleman Disease, Laparoscopy, Lymphoproliferative Disorders, Lymph Node.

Cite This Article: Akin E, Harmantepe AT, Elcin B, Dülger UC, Senol M, Altintoprak F. Castleman's Disease with a Rare Paraduodenal Localization Treated by Laparoscopy: Case Report and Literatur Review. EJMA 2022;2(1):20–22.

astleman Disease (CD) is an extremely rare lymphoproliferative disorder.[1] The pathogenesis of this rare lymphoproliferative disorder is hyperplasia of lymph cells. [2] CD characterized by hyperplasia of lymph cells is mostly located in the mediastinum, less frequently in the paraduodenal region, and complete surgical resection is recommended for its treatment.[3] There is no consensus on the ideal choice between minimally invasive or open surgery for complete surgical resection, especially in cases of paraduodenal CD.[4] There are very few studies in the literature reporting that the rare CD is treated with minimally invasive surgery.[5-7] This study is aimed to present the characteristic radiological features of the patient who was operated for a paraduodenal mass and reported as CD in histopathology, the details, and results of the minimally invasive surgical treatment.

### **Case Report**

Physical examination revealed epigastric tenderness in a 56-year-old female patient who was admitted to the hospital with the complaint of abdominal pain. No specific value was found in the patient's laboratory parameters. Abdominopelvic Computed Tomography showed a well-defined mass of approximately 3x3x2 in size, located in the paraduodenal region adjacent the distal end of the pancreas and shown in Figure 1A,1B, and 1C. Elective surgery with the laparoscopic method was planned. Pneumoperitoneum was created with a Veress needle below the umbilicus, A 52 cm/10 mm/30 ° video endoscopy was inserted through a 10 mm trocar into the abdomen, two ports, 10 mm and 5 mm from the right and left the pararectal region on the mid-clavicular line, were placed in the abdomen under direct

Address for correspondence: Emrah Akin, MD. Sakarya Universitesi Tip Fakultesi Genel Cerrahi Anabilim Dali, Sakarya, Turkey Phone: +90 264 888 40 00 E-mail: emrahakin@sakarya.edu.tr





OPEN ACCESS This work is licensed under a Creative Commons Attribution-NonCommercial 4.0 International License.





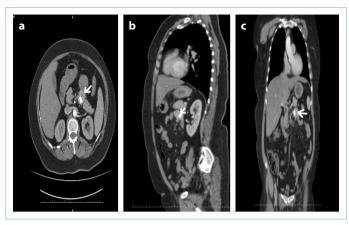
<sup>&</sup>lt;sup>1</sup>Department of General Surgery, Sakarya University Faculty of Medicine, Sakarya, Turkey

<sup>&</sup>lt;sup>2</sup>Department of General Surgery, Sakarya University Educational and Research Hospital, Sakarya, Turkey

<sup>&</sup>lt;sup>3</sup>Department of Pathology, Sakarya University Faculty of Medicine, Sakarya, Turkey

<sup>&</sup>lt;sup>4</sup>Department of General Surgery, Istanbul Technology and Science University, Sakarya, Turkey

EJMA 21



**Figure 1.** Radiological images of the mass, white arrows show the para-duodenal lesion. (a) axial plan (b) sagittal plan (c) coronal plan

vision. In the intra-abdominal exploration, a smooth-circumscribed, soft-consistent mass lesion located in the retroperitoneum, adjacent to the Treitz ligament, and covered partly by the small intestine meso was detected and shown in Figure 2. The mass was separated from the surrounding tissues with sharp dissections by preserving the tail of the pancreas in its cranial neighborhood, the duodenum in its medial and superior neighborhood, and the superior mesenteric artery in the posterior deep line. Artery and vein formed by neovascularization were seen separately in the tumoral mass, was cut with hem-o-lock silicone-coated clips. The mass was resected unblocked and removed from the abdomen. A silicone Jackson-Pratt drain was placed in the lodge and after bleeding control, the trocars were removed, the abdomen was desufflated and the operation was terminated. The patient, whose postoperative period was uneventful, tolerated oral intake on the first day and was discharged on the second day. In the histopathological evaluation, atretic germinal centers are traversed by pen-

