Rare and unusual spindle cell tumour

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CLINICAL QUESTION

A 50-year-old woman presented with a very hard goose-egg size bulky mass of the uterine cervix, involving the uterine corpus and vaginal wall. Histology specimens were obtained via needle biopsies of the cervix. Review the high-quality, interactive digital Aperio slides at http://virtualacp. com/JCPCases/jclinpath-2019-206144_A and http://virtualacp.com/JCPCases/jclinpath-2019-206144 B and consider your diagnosis.

WHAT IS YOUR DIAGNOSIS?

- A. Chronic cervicitis
- B. Soft tissue sarcoma
- C. Spindle cell carcinoma
- D. A spindle cell variant of diffuse large B-cell lymphoma (DLBCL)
- E. Spindle cell melanoma
- The correct answer is after the discussion.

DISCUSSION

The adnexa was the most common site (56%) of primary genital tract lymphoma, followed by the corpus (15%) then the cervix (11%).¹ DLBCL evidently accounts for most primary lymphomas of the cervix (63%).¹ Spindle cell variant (sarcomatoid variant) DLBCL of the genital tract is extremely rare. There are a few case reports in the literature.^{2–4} Spindle cell lymphoma is associated with somatic immunoglobulin and *BCL6* gene mutations as well as BCL6 protein expression, and thus neoplastic spindle cells closely reflect B cells residing in the germinal centre.⁵ This indicates that spindle cell lymphoma is a good responder to chemotherapy and has a good prognosis.

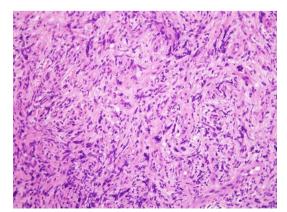


Figure 1 Spindle cells proliferate showing vague storiform pattern with fibrosis and small lymphocytes infiltration in the background.

Take home messages

- Diffuse large B-cell lymphoma (DLBCL) with an unusual prominent spindle cell morphology with marked sclerosing of genital tract often leads to misdiagnosis.
- On clinical presentation, most patients with genital lymphoma exhibit abnormal genital bleeding and have a large bulky mass of the cervix resulting in what is known as a 'barrelshaped' cervix.
- Spindle cell variant DLBCL presented as a subepithelial mass without obvious ulceration or epithelial abnormality; therefore, the diagnosis may require deep cervical needle biopsy.
- Immunohistochemical analysis can efficiently distinguish spindle cell variant DLBCL from other spindle cell neoplasms.

On clinical presentation, most patients with genital lymphoma exhibit abnormal bleeding and have a large bulky mass of the cervix resulting in what is known as a 'barrel-shaped' cervix on pelvic examination.

Spindle cells with a vague storiform pattern of DLBCL are not familiar to pathologists. The diagnosis of this condition may be challenging. The differential diagnosis of spindle cell lymphoma of the uterine cervix includes chronic cervicitis, soft tissue sarcoma, spindle cell carcinoma, spindle cell melanoma and dendritic cell tumour. However, morphological observations and immunohistochemical analysis can efficiently distinguish malignant lymphoma from these other lesions.

The diagnosis of this variant of cervical lymphoma may require deep needle biopsy. The most important point is that it is a lymphoma with an unusual morphology, composed of spindle cells. A prompt and accurate diagnosis can lead to immediate treatment without requiring surgery.

CORRECT ANSWER

D. A spindle cell variant of diffuse large B-cell lymphoma (DLBCL)

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Virtual case of the month

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