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A Successful Giant Esophageal Gastrointestinal Stromal Tumor Enucleation with VATS : A Case Report

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Case Report

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Abstract

Background

Esophageal gastrointestinal stromal tumor (GIST) is exceedingly rare, accounting less than 1% of all GISTs with no distinctive clinical guidelines. Masses exceeding 5 cm have traditionally been managed by esophagectomy due to concerns about mucosal integrity and oncological safety.

Case presentation :

A 43-year-old woman came to our clinic with incidentally discovered giant esophageal mass, suspected to be leiomyoma, later than diagnosed with 9.4cm sized esophageal GIST. Three-port video assist thoracoscopic surgery (VATS) enucleation was performed without preoperative biopsy. Mucosal integrity and azygos vein were well preserved, and there was no evidence of tumor rupture on histologic findings. Post operative esophagography showed no leakage. The patient was discharged on third postoperative day without any complications. CD117 positivity confirmed GIST diagnosis and the patient had no evidence of recurrence on seventh week follow up and adjuvant imantinib was not required.

Conclusions

Current case challenges the conventional 5cm threshold of mass size for esophagectomy and demonstrates VATS enucleation is feasible for giant esophageal GISTs exceeding 9 cm. It also highlights CD117's essential role in distinguishing GIST from leiomyoma.

Background

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal sarcomas of the gastrointestinal tract, with an estimated incidence of 10–15 per million per year [1]. Esophageal GISTs are even rarer, accounting for approximately 0.7% of all GISTs [2]. Given this rarity, no specific surgical guidelines currently exist; however, both the National Comprehensive Cancer Network (NCCN) and European Society for Medical Oncology (ESMO) recommend complete tumor resection without rupture [1, 3].

Generally, enucleation has been performed for esophageal mesenchymal tumors smaller than 65mm, while larger masses have been managed by esophagectomy owing to concerns regarding mucosal integrity and tumor rupture [4]. Although esophagectomy offers complete resection, it carries significant perioperative morbidity and leads to reduced long-term quality of life. Recently, accumulating case reports suggested that minimally invasive surgical enucleation may remain a feasible and organ-preserving option for larger lesions[5–8].

Herein, we report a case of a giant, 9.4 cm esophageal GIST successfully treated by VATS enucleation while preserving the azygos vein and esophageal integrity.

Case Report

A 43-year-old woman was referred to our clinic due to an incidentally discovered huge esophageal mass. Esophagogastroduodenoscopy and endoscopic ultrasound (EUS) revealed a homogenous and hypoechoic mass arising from the muscularis propria layer at 23 from the incisor teeth (Fig. 1A). Chest computed tomography (CT) revealed a huge, about 6 cm-sized esophageal submucosal mass in the upper esophagus, and it was suspected to be a submucosal leiomyoma (Fig. 1B). Video-assisted thoracoscopic surgery (VATS) mass enucleation with three-ports access was planned without preoperative biopsy.

The patient was placed in a left lateral decubitus position under general anesthesia with one-lung ventilation. Two 5-mm ports and one 12-mm port were placed. A large, protruding, inverted L-shaped and well capsulated mass surrounded by the azygos vein was revealed (Fig. 2). With careful dissection of the pleura and the outer longitudinal muscular layer of the mid-esophagus using harmonic scalpel (Harmonic Ace; Ethicon Endo-Surgery, Cincinnati, OH, USA), the encapsulated mass was enucleated while preserving the azygos vein and the integrity of the esophagus (Fig. 2B). The continuous closure of the outer muscular layer of esophagus with a 3 – 0 Vicryl V-Loc suture was then performed (Fig. 2C). A 20 French chest tube was inserted via a 5-mm port site before closing the wound. The surgery took 143 minutes, then the patient was sent to the recovery room.

