

## Vaginal Mass in a 13-year-old Girl

A 13-year-old girl, otherwise healthy, presented with complaints of a painless vaginal mass associated with intermittent abnormal vaginal bleeding and discharge over a few weeks. External examination of genitalia was unremarkable and pubertal development was appropriate for her age. No obvious abnormality was detected on recto-abdominal examination. No transvaginal examination was attempted in view of the patient's age and she was directly referred to a trans-perineal ultrasound scan that showed an ill-defined lobulated heterogeneous vaginal mass. On further evaluation with dynamic contrast magnetic resonance imaging (MRI) of the pelvis, a lobulated polypoid mass was noted protruding from the cervical orifice into the upper two thirds of the vagina. This mass demonstrated heterogeneous high T2 (Fig. 1a) and low T1 (Fig. 1b) signal intensity with mild central enhancement on delayed post contrast sequences (Fig. 1c). The rest of the pelvic organs were unremarkable.

Later, the patient underwent vaginal examination under anaesthesia followed by colposcopy and hysteroscopy that showed a fleshy polypoidal mass protruding from the endocervix into the vagina, which was subsequently resected. The vagina and uterine cavity were found to be unremarkable.

What is the diagnosis of the mass?

- A. Benign endocervical polyp
- B. Mullerian adenosarcoma
- C. Embryonal rhabdomyosarcoma
- D. Endometrial stromal sarcoma
- E. Haemangioma/venous malformation

The gross specimen appeared fleshy and gelatinous in consistency with dark haemorrhagic/ necrotic periphery and grey-white to tan central area, measuring 4.2 cm x 3.4 cm x 2.1 cm in size (Fig. 2). The final pathology revealed a Mullerian adenosarcoma (Figs. 3a and 3b) of endocervical origin.

Further investigations, including abdominal ultrasound as well as computed tomography (CT) thorax and abdomen were negative for nodal and distant metastasis. Clinically, there was no evidence of residual disease at the time of discharge. However, the patient was kept on close follow-up in view of high potential for recurrence of the primary lesion.

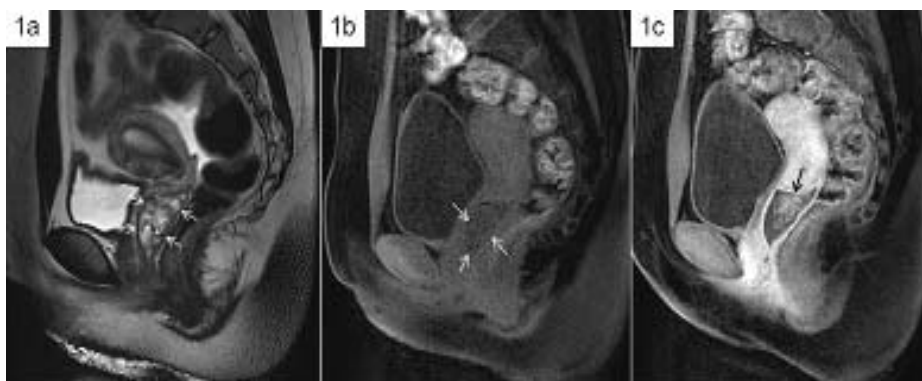


Fig. 1. MRI pelvis sagittal T2-weighted image (A) demonstrates a heterogeneously hyperintense lobulated mass in the upper two-thirds of the vagina (arrows) that is slightly low signal on T1-weighted images (B). Post contrast sagittal T1 fat saturation image (C) demonstrates delayed central enhancement of the lesion (arrow) close to the uterine cervix.

