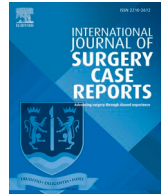




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Case report

Laparoscopic resection of ganglioneuroma from the hepatoduodenal ligament: A case report

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ABSTRACT

Introduction and importance: Ganglioneuromas are extremely rare, slow-growing, benign tumors that arising from Schwann cells, ganglion cells, and neuronal or fibrous tissue. Their malignant degeneration occurs very rarely, complete surgical removal is recommended to eliminate possible symptoms or to prevent possible malignant transformation. Reviewing the literature, there is currently insufficient data available on laparoscopic resection of retroperitoneal ganglioneuromas.

Case presentation: 20-year-old young woman with no previous medical history or regular medication use complaints of abdominal pain. Abdominal CT scan found a cystic mass measuring up to 50 mm in diameter with a thick fluid density and no contrast accumulation, was identified in the porta hepatis region extrahepatically. Ultrasound-guided biopsy was performed, histopathological finding revealed mature benign neurogenic tumor tissue consisting of mature ganglion cells, mature Schwann cells, and branching stroma.

Clinical discussion: A laparoscopic surgery was performed, the 5 cm large tumor was excised from the hepatoduodenal ligament. The tumor was removed from the region of the inferior caval vein, portal vein, and the common and proper hepatic arteries. Final histological diagnosis is ganglioneuroma of the hepatoduodenal ligament. After uneventful postoperative period, the patient was discharged home on the 6th day.

Conclusions: Retroperitoneal tumors were previously excised during laparotomy. However, in recent decades, with the development of laparoscopic surgical techniques and tools, laparoscopic removal of some retroperitoneal tumors seems to be the ideal approach. The use of laparoscopy improves visibility of the relationship of the tumor to the surrounding, often vital, structures. Based on a review of the international literature and our own experience, laparoscopic ganglioneuroma resection is the recommended procedure with careful patient selection, as well as appropriate preoperative imaging and diagnostics, and with adequate expertise.

1. Background

Ganglioneuromas are extremely rare (1/1.000.000), slow-growing, benign tumors that may arise from Schwann cells, ganglion cells, and neuronal or fibrous tissue. It was first described in 1870 by Loretz. It most often occurs in children or young adults, with 60 % of all detections occurring before the age of 20. The majority of ganglioneuromas are diagnosed accidentally, as they are usually asymptomatic and tend to grow slowly. They can remain clinically silent until accidentally discovered or symptoms arise due to mass affect. There is limited data on laparoscopic resection of ganglioneuromas, however laparoscopic excision, after adequate examination is recommended procedure.

2. Case presentation

20-year-old young woman with no previous medical history or regular medication use complaints of abdominal pain and discharge from the navel, attributed to her menstrual cycle. Abdominal ultrasound revealed solid mass in the hilum of her liver measuring a maximum of 46 mm in diameter. This inhomogeneous structure shows no significant vascularization, a finding which is not consistent with an abscess. Its dignity was uncertain, and suggested as aconglomeration of lymph nodes.

Gynecological examination was unremarkable. Abdominal CT scan recommended by the internalist found a cystic mass measuring up to 50 mm in diameter with a thick fluid density (31 HU) and no contrast

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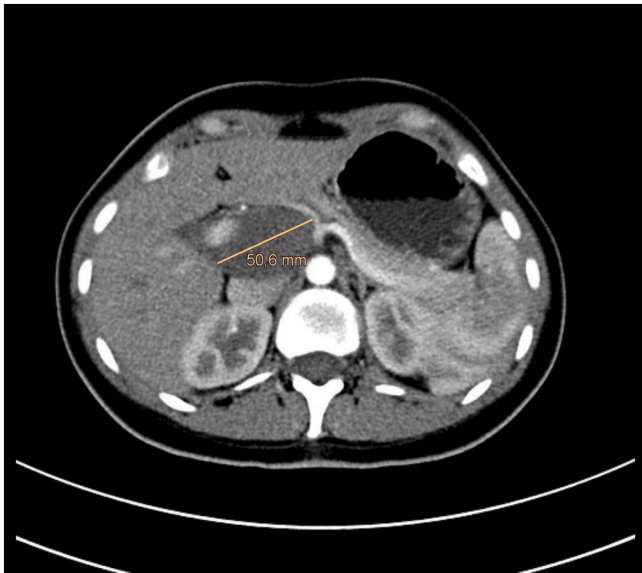


Fig. 1. CT picture of 50 mm large mass in the porta hepatis region.