On postoperative day 0, the total drain was 11 cc, and 42 cc was drained the next day and removed after the esophagogram. Soft diet was start after the esophagogram revealed no leakage or stenosis (Fig. 3A). The patient was discharged on postoperative day 3 without any complications. There was no change in laboratory tests until the day of discharge. Pathologically, the tumor was 9.4x5.8cm sized and showed positive immunoreactivity for C-KIT, smooth muscle actin, and desmin (Fig. 4). The mitotic rate was 0/50 High-power fields (HPF). Therefore, the patient was diagnosed with an esophageal GIST of pathologic stage T3. She was then referred to an oncologist. According to the pathologic findings, adjuvant imatinib was not needed. At the 7-weeks follow-up, the patient was doing well with no dysphagia and no evidence of recurrence on chest CT (Fig. 5).

Discussion

Esophageal GISTs are exceedingly rare, accounting for less than 1% of all GISTs, and there is no definite diagnostic and surgical guidelines [2, 9]. This case is notable on two counts: the successful VATS enucleation of a large 9.4 × 5.8 cm esophageal GIST, and an immunohistochemical profile that illustrates the critical role of CD117 in confirming the diagnosis.

The preoperative differentiation of esophageal GIST from leiomyoma is inherently difficult, as both arise from the muscularis propria and share overlapping imaging characteristics on CT and EUS. In this case,

the lesion was presumed to be a leiomyoma preoperatively and biopsy was deferred to avoid mucosal seeding and adhesion that may compromise subsequent surgical dissection. Postoperative immunohistochemistry demonstrated positive immunoreactivity for C-KIT (CD117), SMA, and desmin, and negative reactivity for CD34. Esophageal GIST express positivity for CD117 (nearly 100%), and up to 80% positivity for CD34, while esophageal leiomyoma express negativity for CD117 and CD 34[10, 11]. Diffuse CD117 positivity therefore remains the decisive discriminating marker for GIST, irrespective of concurrent smooth muscle marker expression.

Historically, tumors exceeding 65mm were considered to require esophagectomy, given concerns regarding mucosal integrity and seeding risk during enucleation [4]. Present case challenges this conventional threshold. Using a three-port right-sided VATS approach, the 9.4 cm encapsulated tumor was enucleated in 143 minutes, with preservation of both the azygos vein and esophageal mucosal integrity—representing one of the largest esophageal GISTs reported to have been managed by VATS enucleation. In a series of five patients with low- to intermediate-risk esophageal GISTs managed by VATS enucleation with a median follow-up of 5.5 years reported no recurrences, supporting the oncological adequacy of this organ-preserving approach [12]. Postoperative esophagography confirmed the absence of leakage, and the patient was discharged on postoperative day 3 without complications, affirming both the safety and efficiency of minimally invasive enucleation.

Regarding adjuvant therapy, current NCCN guideline reserve adjuvant imatinib for high-risk patients defined by tumor size over 10 cm, 5 > 10/50 HPF, combined size over 5 cm with mitotic count over 5/50 HPF [3]. With a mitotic rate of 0/50 HPF, complete R0 resection without rupture, and a tumor diameter of 9.4 cm—below the 10 cm high-risk threshold—our patient did not fulfill high-risk criteria, and adjuvant imatinib was withheld accordingly. No evidence of recurrence was identified at 7-week follow-up.

Conclusion

In conclusion, this case demonstrates that VATS enucleation is feasible and oncologically appropriate for giant esophageal GISTs exceeding 9 cm, and that CD117 immunoreactivity remains the cornerstone of diagnosis when distinguishing esophageal GIST from leiomyoma.

Abbreviations

GIST
gastrointestinal stromal tumor
CT
Computed Tomography
VATS
Video assist thoracoscopic surgery
HPF
High Power fields

EUS
Endoscopic Ultrasound

Declarations

Ethics approval and consent to participate :

Not applicable.

Consent to participate :

Not applicable.

Consent for publication :

Appropriate written informed consent was obtained for the publication of this case report and its accompanying images.

Competing interests :

Nothing to declare.

Conflicts of Interests :

None declared.

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Author Contribution

JSY and JHK wrote the main manuscript text. SJK, TYK, KHK prepared figures 1-3, 5 and edited manuscript. KYJ prepared figure 4. JSY and JHK conducted the operation and postoperative care of the patient. All authors read and approved the final version of the manuscript.

