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Resident Quiz Case

Uterine Mass! Spectrum of Disease

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Case History

A 45-year-old female para 4 presented in surgical Outpatient Department at Abbas Institute of Medical Sciences, Muzaffarabad with history of hypogastric pain for 9 months associated with pervaginal bleeding and feeling of lower abdominal heaviness. She had also started noticing a mass lower abdomen gradually increasing in size. The ultrasound abdomen reported huge fibroid in the uterus. She was Hep-C positive with normal routine laboratory investigations. s.

Gross Description

Received in formalin was 15x9.5x9cm firm to hard tan white uterus (Fig.1) without grossly visible cervix and no attached adnexae. There was 11x7x5.4cm tumor. The cut sections showed whorling pattern, friability, completely distorting and pushing endometrial cavity to one side.

Please see microscopic pictures 1-3

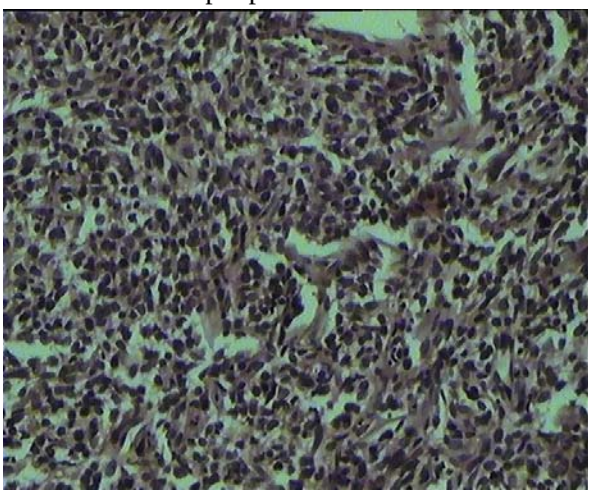

Figure.1

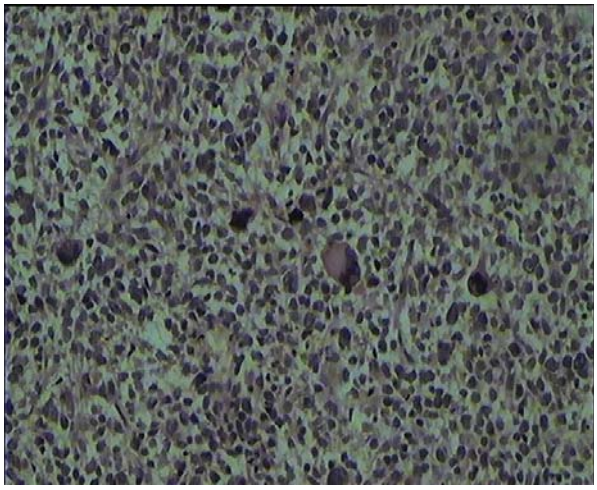

Figure 2

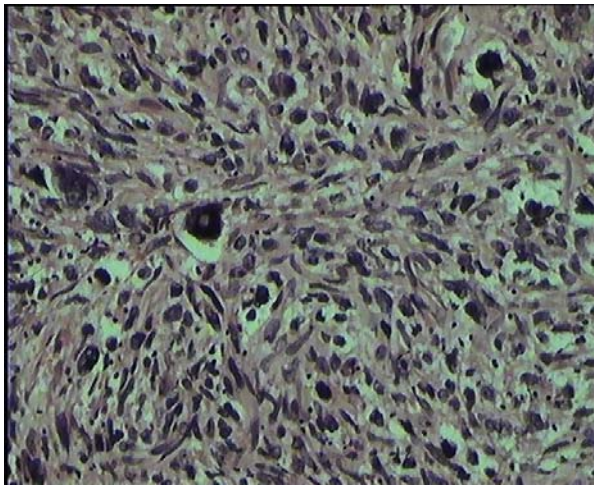

Figure 3

Questions

1. What is the most likely diagnosis?
2. What are your differentials in relation to site, type of lesion and case history of the patient?
3. How would you confirm the diagnosis?

Answers

The most likely diagnosis is leiomyosarcoma. This rare tumor of uterine smooth muscle accounts for 1% to 2% of all uterine malignancies.[1]

The diagnosis of uterine sarcomas is made from histologic examination of the tumor with special emphasis on the cellularity, mitotic counts and nuclear atypia. The differential diagnosis includes other malignant tumors including poorly differentiated endometrial carcinoma, cervical carcinomas, Yolk Cell tumor, mixed Mullerian tumor, choriocarcinoma, lymphoma and metastatic tumors. Confirmation of the origin of the tumor is based on positive stains for Smooth muscle actin, myosin, desmin, Ki-67, ER, PR and EMA. p53 mutations and p16 over expression is noted in 25-47% patients.

Discussion

Peak age is 40-69 years with mean age of 54. High risk factors include prior pelvic radiation (10%-25% of cases), use of long-term adjuvant tamoxifen in women with breast cancer[2-3] and 1% to 2% in postmenopausal women. Parity, early menarche or menopauses are not associated with increased risk of developing leiomyosarcoma. These patients usually present with abnormal uterine bleeding as our patient presented having abnormal pervaginal bleeding. It is a rapidly growing tumor with a doubling time of four weeks as noticed by our patient. 5 year survival rate is 40% with only 10% if anaplastic. Minimal survival if tumor extends beyond uterus. This tumor is very aggressive even if confined to uterus. Recurrence rate of 53% to 71% is documented. Tumor size, tumor grade and stage are major prognostic parameters.[4] Patients with germ line mutations of fumarate hydratase have increased risk for uterine leiomyosarcomas and uterine leiomyomas.[6] Treatment includes total abdominal hysterectomy and debulking of tumor, chemotherapy with doxorubicin or docetaxel/ gemcitabine is used for advanced or recurrent disease. Radiotherapy may be useful.

Surgery is the only treatment and prognosis depends on disease extend at the time of diagnosis, tumor size >5cm and mitotic index >10/ high power field. Nonrandomized studies have reported improved survival after adjuvant chemotherapy with or without radiation therapy. The value of pelvic radiation therapy has not been established. Current studies consist primarily of phase-II chemotherapy trials for patients with advanced disease.[5]

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