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Case Report

Extragastrintestinal stromal tumor presenting as an exophytic prostatic mass [☆]

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ABSTRACT

Extragastrintestinal stromal tumors (EGISTs) are uncommon manifestation of gastrointestinal stromal tumors originating from cells outside the gastrointestinal tract. Documented sites of EGISTs include the omentum, mesentery, retroperitoneum, and prostate gland. Prostatic EGISTs are rare entities, which have been sporadically observed, yet all of them were found to be confined within enlarged prostates. We herein report a rare case of EGIST in a 66-year-old man, presenting as a large exophytic prostatic mass.

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Introduction

Gastrointestinal stromal tumors (GISTs) are an uncommon type of tumor of the digestive tract. Estimated annual incidence of GIST is approximately 11–15 per million [1,2]. GISTs typically arise from GI wall, with the stomach, small bowel, and large bowel being the most common sites [3]. However,

around 5% of GISTs originate from cells outside the GI tract, which are termed extragastrintestinal GIST (EGIST) [4]. Common sites of EGISTs are omentum, mesentery, and retroperitoneum [5–7]. EGISTs arising primarily within the prostate gland are rare [8–14]. In all of the reported cases of prostatic EGISTs, the tumors were located within enlarged prostate glands. We herein report a rare case of prostatic EGIST that manifests as an exophytic prostatic mass.

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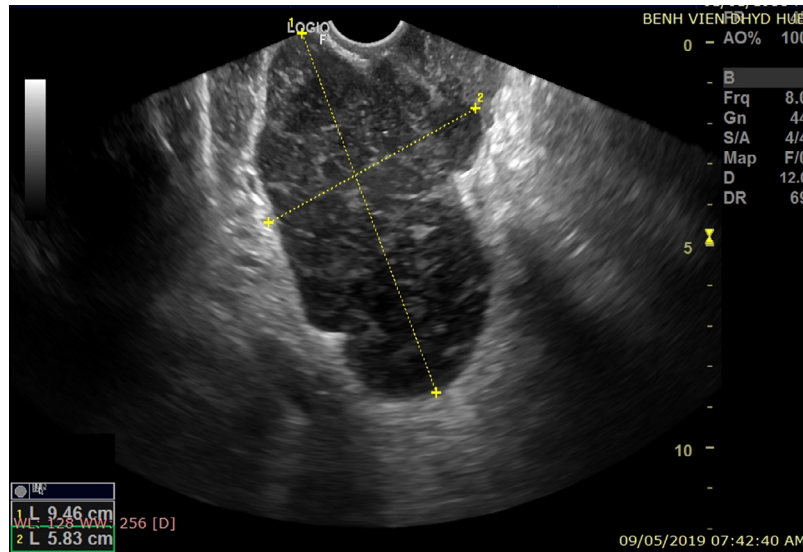


Fig. 1 – Endorectal ultrasound shows a heterogeneous soft tissue mass behind the bladder with lobulated contour.

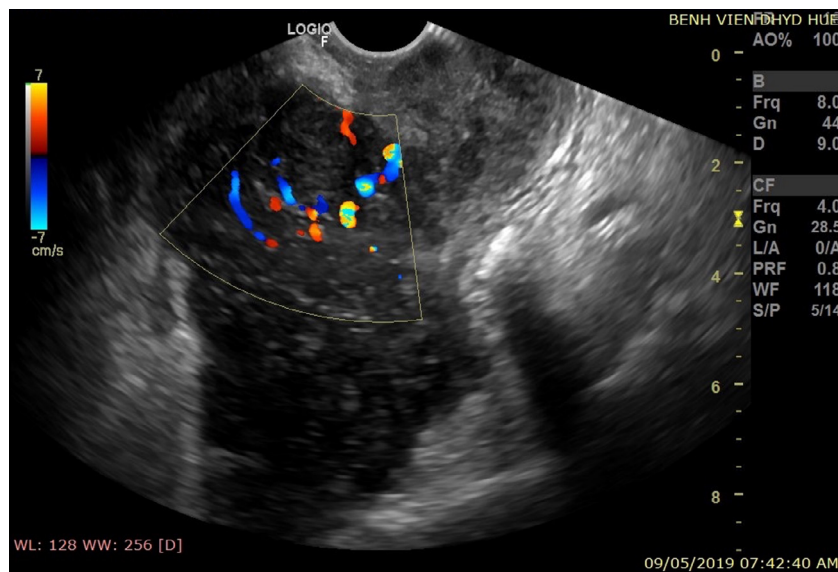


Fig. 2 – Doppler image shows increased vascularization at the periphery of the mass.

