Extragastrointestinal stromal tumor arising in the rectovaginal septum

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Citation

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Abstract

Extraintestinal gastrointestinal stromal tumor of the rectovaginal septum presenting as a vaginal mass is unusual. We describe a case in a 45 year old woman who presented with a recurrent vaginal mass. This tumor was characterized by intersecting fascicles of spindle cells, no necrosis and a mitotic count of 8/50 high power fields. Immunohistochemistry revealed diffuse cytoplasmic positivity for CD117 (c-Kit). An accurate diagnosis is clinically important as these tumors recur inspite of complete excision. Kit tyrosine inhibitor (imatinib) is the treatment of choice in recurrent and advanced disease.

INTRODUCTION

Gastrointestinal stromal tumors (GIST) are neoplasms arising from the interstitial cell of Cajal and are positive for CD117 (c-Kit). Extraintestinal gastrointestinal stromal tumors (EGISTs) are uncommon and unique neoplasms, usually involving the mesentery, omentum, retroperitoneum, rarely bladder [1] and inguinal hernial sac. EGISTs arising in the rectovaginal septum and presenting as a recurrent vaginal mass is unusual. Till date only 9 cases of EGIST presenting as vaginal mass have been reported [1] of which 4 cases presented as recurrent tumors. Histologically, these tumors are composed of spindle cells arranged in a fasicular pattern and hence mimic other vaginal spindle cell neoplasms. The diagnosis of EGIST has to be entertained in the differential diagnosis of recurrent vaginal spindle cell neoplasms as the clinical management is different for these tumors.

CASE SUMMARY

A 45 years old woman presented with history of chronic pelvic pain . She was operated 5 years earlier for a vaginal cyst which was reported as neurilemmoma. Pelvic examination revealed whitish discharge and a firm non tender mass measuring 10x8cms, arising from the rectovaginal septum and bulging through posterior vaginal wall. The rectal mucosa was uninvolved. Ultrasonography revealed a well defined lobulated mass posterior to urinary bladder with heterogenous echotexture. Fine needle aspiration cytology (FNAC) of the mass was reported as spindle cell tumor, possibly of benign nature. On excision, the mass was not well delineated , but adherent to rectum and posterior vaginal wall.

PATHOLOGICAL FINDINGS

The resected specimen consisted of multiple grey white bits of tissue, largest measuring 4.5 x2.5 x1.5 cms. Cut section revealed grey white and hemorrhagic areas with no apparent whorling pattern. Microscopy revealed a tumor composed of spindle cells arranged in short and long fascicles interspersed with hypocellular areas and a mitotic count of 8/50HPF. The tumor cells showed moderate cytoplasm , elongated vesicular nuclei and inconspicuous nucleoli (Fig 1). No necrosis was observed. The tumor cells showed strong cytoplasmic positivity for CD117 (Fig 2). They were negative for desmin and S-100 (Fig 3 & 4).

Figure 1

Figure 1: Spindle shaped tumor cells arranged in fascicles (H&E 400)