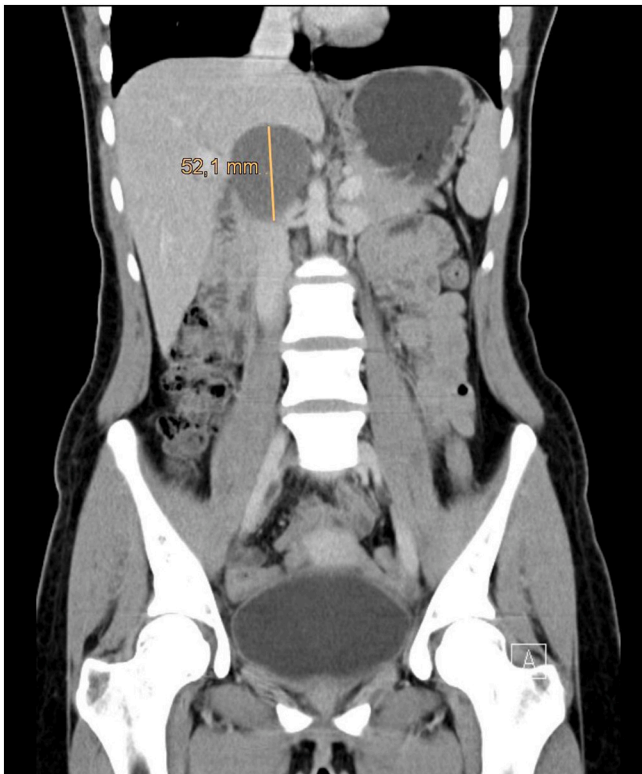


Fig. 2. The mass displacing the surrounding structures.

accumulation, was identified in the porta hepatis region extrahepatically (Fig. 1). The mass displaced the surrounding structures in an arc, but the surroundings were peaceful (Fig. 2). Locoregionally pathological lymph nodes unidentifiable.

Ultrasound-guided biopsy was performed which revealed at the level of the hepatic hilum, the well-defined, cystic pattern noted on CT appeared more solid in ultrasound examination.

Histopathological finding revealed mature neurogenic tumor tissue consisting of mature ganglion cells, mature Schwann cells, and branching stroma. Positivity for S100 and Synaptophysin was observed under immunohistochemical staining. In addition to background positivity expressed with GFAP, mature ganglion cells showed expression. C-kit and DOG1 were negative. There were no signs of malignancy in the tissue. Therefore, the observed lesion may likely correspond to a mature ganglioneuroma.

To complete the staging process a CT scan of the chest was performed which did not reveal any abnormalities indicative of metastasis in the chest.

Oncology Advisory Board recommended surgical intervention.

A laparoscopic surgery was performed under endotracheal anesthesia, during which the 5 cm large tumor was excised from the hepatoduodenal ligament, using 4 ports (Fig. 3). The tumor was removed from the region of the inferior caval vein, portal vein, and the common and proper hepatic arteries using an ultrasonic high-energy device, and was then placed in an endobag. The nerve fiber leading up to the lesion was cut between plastic clips (Fig. 4).

Despite the lesion significantly surrounding the common hepatic artery, the vessel was not damaged anywhere during preparation. The specimen was removed through the 5 cm enlarged wound of the working port of the right hand. The operation time was 140 min, and the total blood loss was 25 ml.

The removed specimen: (Fig. 5).

After uneventful postoperative period, the patient was discharged home on the 6th day.