Case report

A 66-year-old man presented to the Department of Urology with the chief complaint of mild dysuria for 2 years. A digital rectal examination revealed a large mass with smooth, bulging contour, presumably an enlarged prostatic mass. The serum prostate-specific antigen level was 1.93 ng/mL.

The patient underwent endorectal ultrasound, which revealed a $9.5 \times 6.3 \times 5.8$ cm lobulated soft tissue mass located posterior to the urinary bladder (Fig. 1). The mass was heterogeneous with areas of cystic change. Doppler images showed increased vascularization at the periphery of the mass (Fig. 2). Pelvic magnetic resonance imaging demonstrated a giant, well-defined mass posterior to the bladder, which is con-

tiguous with the prostate. The mass was heterogeneously hyperintense on T2W images and hypointense on T1W images (Figs. 3 and 4). The solid components of the mass showed high-signal intensity on diffusion-weighted images and low apparent diffusion coefficient (ADC) values, consistent with restricted diffusion (Figs. 5 and 6). The tumor showed robust but heterogeneous enhancement after intravenous administration of gadolinium with cystic changes at peripheral area (Figs. 7 and 8). The “claw sign” was evident, suggesting a prostatic origin, whereas the prostate itself was small and intact (arrow, Fig. 8). MR spectroscopy demonstrated an elevated citrate peak at the peripheral solid zone (Fig. 9).

The patient underwent an ultrasound-guided transrectal prostatic needle biopsy, which revealed spindle cells prolifera-



Fig. 3 – Sagittal T2W image shows a large well-defined mass with heterogeneous hyperintensity.

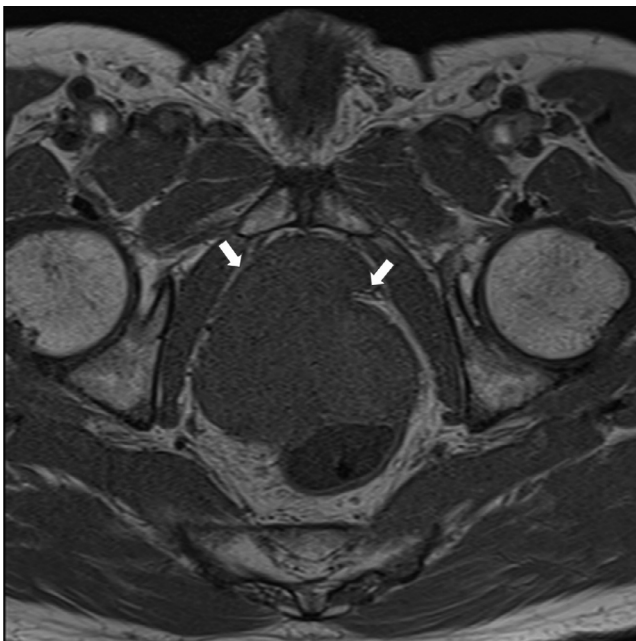


Fig. 4 – The mass was in continuity with the prostate (arrows) and was isointensive on axial T1W image.

tion in continuity with normal prostate tissue (Figs. 10 and 11). At surgical observation, the mass extended cranially and was separate from the rectum. Postoperative pathological study revealed a stromal tumor with moderate risk (9.5 cm, 3/50 HPF) according to 2006 Miettinen and Lasota classification [15] (Fig. 12), which was subsequently confirmed by immunohistochemistry. Immunohistochemistry results were as follows: CD117 (+), CD34 mild (+), Desmin (–), Vimentin (+), S100 mild (+), Ki67 sporadic (+) (Figs. 13–18).

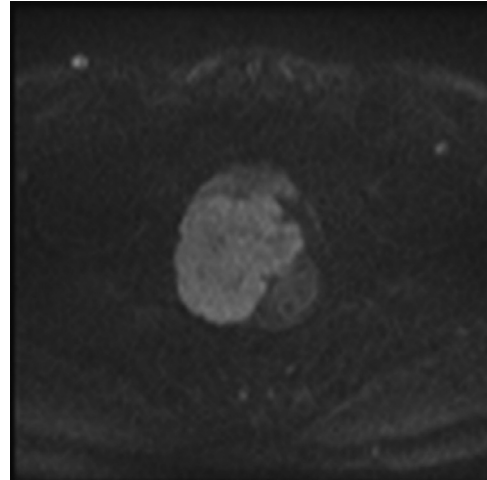


Fig. 5 – The mass had high signal intensity on diffusion-weighted images.