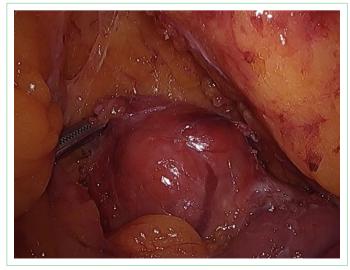
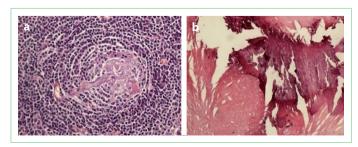


Figure 2. The mass on the para-duodenal region.

etrating hyalinized venules (lollipop follicles) and mantle zone surrounded by small lymphocytes in a concentric pattern (onion skin appearance) was observed and shown in Figure 3A. Areas of heterotopic calcification of the central part of the tumor and hyaline deposits were shown in Figure 3B. The pathological diagnosis was reported as Castleman Disease.

#### **Discussion**

CD was first described as a lymphoproliferative disorder in 1954 by Benjamin Castleman, a pathologist at Massachusetts General Hospital.[8] The characteristic features of CD have been shown to be a benign, localized thymoma-like enlargement involving hyperplastic lymph nodes in the anterior mediastinum. In clinical and radiological definitions, the CD can be classified as unicentric or multicentric depending on the number of lymph nodes involved.[9] Although the etiology is not fully known, chronic low-grade inflammation, immunodeficiency, and dysregulated autoimmunity are thought to be possible mechanisms in the pathogenesis of CD. The critical role of inflammatory mediators such as interleukin 6 (IL-6) or interleukin 10 (IL-10) and human herpesvirus 8 (multicentric variant only) has been demonstrated in preclinical animal models.[10] The unicentric CD is most common in the chest (70%), neck (15%), and rarely in the abdomen-pelvis (12%) and axillary (3%) lymphoid tissues.[11] CD is usually asymptomatic and diagnoses incidentally on the chest or abdominopelvic imaging performed for other ethological reasons.[12] While dyspnea, cough, hemoptysis, and chest pain may be present in thoracic disease, vomiting, dyspepsia, abdominal or lumbar pain may be present in abdominalretroperitoneal disease due to adjacent organ compression. In the abdominopelvic location, the paraduodenal region is so rare and clinically presents with abdominal pain. Diagnosis is made by demonstrating a single, homogeneous, soft, satellite-free intra-abdominal mass on Computed Tomography. However, these features are not unique to CD. Mesenchymal tumors such as GIST, paraganglioma, schwannoma, which are more common in preoperative radiological differential diagnosis, can be considered. The standard treatment



**Figure 3.** Pathological examples **(a)** onion skin appearance **(b)** heterotopic calcification.

for CD is en-block surgical resection, which is a curative approach without recurrence in almost all cases. Controversy still exists between choosing open surgery or minimally invasive surgery for the surgical method. [9] The minimally invasive method can be preferred in appropriate cases in order to benefit from its advantages in experienced centers. The laparoscopic method promises advantages such as less pain, shorter hospital stay, and faster return to work.[13] In our opinion, the laparoscopic method contributed to the visibility and preservation of organ neighborhoods and vascular structures with its x3 enhanced visualization. In cases with uncertain diagnoses, the field of laparoscopic surgery is expanding with both its diagnostic and therapeutic advantages. However, there are studies suggesting that laparoscopy should be used cautiously or not, due to the high risk of rupture and hemorrhage, especially in retroperitoneal masses larger than 6 cm.[14,15] Accordingly, it is important to choose a suitable patient in the selection of the method. In addition, it is a limited method since it requires experience and technical equipment to be applied in rare anatomical localizations. In this case, complete surgical resection was performed with the laparoscopic method. The patient, who had a very uneventful postoperative period, was discharged on the 2<sup>nd</sup> day.