Histology showed a nodule with a fibrotic capsule containing mature ganglion cells visible within a watery, scarred stroma. Immunohistochemical examination demonstrated stromal and ganglion cells with both Synaptophysin and S-100 positivity. Stromal cells also showed strong SOX10 expression and mild Chromogranin A positivity was found in ganglion cells. The MIB-1 index was less than 1 %. In addition, a tumor-free lymph node was visible. There are no signs of malignancy.

Final histological diagnosis is ganglioneuroma of the hepatoduodenal ligament.

HEx20 (Fig. 6), S100x40 (Fig. 7), SOX10x40 (Fig. 8).

Our patient currently does not have any complaints and does not require further oncological treatment during the postoperative follow up.

3. Discussion

Ganglioneuromas are extremely rare, slow-growing, benign tumors that arising from Schwann cells, ganglion cells, and neuronal or fibrous tissue.

Tumors are most often diagnosed in patients between the ages of 10–29. They can develop along the sympathetic nerve chain, but are

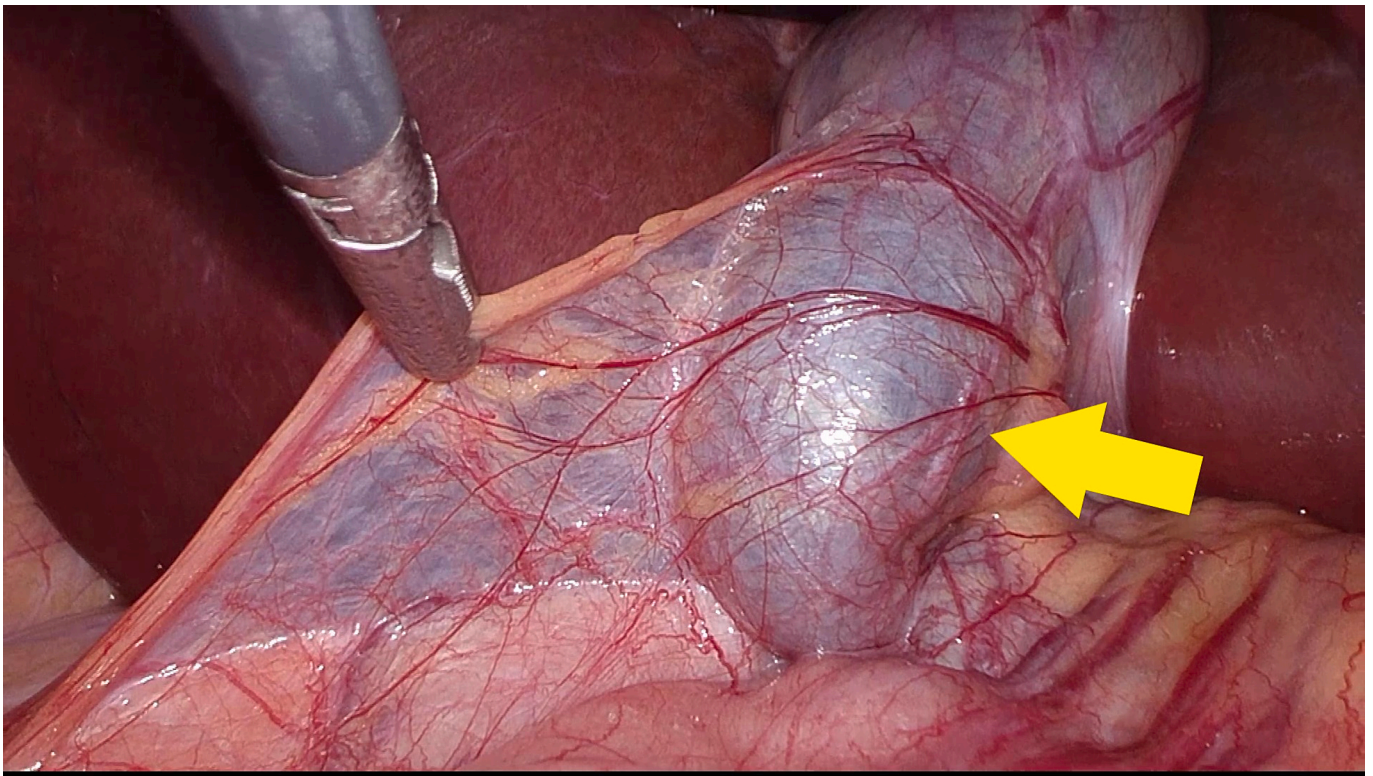


Fig. 3. Intraoperative picture of the tumor indexed in the hepatoduodenal ligament.

predominantly found in the posterior mediastinum (42 %), retroperitoneal region (37.5 %), in the adrenal glands and in the cervically retropharyngeal regions (2–5 %) [1]. The incidence rate is higher in women than in men, by a ratio of approximately 3:2. They are usually discovered incidentally because they are usually asymptomatic and grow slowly.

In terms of imaging, computed tomography (CT) and magnetic resonance imaging (MRI) are the gold standard procedures for establishing a diagnosis and assessing the extent of the tumor. In the absence of specific radiologic features, biopsies (ultrasound-guided fine-needle aspiration or CT-guided core biopsy) can provide a histologic diagnosis that can aid in planning treatment and surgery [2].