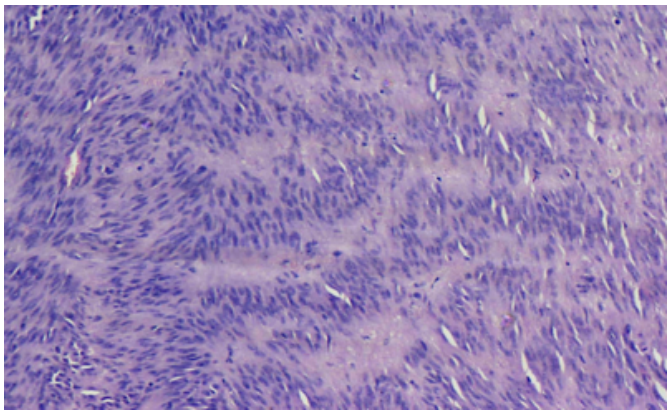


Figure 2

Figure 2: CD 117 positive tumor cells

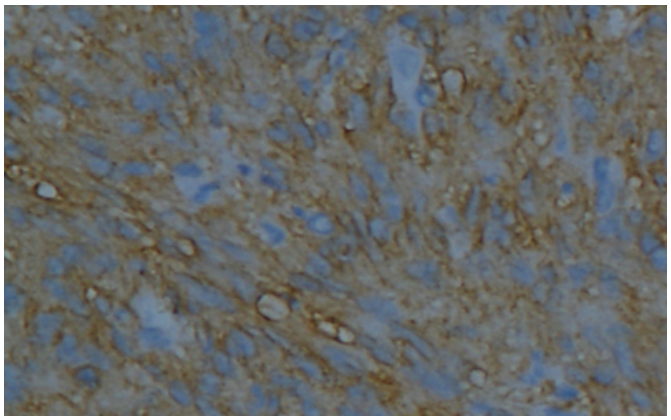


Figure 3

Figure 3: Desmin negative tumor cells

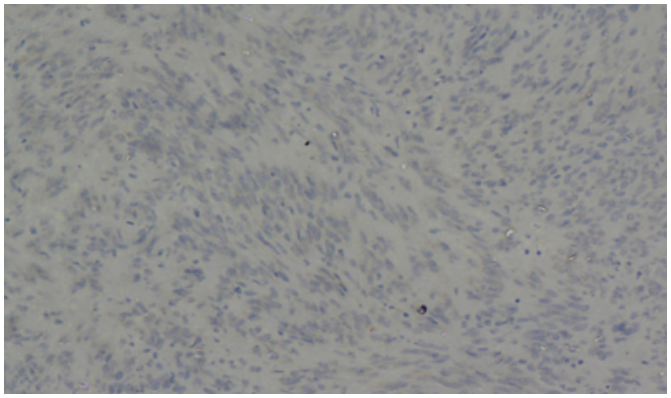
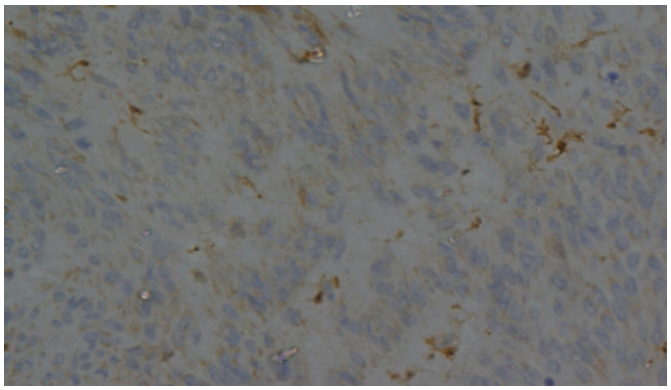


Figure 4

Figure 4: S-100 negative tumor cells



DISCUSSION

5-7% of all GISTs occur as primary tumors outside the GI tract [1]. EGISTs are uncommon neoplasms and the largest series document 48 cases [2]. Our case is unique because of unusual anatomic location and presentation. Only 9 cases of GIST presenting as vaginal mass has been documented. The spindle cell pattern exhibited by GISTs causes diagnostic confusion as it mimics other spindle cell mesenchymal tumors in the vagina. This can easily be resolved by the help of immunohistochemical marker CD117 (c-kit) which exhibits positivity within the cell cytoplasm and accentuation of the golgi region . Other smooth muscle and neural tumors were ruled out as the tumor was desmin and S-100 negative.

Reith et al [2] reported that frequent mitotic activity (>2/50 HPF), high cellularity and the presence of necrosis indicate a potentially aggressive clinical course for EGIST and that 92% of patients with two or more of these three features have a poor outcome. The current definitive treatment for EGIST is surgical resection , but in majority of the patients, the tumors recur despite complete resection [3]. An accurate diagnosis is mandatory for EGIST as these are tumors with an aggressive course and a potential for recurrence inspite of complete surgical excision. In addition, the treatment strategy includes kit tyrosine inhibitor, Imatinib which is used for recurrent and advanced disease. This case is highlighted for its unusual location and presentation as well as to caution pathologists when analyzing spindle cell lesions of the vagina as a correct diagnosis is essential for the management of these tumors.

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