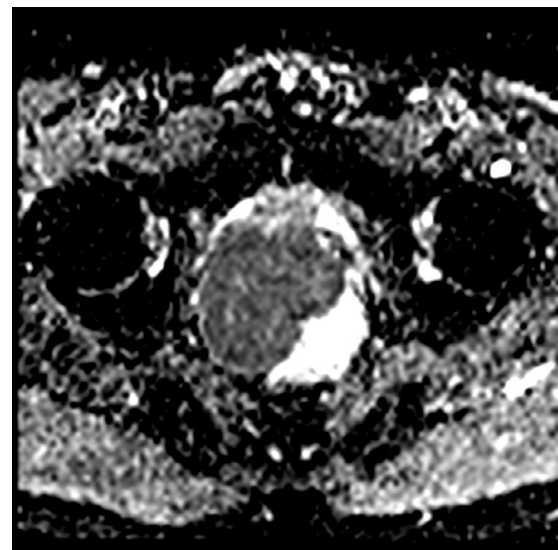


Fig. 6 – The mass showed low ADC values consistent with diffusion restriction.

Discussion and conclusion

GI stromal tumors are a heterogeneous group of mesenchymal tumors of the GI tract [5]. They arise from the interstitial cells of Cajal [7]. GISTs are typically found in patients over 40. Around 70% of these tumors are symptomatic [2]. The common symptoms are GI bleeding and gastric discomfort [5]. Bowel obstruction is uncommon. Radiologic appearance varies according to tumor size and location. Tumors usually appear as a well-defined mass originating from GI wall. Endoluminal extension or exophytic growth are not uncommon. On computed tomography, tumors usually have heterogeneous soft tissue density. Cystic degeneration or necrosis are com-

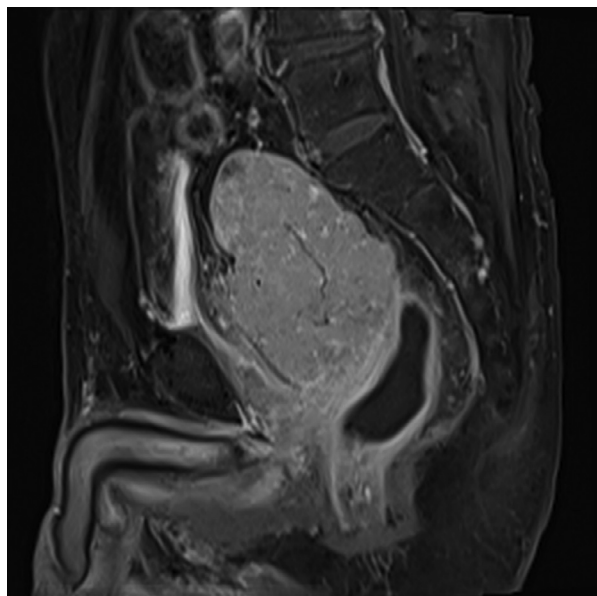


Fig. 7 – Sagittal T1W after Gadolinium intravenous administration. The tumor showed robust enhancement.

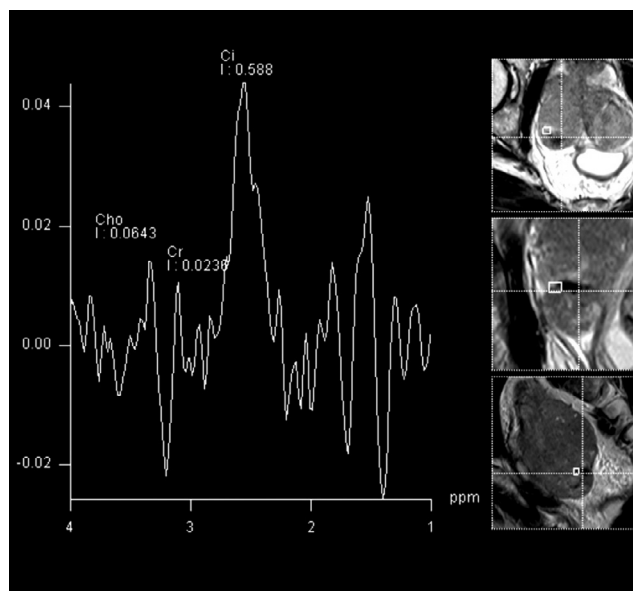


Fig. 9 – MR spectroscopy shows an elevated peak of citrate at the peripheral solid zone.