Ganglioneuromas show neuroendocrine potential, which can be attributed to their origin in the sympathetic nervous system, but the majority of them are hormonally inactive. Microscopically, the absence of mitotic figures, intermediate cells, neuroblasts, or necrosis distinguishes ganglioneuroma from ganglioneuroblastoma and paraganglioma.

Their malignant degeneration occurs very rarely, so there is no need for neoadjuvant or adjuvant oncological treatment [3].

Based on these, complete surgical removal is recommended to eliminate possible symptoms or to prevent possible malignant transformation. In addition, subsequent long-term follow-up (including imaging controls) is mandatory in order to prevent possible recurrence, especially if only partial tumor removal was achieved [4].

After complete tumor resection, the prognosis is excellent, although surgical morbidity must be taken into account, especially in the case of a large ganglioneuroma.

Reviewing the literature, there is currently insufficient data available on laparoscopic resection of retroperitoneal ganglioneuromas. Retroperitoneal tumors were previously excised during laparotomy. However, in recent decades, with the development of laparoscopic surgical techniques and tools, laparoscopic removal of some retroperitoneal tumors seems to be the ideal approach. Between 2008 and 2021, laparoscopic resections of 12 retroperitoneal ganglioneuromas were published [5]. Average age of the patients is 33 years (22–60 years), average tumor size is 50 mm (40–90 mm), average operation time is 170.5 min (80–350 min), average blood loss is 21.5 ml (0–250 ml), average hospital stay is 7 days (3–12 days).

4. Conclusions

The use of laparoscopy improves visibility of the relationship of the retroperitoneal tumor to the surrounding, often vital, structures, and helps to perform a more precise dissection [6]. The use of laparoscopic ultrasonic high-energy cutting devices and various retractors allow manipulation of the tumor mass and safe dissection in tight spaces. Furthermore, laparoscopic surgery is associated with fewer postoperative complications, including less blood loss, fewer postoperative adhesions, less postoperative pain and faster recovery and shorter

