

# Adenoma Malignum in Peutz Jegher Syndrome: Case Report

## Case Report

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### Abstract

Adenoma malignum, also known as gastric-type mucinous adenocarcinoma of the cervix, is a rare variant of mucinous adenocarcinoma, previously referred to as minimal deviation adenocarcinoma. It accounts for approximately 1–3% of all cervical adenocarcinomas. Adenoma malignum is often associated with Peutz-Jeghers syndrome and mucinous tumors of the ovary. This case is unique due to its prolonged indolent clinical course, with no evidence of local invasion or metastasis over a period of six years, underscoring the deceptively benign clinical, radiological, and pathological appearance of this malignancy. We emphasize the subtle diagnostic clues that can facilitate early recognition of this rare but aggressive entity.

**Keywords:** Adenoma malignum; Cervical cancer; Peutz–Jeghers syndrome

## Introduction

Adenoma malignum of the cervix is a rare subtype of cervical adenocarcinoma. This tumor is characterized by its deceptively benign appearance on imaging and histopathology, often resulting in delayed diagnosis. Adenoma malignum may arise de novo or in association with Peutz–Jeghers syndrome or hereditary intestinal polyposis. We report a case of adenoma malignum in a patient with known Peutz–Jeghers syndrome, with a six-year interval between the initial presentation and definitive diagnosis.

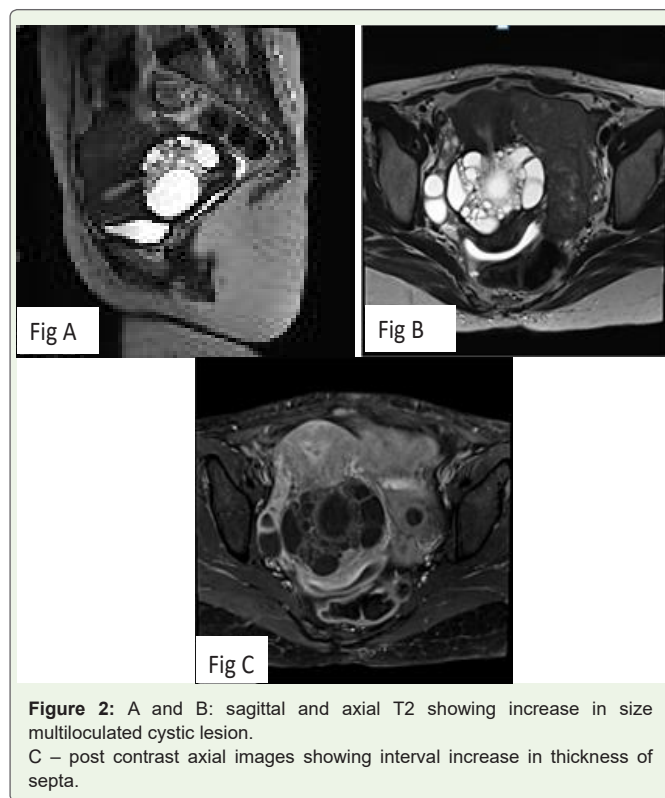
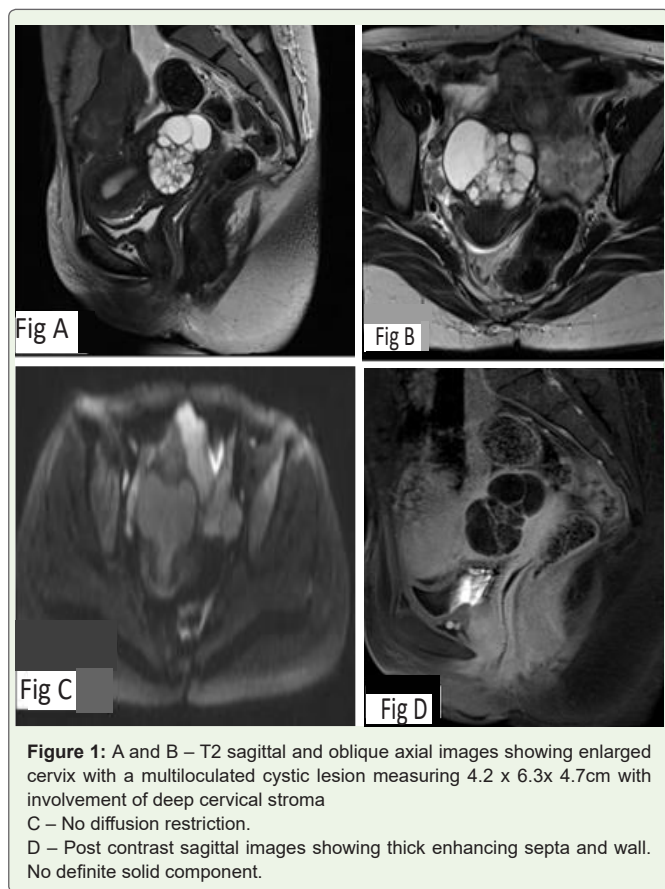
## Case Report