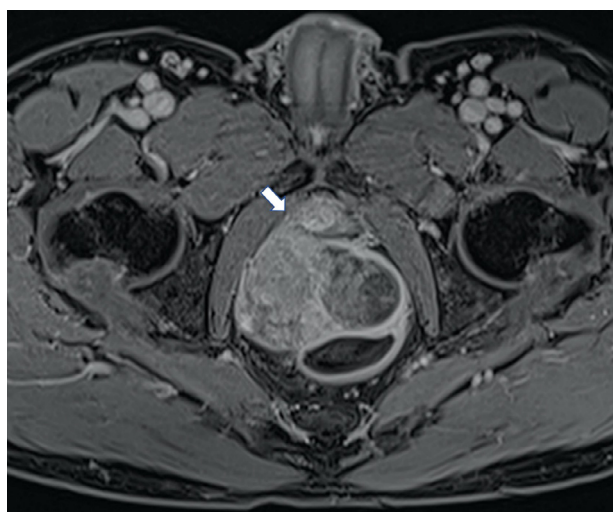


Fig. 8 – Axial T1W after Gadolinium intravenous administration. The tumor showed heterogeneous enhancement. The cystic component did not enhance. Claw sign (arrow) suggesting prostatic origin.

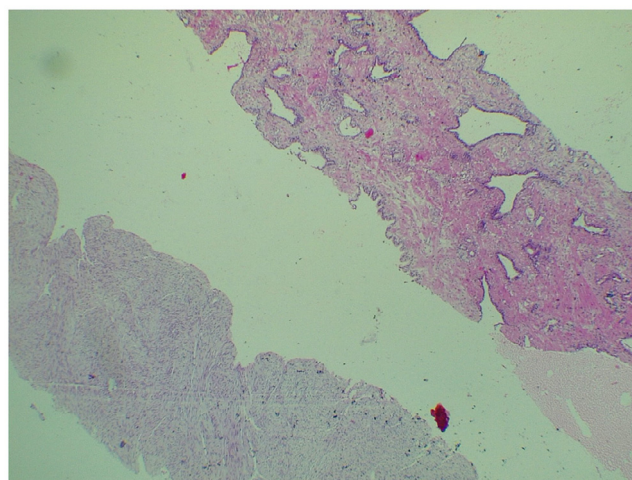


Fig. 10 – Biopsy histopathology shows the tumor cells (bottom left corner) and the prostate tissue (upper right corner), H.E x 40.

mon in large tumors. Calcification is uncommon. GISTs usually show strong enhancement [16]. On magnetic resonance imaging (MRI), GISTs are usually heterogeneous due to cystic degeneration or hemorrhage. Tumors are hypointense on T1W imaging and hyperintense on T2W imaging. Solid component may show restricted diffusion. Peripheral enhancement is typical with central areas of nonenhancement due to necrosis or cystic degeneration [17]. GISTs are characterized by their expression of KIT (CD117), a tyrosine kinase growth factor receptor [15].

Extragastrintestinal stromal tumors share similar histological and immunophenotypic features as GISTs. However, EGISTs may exhibit a more aggressive clinical course. Metastasis may present at the time of initial diagnosis [7]. Radiologic diagnosis of EGISTs is challenging. Differential diagnosis includes GI lymphoma, carcinoid, and GI carcinoma. Prostatic EGISTs are extremely rare. To our knowledge, only 14 cases have been reported in the English literature [13]. Therefore, our case report should be the 15th case of reported prostatic EGISTs in the literature. For prostatic EGISTs, differential diagnosis should also include prostatic cancer and GIST arising from the rectum and invading the prostate. Patients with prostatic EGISTs usually show normal or mild elevated

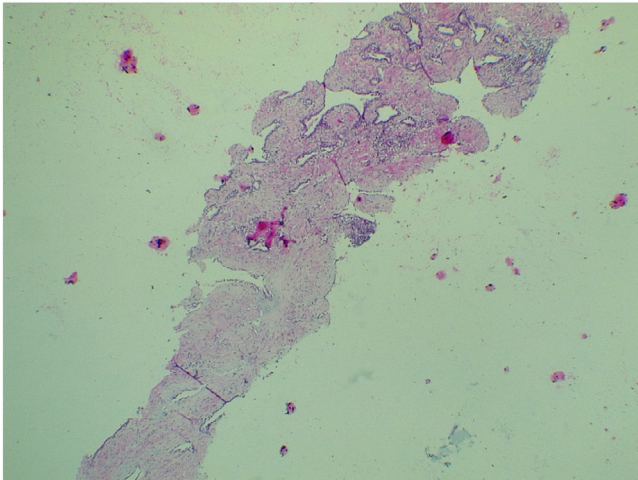


Fig. 11 – Biopsy histopathology shows the tumor (bottom right corner) in continuity with the prostate tissue (upper left corner), H.E \times 40.