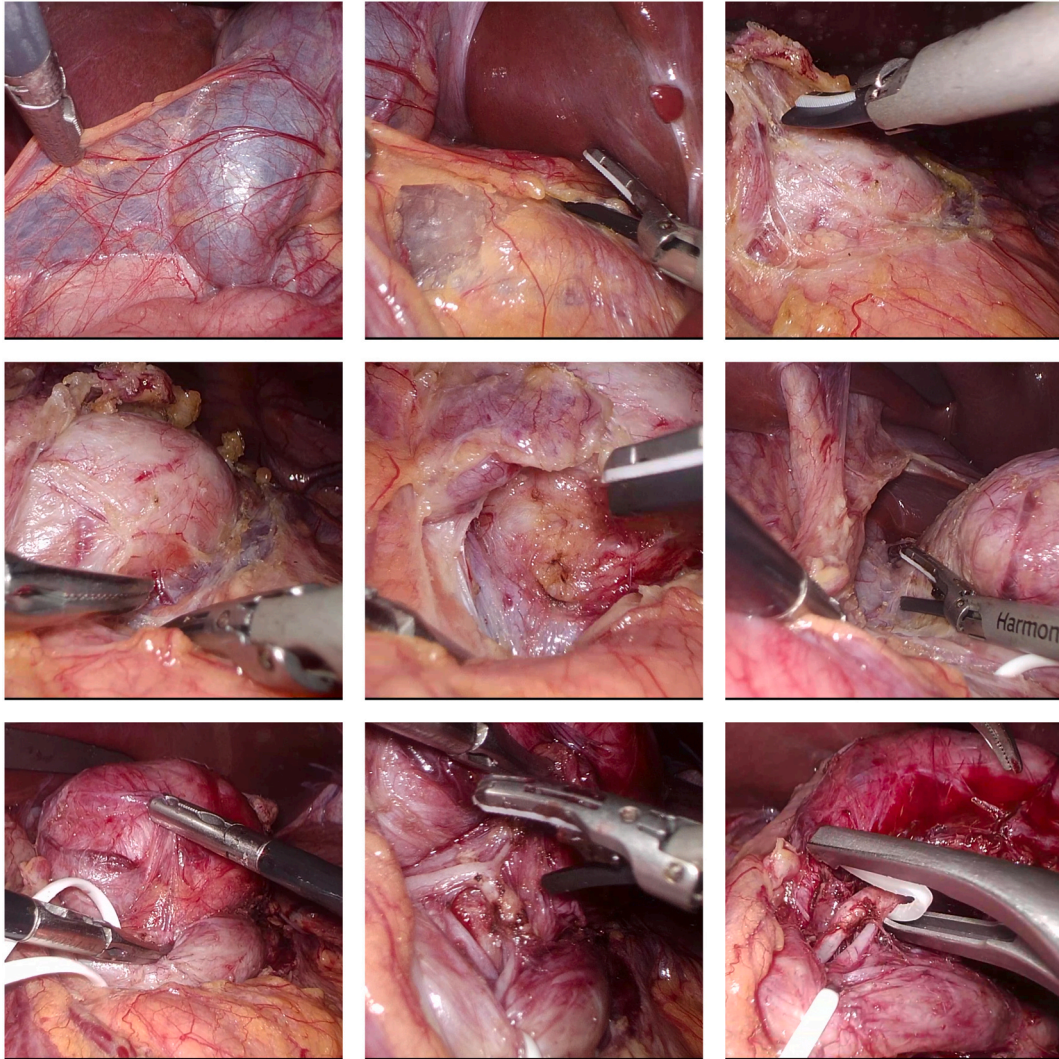


Fig. 4. Intraoperative pictures of tumor removal.



Fig. 5. The tumor after removal.