#### **Conclusion**

The diagnosis of Castleman's Disease, which is a rare lymphoproliferative disorder in paraduodenal masses, can be considered and laparoscopic surgery can be considered for its treatment.

#### Disclosures

**Informed Consent:** Written informed consent was obtained from the parents of the patient for the publication of the case report and the accompanying images.

**Peer-review:** Externally peer-reviewed. **Conflict of Interest:** None declared.

**Financial Disclosure:** The authors declared that this study has received no financial support.

**Authorship Contributions:** Concept – E.A., F.A.; Design – F.A.; Supervision – A.T.H., M.S.; Materials – A.T.H.; Data collection and/or processing – U.C.D.; Literature search – N.F., R.C.; Writing – E.A.; Critical review – F.A.

#### References

1. Fajgenbaum DC. Novel insights and therapeutic approaches in idiopathic multicentric Castleman disease. Hematol Am Soc

- Hematol Educ Program 2018;2018:318-25.
- Polizzotto MN, Uldrick TS, Wang V, Aleman K, Wyvill KM, Marshall V, et al. Human and viral interleukin-6 and other cytokines in Kaposi sarcoma herpesvirus-associated multicentric Castleman disease. Blood 2013;122:4189–98.
- Bejjani J, Lemieux B, Gariepy G, Younan R. Complete anemia reversal after surgical excision of mesenteric hyaline-vascular unicentric Castleman disease. Can J Surg J Can Chir 2009;52:197–8.
- 4. van Rhee F, Greenway A, Stone K. Treatment of idiopathic castleman disease. Hematol Oncol Clin North Am 2018;32:89–106.
- Serpa Carmoma F, Cueva M, Serpa Carmoma F, Molina GA, Yu Lee A, Flores E. Castleman's disease: Laparoscopic resection of unicentric disease in the hepatic hilum: A case report. Ann Med Surg (Lond) 2021;67:102494.
- Bracale U, Pacelli F, Milone M, Bracale UM, Sodo M, Merola G, et al. Laparoscopic treatment of abdominal unicentric castleman's disease: a case report and literature review. BMC Surg 2017;17:38.
- 7. Sawaya Z, Semaan DB, Nicolas G, Dib A, Tayar C. Unicentric Castleman's disease: laparoscopic approach of a para-duodenal retroperitoneal mass. Am J Case Rep 2020;21:e918444.
- 8. Castleman B, Towne VW. Case records of the Massachusetts General Hospital: Case No. 40231. N Engl J Med 1954;250:1001–5.
- 9. Yoshizaki K. A reference guide for management of Castleman disease. Rinsho Ketsueki 2017;58:97–107.
- 10. Yoshizaki K, Matsuda T, Nishimoto N, Kuritani T, Taeho L, Aozasa K, et al. Pathogenic significance of interleukin-6 (IL-6/BSF-2) in Castleman's disease. Blood 1989;74:1360–7.
- 11. van Rhee F, Voorhees P, Dispenzieri A, Fosså A, Srkalovic G, Ide M, et al. International, evidence-based consensus treatment guidelines for idiopathic multicentric Castleman disease. Blood 2018;132:2115–24.
- 12. Yu MH, Lee JM, Baek JH, Han JK, Choi B-I. MRI features of gastrointestinal stromal tumors. AJR Am J Roentgenol 2014;203:980–91.
- 13. Guidelines for diagnostic laparoscopy. Society of American Gastrointestinal Endoscopic Surgeons (SAGES). Surg Endosc 1993;7:367–8.
- 14. Lefor AT. The role of laparoscopy in the treatment of intra-abdominal malignancies. Cancer J Sudbury Mass 2000;6:159–68.
- 15. Kriger AG, Berelavichus SV, Son AI, Gorin DS, Akhtanin EA, Kaldarov AR, et al. Surgical treatment of retroperitoneal masses. [Article in Russian]. Khirurgiia (Mosk) 2017;1:15–26.