A 45-year-old nulliparous woman, diagnosed with Peutz–Jeghers syndrome (PJS) during adolescence based on perioral pigmentation and multiple gastrointestinal polyps, presented with mild mucinous vaginal discharge. Her past medical history was significant for

multiple admissions due to small bowel obstruction and surgical interventions for polyp-associated intussusception.

On per-vaginal examination, the cervix appeared bulky with mucoid discharge. Transvaginal ultrasound revealed a multiloculated cystic lesion within the cervix. Pelvic MRI was subsequently performed, showing an enlarged cervix with a multiloculated cystic lesion that was T2 hyperintense and T1 hypointense, extending into the deep stroma. Post-contrast imaging demonstrated thick enhancing septa (Figure 1).

Given the patient's background of PJS, a differential diagnosis of adenoma malignum was suggested, despite the absence of a solid component. The patient, however, declined surgical intervention and opted for regular follow-up. The lesion remained stable for nearly three years, after which she was lost to follow-up.



Three years later, she re-presented with profuse mucinous vaginal discharge. Local examination again revealed a bulky cervix, and repeat MRI was performed for reassessment (Figure 2). Compared with the initial MRI, there was mild further enlargement of the lesion with stromal expansion. Post-contrast imaging showed a minimal increase in wall thickness, but no definite solid component was identified. There was no evidence of parametrial extension, pelvic, or inguinal lymphadenopathy. In view of the progressive nature of the lesion and persistent symptoms, adenoma malignum was strongly suspected, and the patient consented to surgery.

Unlike conventional cervical malignancies, adenoma malignum predominantly involves the deep cervical stroma while sparing the mucosa; therefore, surface biopsy may be non-diagnostic and was not attempted.

She subsequently underwent total abdominal hysterectomy.

Gross examination revealed an enlarged cervix with a multiloculated cystic neoplasm measuring 5.6 x 4.5 x 4.5 cm. The cysts ranged in size from 0.3 to 2 cm and contained viscous fluid. The inner walls of the cysts were smooth, with no solid or papillary areas identified. Microscopy showed well-formed glands and cysts of variable sizes haphazardly distributed within the stroma, with minimal desmoplastic response. The tumor cells exhibited minimal cytologic atypia and abundant pale eosinophilic to clear cytoplasm with distinct cell borders. The tumor involved the deep third of the cervical stroma but spared the resection margins (clearance of 0.1 cm). The histopathological features were consistent with gastric-type adenocarcinoma circumferentially involving the cervix, confirming the diagnosis of adenoma malignum. The tumor was staged as FIGO IB2, TNM T1b2N0.

As the surgical margins were negative, the multidisciplinary tumor board recommended follow-up. PET-CT excluded distant metastases and other malignancies associated with PJS. At present, the patient is two years post-surgery and remains disease-free, with no evidence of metastasis.

**Discussion**

Adenoma malignum, also known as gastric-type mucinous adenocarcinoma of the cervix, is a rare variant of mucinous adenocarcinoma, previously termed minimal deviation adenocarcinoma. It accounts for 1–3% of all cervical adenocarcinomas and is often associated with Peutz–Jeghers syndrome and mucinous ovarian tumors [1].

Classically, adenoma malignum has been described as a multilocular cystic lesion with enhancing solid components, cervical enlargement, and deep stromal invasion. In our case, cervical expansion and deep stromal invasion were present; however, solid components were not appreciable, even on retrospective review. The initial MRI revealed a multiloculated cystic cervical lesion with enhancing septa and walls, but without a solid component—features that overlap with benign lesions such as complex nabothian cysts, tunnel clusters, or deep endocervical glandular hyperplasia [2]. Nonetheless, the involvement of deep stroma, along with intense

enhancement of septa and a gradual increase in septal thickness and cyst size on follow-up MRI, raised concern and ultimately led to surgical excision and a definitive diagnosis of adenoma malignum.