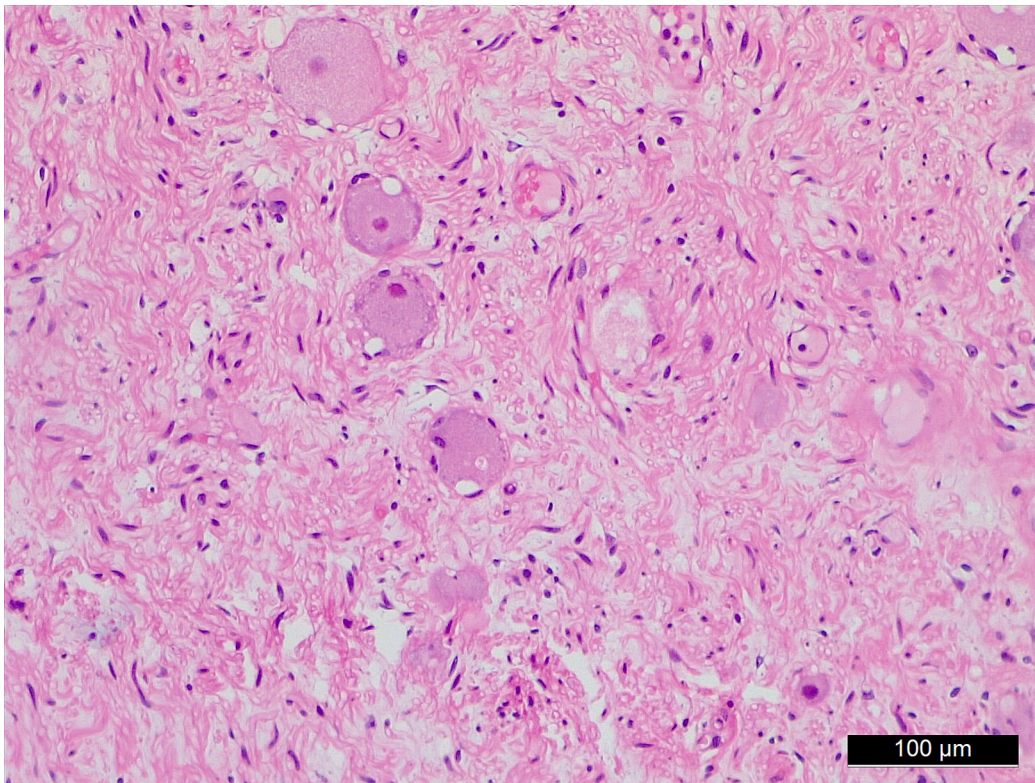


Fig. 6. Histological picture HEx20.

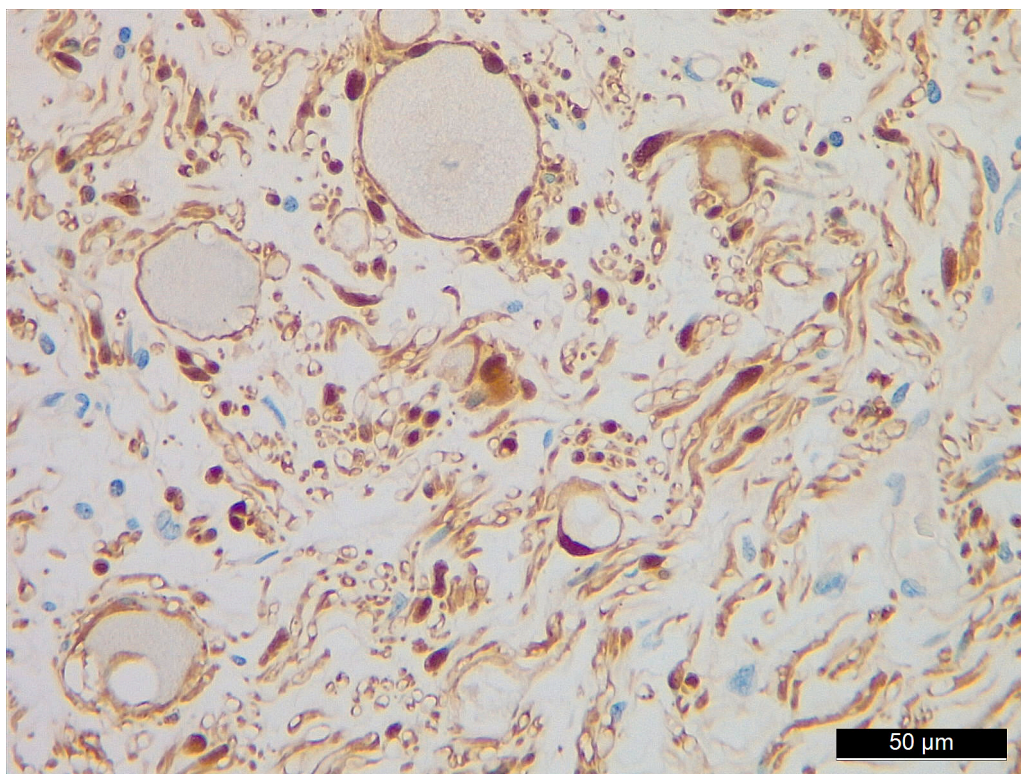


Fig. 7. Histological picture S100x40.

hospital stay compared to an open surgery [7].