Answer: B

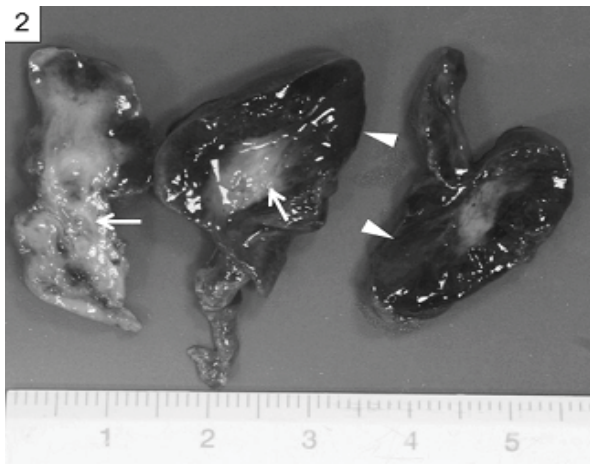


Fig. 2. Gross postsurgical specimen shows cut surfaces of the polypoid lesion demonstrating outer zone of haemorrhagic necrosis (arrowheads) and inner pale tan tissue (arrows).

## Discussion

Adenosarcoma is an intermediate form of mixed Mullerian tumour with the benign adenofibroma at one end and aggressive carcinosarcoma at the other end of the spectrum.<sup>1</sup> Mullerian adenosarcoma is a biphasic tumour comprising benign glandular epithelium and low-grade malignant stromal component, usually involving the uterine corpus with rare occurrence in the uterine cervix, as in our case. Even rarer is occurrence in the extrauterine sites, such as in the vagina, ovary, fallopian tube and peritoneal surface. Cervical origin accounts for about 2% to 9% of all Mullerian adenosarcoma locations; and the younger age of presentation is more common with cervical lesions than other locations.

So far, only a few cases of cervical adenosarcomas have been reported in adolescents and paediatric patients in the literature, with the youngest patient reported being a 10-year-old girl.

Pelvic irradiation, hyperestrogenism, long-term oral contraceptive use and treatment with Tamoxifen have been found to be associated with Mullerian adenosarcoma of the uterus in elderly patients. However, these may be coincidental and no definite causal relationship or proven aetiological factors have been established. Unlike in cervical adenocarcinoma, human papilloma virus has not been implicated in the aetiology and prognosis of these mixed Mullerian tumours.

The patient may present with an abnormal vaginal discharge and/or bleeding with or without an introital mass. On MRI, typical low-grade adenosarcoma appears as a solitary polypoid mass containing a few tiny scattered cysts and rarely haemorrhagic foci. The solid component of the lesion is heterogeneously hyperintense on T2-weighted images and iso/hypointense on T1-weighted images relative to the myometrium.<sup>2</sup> The lesion is usually less vascular as demonstrated by the colour Doppler ultrasound and may show delayed heterogeneous enhancement on dynamic contrast MRI scan. Most of these MRI features were well demonstrated in our case.

Histologically, this mixed tumour is composed of benign glands distributed in a malignant stroma. The glands may be cystically dilated or show cleft like spaces, and are lined by bland epithelial cells usually of endometroid type. The stroma is cellular in general and characteristically shows cellular condensation around glands and beneath epithelium typical of Mullerian adenosarcoma. In most