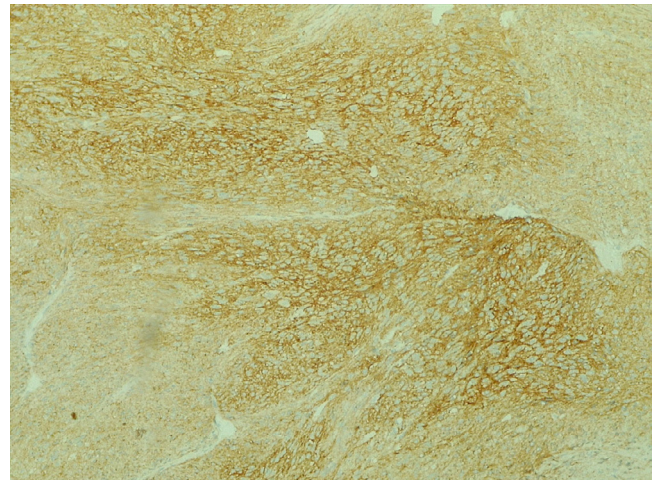


Fig. 13 – Immunohistochemistry staining strong positive for CD117, IHC \times 100.

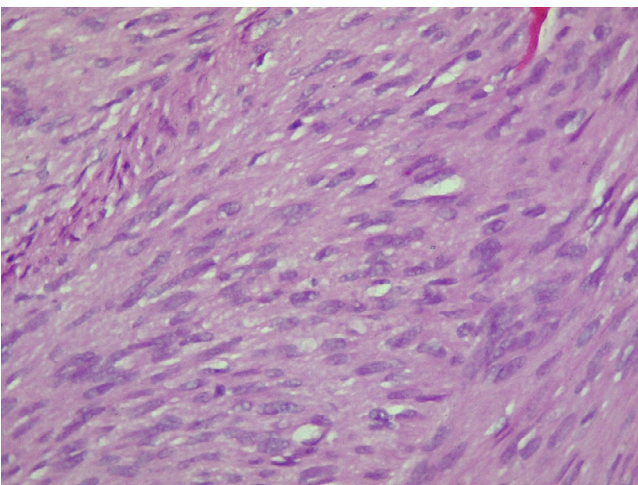


Fig. 12 – Pathological result shows spindle cell proliferation, HE \times 400.

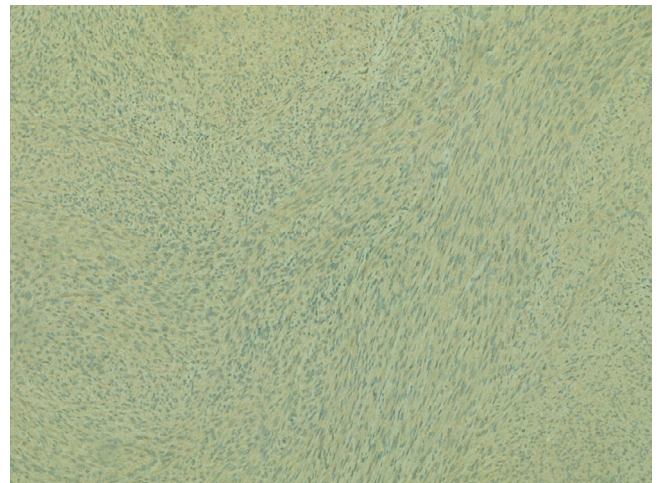


Fig. 14 – Mild diffuse CD34 expression of the tumor cells, IHC \times 100.

prostate-specific antigen level [13]. MRI with gadolinium intravenous administration provides excellent soft tissue differentiation, hence useful in accessing pelvic masses. The “claw sign” is useful in determining the origin of the tumor. Furthermore, magnetic resonance spectroscopy may provide an adjunct to conventional MRI in analysis of metabolic changes within tumors. Although citrate can be found in different tissues, prostate gland shows characteristically high citrate concentration [18]. Therefore, an elevated citrate peak in MR spectroscopy is highly suggestive of prostatic origin [19].

In conclusion, although EGISTs originating from prostate are rare, they should be included in the differential diagnosis of prostatic masses.