Based on a review of the international literature and our own experience, laparoscopic ganglioneuroma resection is the recommended

procedure with careful patient selection, as well as appropriate preoperative imaging and diagnostics, and with adequate expertise. Careful multidisciplinary team work needed to establish proper preoperative

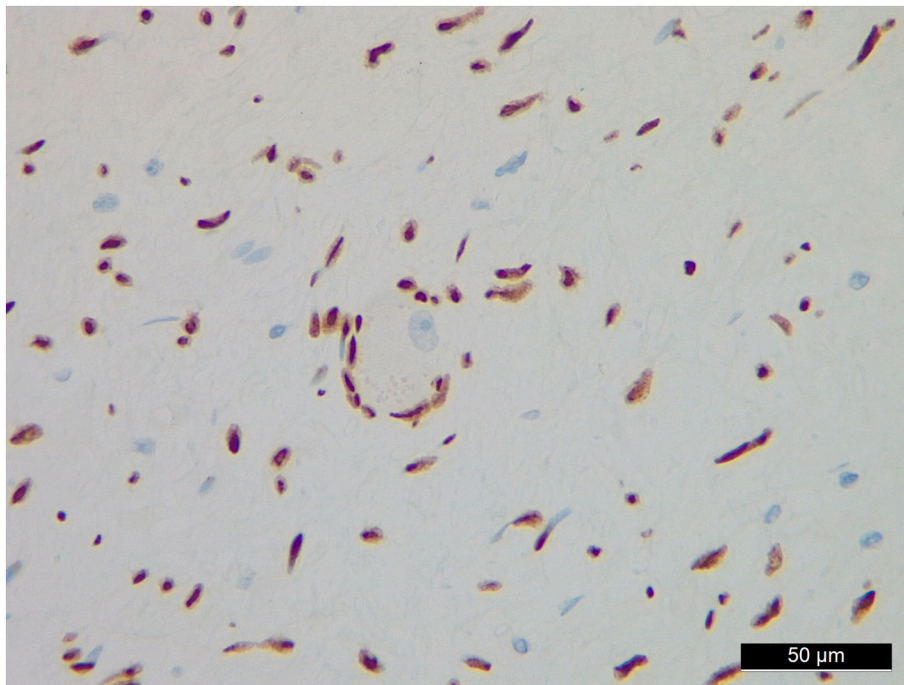


Fig. 8. Histological picture SOX10x40.

diagnosis and patient evaluation and management to achieve successful therapy beside comprehensive postoperative follow up.

List of abbreviations

HU Hounsfield unit

Declarations

The work has been reported in line with the SCARE criteria.

Ethical approval

Ethics committee approval was not given, but the patient consented to the publication of the study, as the study does not contain data on the individual identification or ethnic identification of the patient.

Funding

The study sponsor (University of Debrecen, Hungary) has no involvement in data collection and analysis, and in the writing of the manuscript.

Author contribution

Tamás Felföldi contribute to the publication.
Zsolt Varga contribute to the publication.
Péter Kolozsi contribute to the publication.
Dávid Ágoston Kovács contribute to the publication.
Dezso Tóth contribute to the publication.

Guarantor

The Guarantor is the corresponding author, Tamás Felföldi.

Research registration number

Not applicable.

Conflict of interest statement

The case report does not hurt any of financial or personal interests.

Data availability

All the datasets used in this report about the patient can be found in the digital register of the Surgical Department of the University of Debrecen.

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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