Differentiating benign entities from adenoma malignum is particularly challenging when an enhancing solid component is absent. Several reports have documented benign glandular lesions—including tunnel clusters, deep endocervical glands, deep nabothian cysts, and lobular endocervical glandular hyperplasia—that can closely mimic adenoma malignum both histologically and radiologically [3]. Benign lesions generally do not invade the deep cervical stroma, are smaller in size, demonstrate well-defined margins, and typically lack solid components. The transition from hyperplasia to high-grade malignancy appears to represent a continuous spectrum, with the proportion of solid components within a lesion serving as a key determinant.

Profuse mucinous vaginal discharge is the most common clinical presentation. The risk is heightened in patients with Peutz–Jeghers syndrome, who have a significantly increased lifetime risk of gynecologic malignancies, particularly adenoma malignum. Therefore, close surveillance is warranted in such high-risk populations.

MRI plays a pivotal role in early detection. Adenoma malignum often appears as a multicystic lesion with a characteristic “honeycomb” appearance, thin septations, and variable wall or septal enhancement (Table 1). While the presence of a solid component or nodularity raises suspicion for malignancy, cases such as ours—entirely cystic without a solid nodule—pose diagnostic difficulty.

On MRI, these lesions are usually isointense or slightly hyperintense relative to the uterus on T1-weighted imaging and markedly hyperintense on T2-weighted imaging. A T2 hyperintense solid component, when present adjacent to cysts, may enhance following gadolinium administration. Radiologic–pathologic correlation has shown that these apparent solid areas often represent clusters of minute cysts with edematous cervical stroma rather than true solid tissue [4]. Thus, the absence of a solid component on imaging does not exclude malignancy, as microcysts or stromal edema may mimic cystic spaces. However, true solid nests may be observed in aggressive or dedifferentiated forms.

**Table 1:** Imaging features of adenoma malignum and their clinical significance.

Imaging feature	Significance
Multiloculated cystic lesion	Can mimic benign conditions, but is not specific
Wall and septal enhancement	Benign lesions usually don't enhance significantly.
Irregular or thickened septa	Red flag- Suggests neoplastic process
Infiltrative or ill-defined margins	important clue against benignity
Deep endocervical stromal invasion	important clue against benignity
Large lesion	Larger lesions are more likely malignant
Asymmetry or distortion of cervical architecture	Suggestive of invasion
No diffusion restriction	Common in well-differentiated tumors like adenoma malignum.

In a study by Sugiyama et al., the MR findings of pseudoneoplastic glandular lesions were reported to be almost identical to those of adenoma malignum, making differentiation extremely challenging [5].

Histologically, adenoma malignum is so well-differentiated both structurally and cytologically that a diagnosis of malignancy can only be established based on the presence of distorted, irregularly outlined glands deep within the cervix or atypical cells. Its deceptively benign appearance often leads to diagnostic difficulty.

Routine cytological screening is of limited value in adenoma malignum. Therefore, a deep biopsy is essential when clinical suspicion is high, especially in patients presenting with profuse vaginal discharge and a multilocular cystic cervical mass [6]. Some studies suggest that the prognosis of adenoma malignum is better than that of conventional cervical adenocarcinoma [6]. Reports indicate that up to 60% of adenoma malignum cases with a solid component present with metastases [3]. However, our case demonstrated an indolent course, with no evidence of metastasis even after six years from initial presentation and two years postoperatively.

The mainstay of treatment for adenoma malignum is surgical excision. However, no standardized surgical protocol has been established due to its rarity, frequent preoperative misdiagnosis, and limited data on surgical outcomes [7]. Due to the rarity of the disease, standardized surveillance and follow-up guidelines are not well established. Common sites of recurrence include local pelvic recurrence, pelvic lymph nodes, and para-aortic lymph nodes. MRI is the preferred modality for detecting local recurrence, while CT imaging is recommended for evaluating distant metastasis.

**Conclusion**

Adenoma malignum is a rare entity with significant diagnostic challenges, primarily due to its overlap with benign imaging appearances. The key imaging feature is a multiloculated cystic lesion of the cervix with deep stromal invasion. An enhancing solid component may also be present. A high index of clinical suspicion, particularly in genetically predisposed patients, along with the presence of mucous vaginal discharge is crucial for diagnosis. Early surgical intervention may be warranted to prevent diagnostic delays and improve prognosis.

**Conflict of Interest:** The authors have no conflict of interest.

**Declaration of the patient consent form:** Patient consent form attached.

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