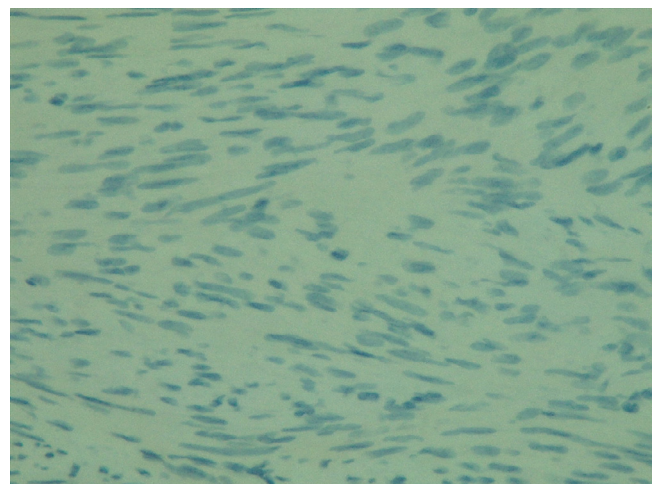


Fig. 15 – Desmin negative, IHC \times 400.

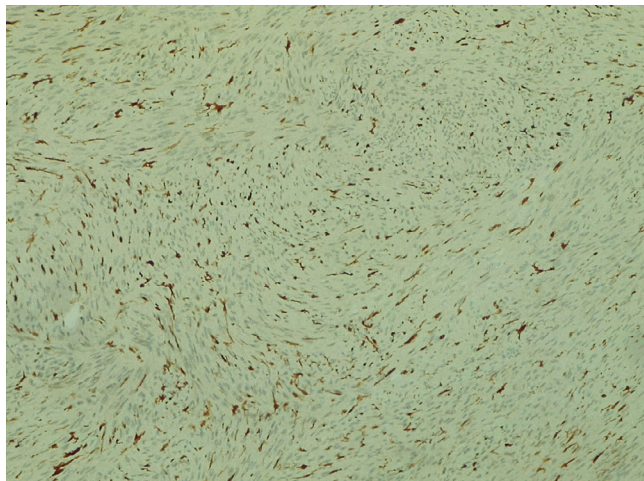


Fig. 16 – S100 mild positive, IHC x 100.

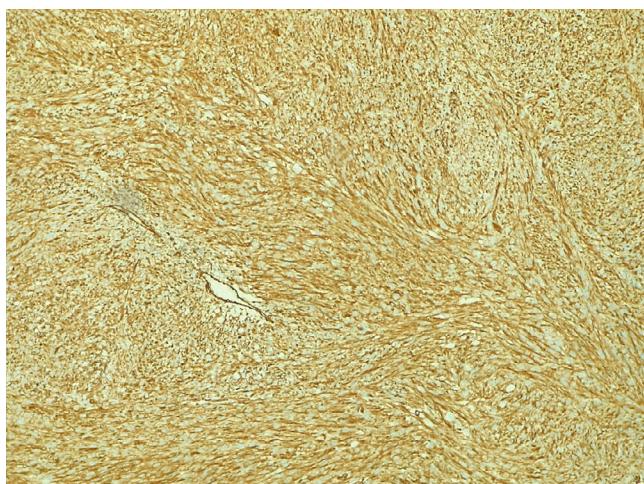


Fig. 17 – Vimentin positive, IHC x 100.

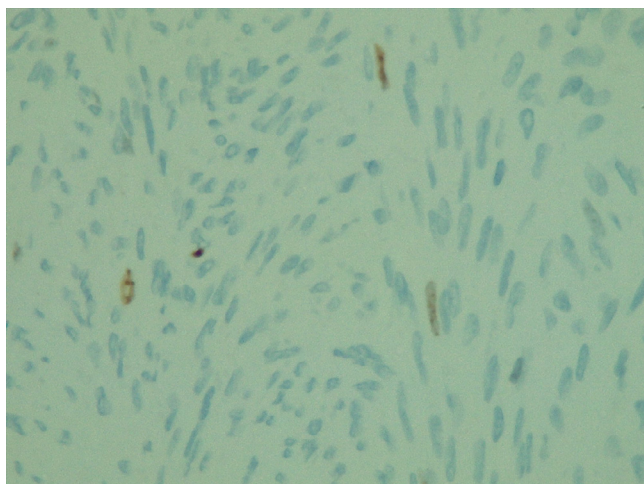


Fig. 18 – Ki67 sporadic positive, IHC x 400.

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