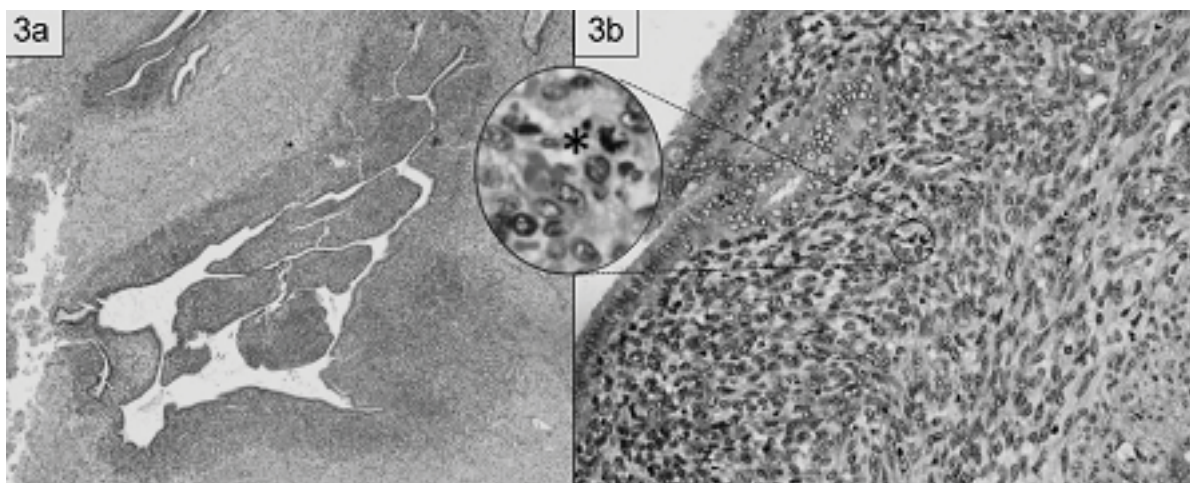


Fig. 3. Low power view of histopathology slides. (A) displays a dense periglandular condensation of stromal cells with polypoidal projections into the gland. (B) shows a high power view of mild nuclear atypia of the stromal cells and a mitotic figure (asterisk in the magnified window).

tumours, the stromal cells show mild to moderate nuclear atypia. Mitotic figures are readily identified in most cases, particularly in the hypercellular stromal cuffs around the glands. The stromal component of this neoplasm may be homologous, i.e. containing indigenous uterine elements or heterologous with differentiation into extrauterine stromal tissues, such as cartilage, osteoid and striated muscle.<sup>3,4</sup> Heterologous elements are reported to be present in 20% to 25% of cases. A more aggressive variant of this entity is Mullerian adenosarcoma with sarcomatous overgrowth (MASO), which is associated with high postoperative recurrence and metastases even when diagnosed and treated at an early stage. Sarcomatous overgrowth occurs in approximately 10% of cases and is defined as pure high-grade sarcomatous component constituting 25% or more of the tumour showing increased cellularity, mitotic activity and nuclear atypia compared to the appearance of the background adenosarcoma.<sup>5</sup>

Differential diagnosis includes benign endometrial/endocervical polyps, adenofibroma, embryonal rhabdomyosarcoma and endometrial stromal sarcoma. In benign polyps and adenofibroma, both the epithelial and stromal components are morphologically benign, showing no stromal hypercellularity, periglandular cuffing, mitoses or nuclear atypia. In young patients, an embryonal rhabdomyosarcoma is an important consideration showing significant morphological overlap with adenosarcoma. In rhabdomyosarcomas, glands are not distributed throughout the tumour and sarcoma cells are more primitive and mitotically active than stromal cells of adenosarcoma. Immunohistochemical markers, such as myogenin and MyoD1, would show more diffuse staining in rhabdomyosarcoma. Whereas, the absence of periglandular cuffing, phyllodes-like architecture and even distribution of glands in the tumour differentiates the endometrial stromal sarcoma from a typical Mullerian adenosarcoma.

Although there is no current consensus with regard to the optimal therapy for cervical adenosarcomas, the aggressive lesions may warrant total hysterectomy and bilateral salpingo-oophorectomy in addition to adjuvant chemotherapy and radiation. Whereas the low-grade tumours, particularly in paediatric and younger patients, may be treated with conservative surgical methods such as wedge resection, cone biopsy or trachelectomy, to preserve the patient's fertility. However, review of the literature shows a high recurrence rate, often late, following conservative surgical management. High-grade malignant features in the stromal component, depth of invasion and sarcomatous overgrowth are some of the predictors for tumour recurrence and poor prognosis.<sup>4,5</sup> In view of the known potential for delayed recurrence with conservative management, long-term close follow-up of these patients is warranted.

## REFERENCES

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