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Availability of data and materials :

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Figures

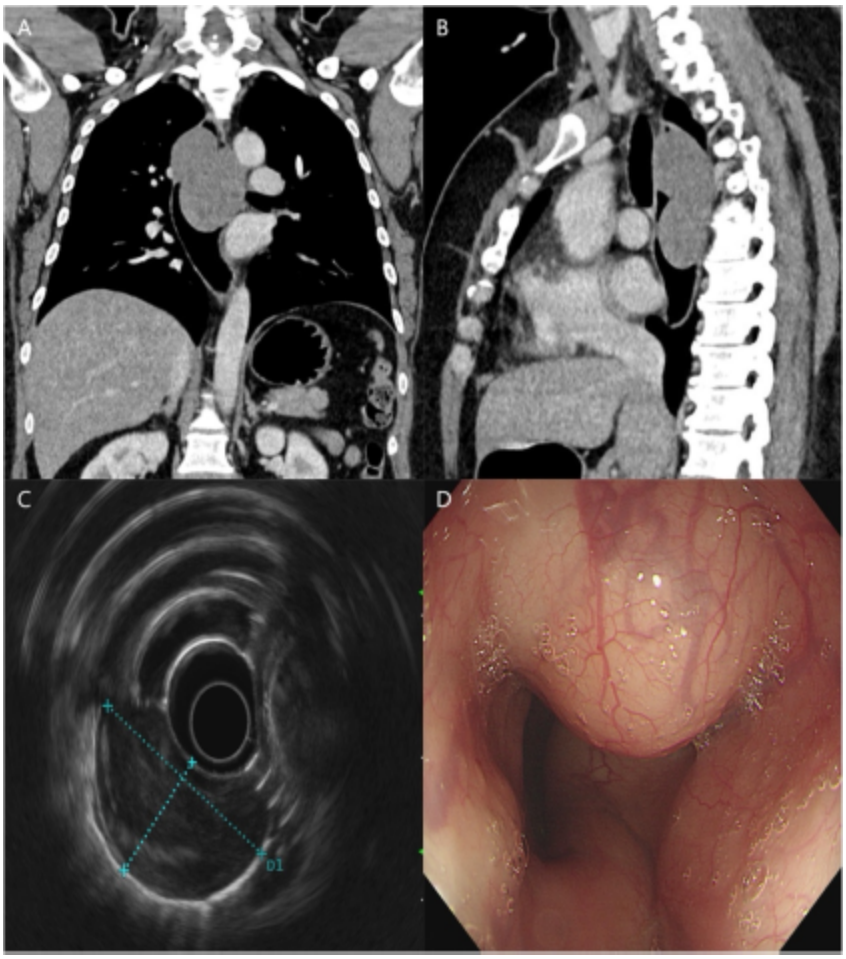


Figure 1

Computed tomographic images of Esophageal GIST and endoscopic images of Esophageal GIST

A : Coronal view of esophageal GIST on enhanced chest CT.

B : Sagittal view of esophageal GISTon enhanced chest CT.

C : EUS showing homogeneous and hypoechoic mass arising from muscularis propria.

D : Endoscopic view showing giant protruding mass of esophagus.

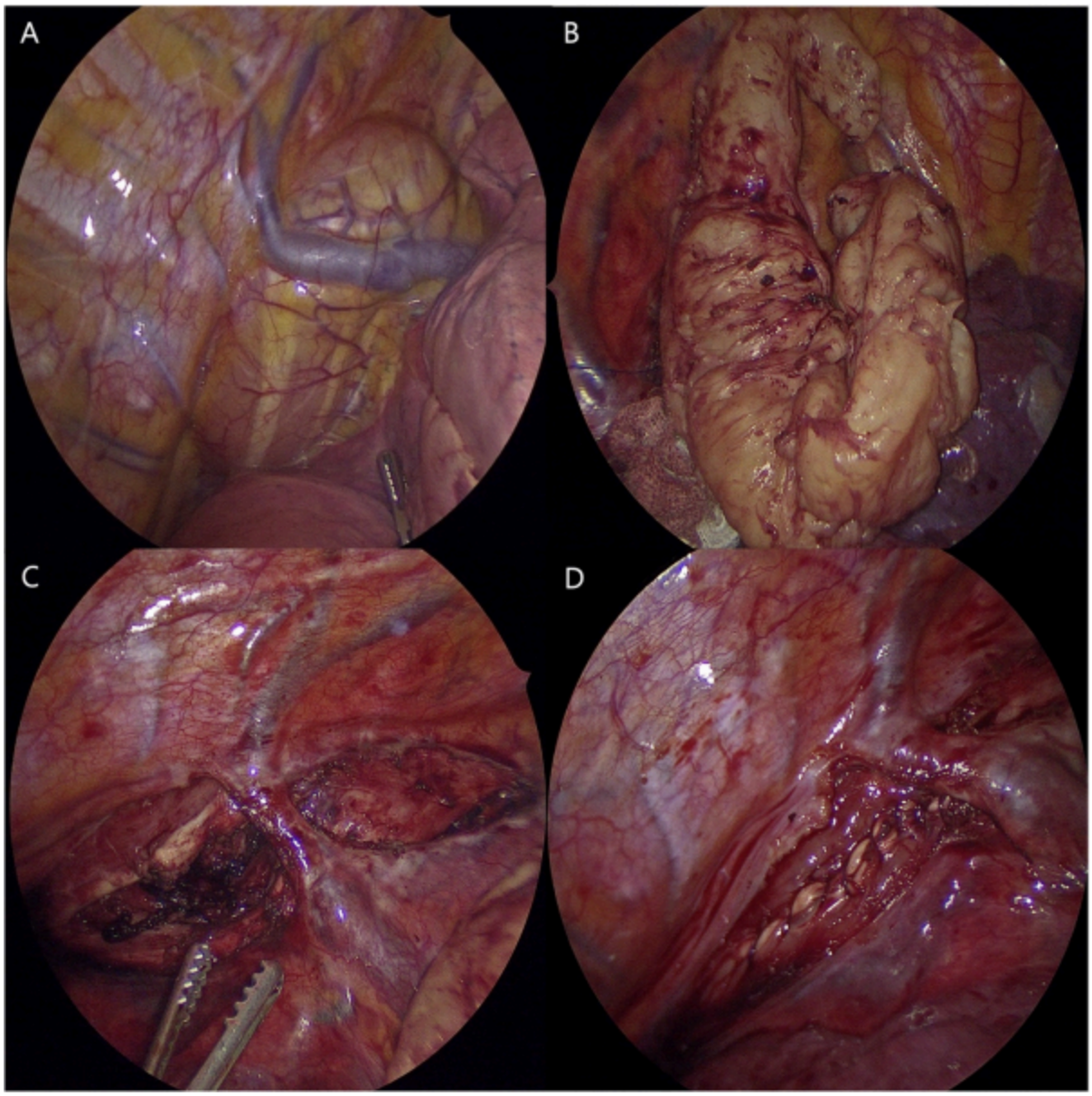


Figure 2

VATS findings of esophageal GIST

A : Esophageal GIST surrounded by azygos vein.

B : Resected esophageal GIST before extraction.

C : VATS view showing well preservedazygos vein and mucosal layer of esophagus.

D : After continuous closer of outer layer of esophagus.

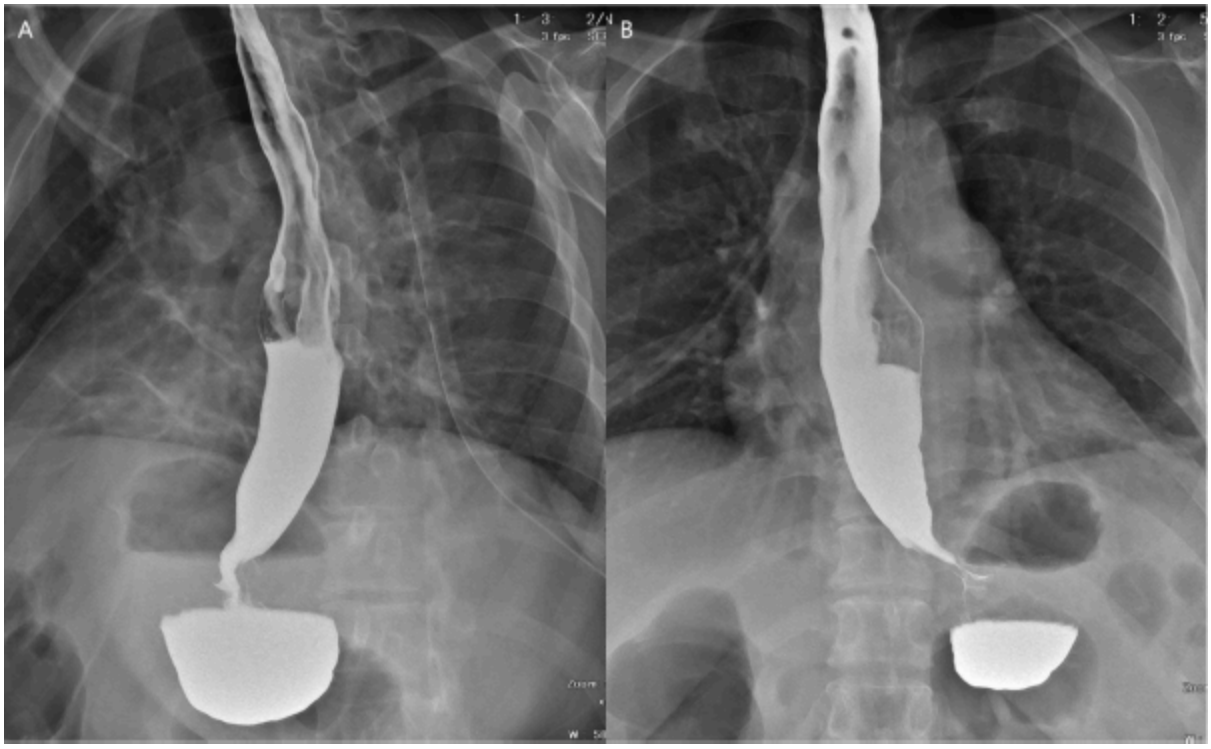


Figure 3

Esophagogram on postoperative day 1

A : Lateral view of esophagogram showing no leakage after surgery on postoperative day 1.

B : PA view of esophagogram showing no leakage after surgery on postoperative day 1.

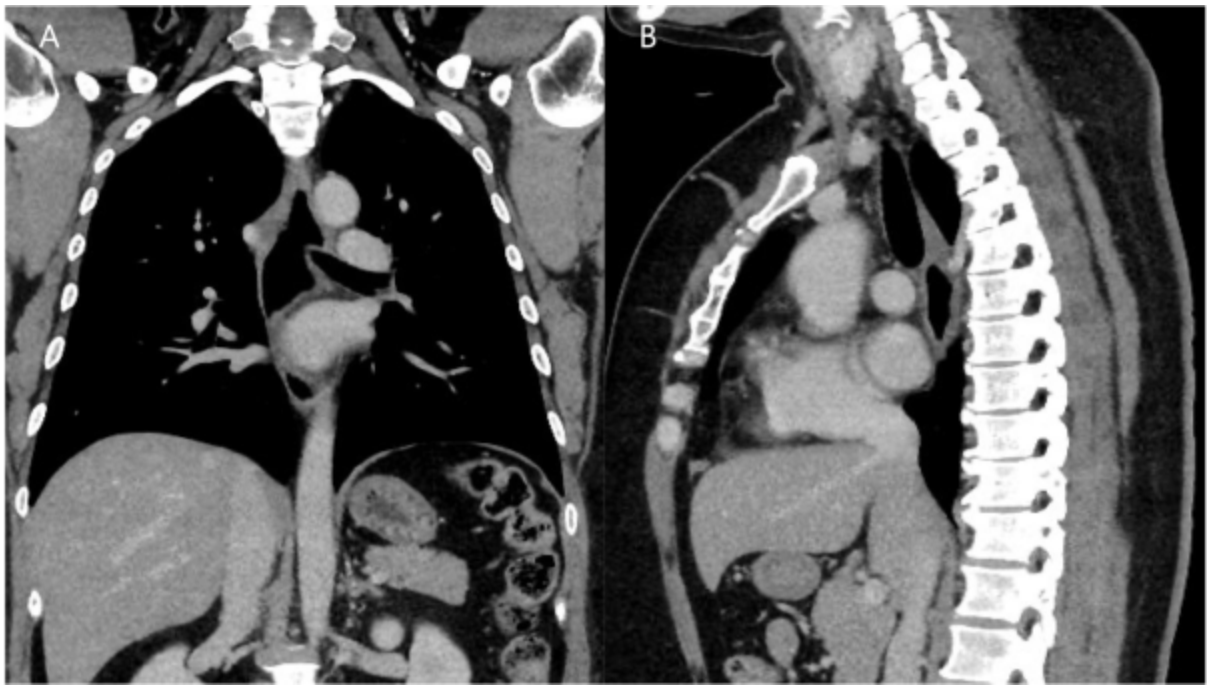


Figure 4

Follow up CT of patient seven weeks after surgery

A : Coronal view of non-enhanced chest CT.

B : Sagittal view of non-enhanced xhest CT.

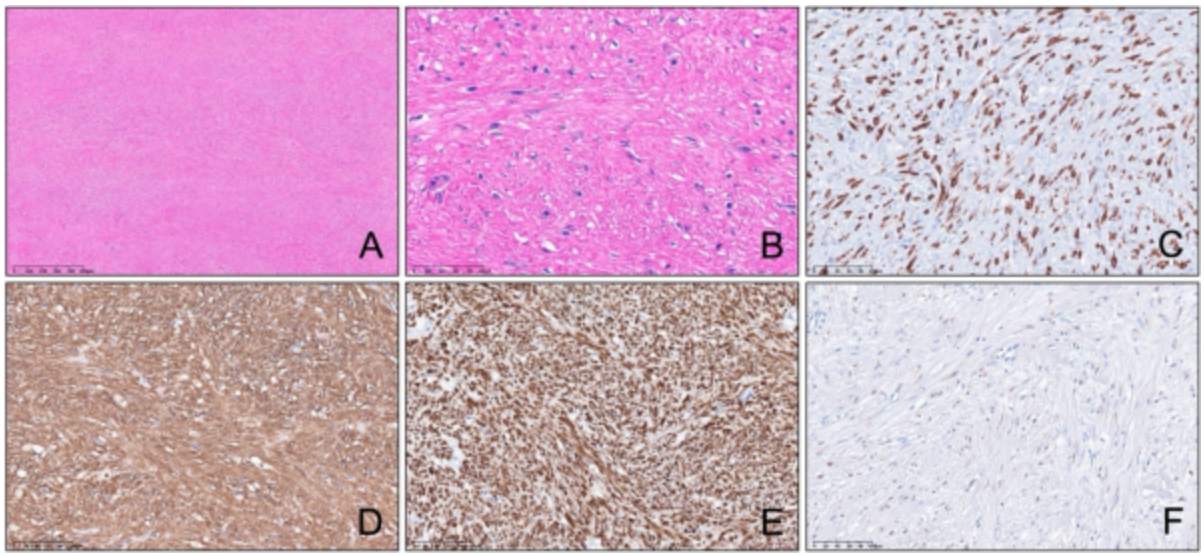


Figure 5

Histologic findings of gastrointestinal stromal tumor.

A and B : The tumor is homogeneous and paucicellular, composed of spindle to oval cells without significant cytologic atypia

C : Tumor cells are positive for C-KIT.

D : Tumor cells are positive for smooth muscle actin.

E : Tumor cells are positive for desmin.

F : Tumor cells are very weakly positive for S100.

Original magnification: A) ×40, B) ×400, C-F) ×200.