Perinodular hydropic degeneration in uterine leiomyoma causing rapid enlargement and mimicking a myxoid smooth muscle tumor: Case report of a diagnostic challenge

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ABSTRACT

Introduction: Hydropic degeneration, sometimes reported as perinodular hydropic degeneration, is one of the various degenerative changes that can be observed in uterine leiomyomas. It can be a clinical and a histopathological diagnostic pitfall.

Case Report: Here we present the case of a 40-year-old woman with leiomyoma uteri with perinodular hydropic degeneration. The tumor almost doubled in size over 2 months. Histopathological examination revealed fascicles, cords and nodules of smooth muscle cells separated by excessive amounts of extracellular material. No necrosis or significant atypia was noticed. Average mitotic activity was very low. A myxoid smooth muscle tumor could be excluded only by alcian blue special stain. The presence of floating vessels in the edematous fluid as well as hyalinization inside the tumor nodules were among the reassuring features supporting the diagnosis.

Conclusion: The pathologist must be aware of this benign entity to avoid overdiagnosis, especially in the view of the alarming histology and the rapid growth. Negative findings with alcin blue special stain help establish the diagnosis.

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1. Introduction

Degenerative changes in uterine leiomyomas are common. Of the changes that can be observed, hydropic degeneration, sometimes reported as perinodular hydropic degeneration, poses a clinical and a histopathological diagnostic challenge [1,2]. If significant amount of edema fluid accumulates in a benign leiomyoma, the separated fascicles and/or bundles of tumor cells will show a multinodular gross as well as microscopic appearance. Here we present the case of a 40-year-old woman with a rapidly enlarging uterine fibroid. The histopathological examination revealed leiomyoma uteri with perinodular hydropic degeneration. The pathologist should be aware of such a diagnostic pitfall.

2. Case presentation

A 40-year-old previously healthy woman, gravida 3, para 3, presented with a 1-year history of menorrhagia and feelings of pelvic discomfort and heaviness. No change in bowel habits nor urinary symptoms was documented. The pelvic examination revealed a large palpable, mobile abdominopelvic mass. Laboratory tests were within normal limits except for mild anemia (Hb 10.6). Ultrasonography revealed a bulky uterus with a hypoechoic, well circumscribed mass in the posterior uterine wall that measured 6 × 6 cm. Two months later, ultrasonography was repeated, and the previously described mass had enlarged in size to 10 × 9 cm. Despite the rapid enlargement, the patient underwent a myomectomy to be able to get pregnant. The procedure was uneventful. Grossly, the mass had a whist cut surface with ill-defined nodularity and scattered small cysts containing watery fluid. The tumor appeared to be oozing the watery fluid more when squeezed. The microscopic examination revealed a smooth muscle tumor with solid areas adjacent to areas showing nodules and fascicles of tumor cells floating in excessive amounts of extracellular material (Fig. 1). The separated fascicles were delicate, and many of the nodules showed hyalinization and/or thin-walled to hyalinized blood vessels. The accumulation of material was focal and in association with hyalinization. In addition, the material was pale to eosinophilic. Red blood cells and inflammatory cells, including occasional hemosiderin-laden macrophages, were seen floating in the surrounding fluid.
in the accumulated fluid. Although dysmorphic or degenerated tumor cells were identified, the atypia was minimal, and coagulative necrosis was not identified. The average mitotic activity was far less than 2/10 HPFs. The solid areas also contained a prominent pale myxoid-like matrix (Fig. 2). Alcian blue special stain for acid mucin was performed and the results were negative (Fig. 2). The diagnosis of leiomyoma uteri with perinodular hydropic degeneration was established, and the patient was doing well at 3 months post-myomectomy.

3. Discussion

Perinodular hydropic degeneration imposes a diagnostic difficulty because of its alarming histological appearance. Moreover, it sometimes behaves in ways that suggest malignancy. Hydropic change is poorly reported as perinodular hydropic degeneration [2–5], and many cases have been documented under names such as multinodular hydropic, multicellular cystic, cystic degenerated, or hydropic leiomyomas [5–8].

3.1. Rapidly enlarging myometrial tumors

A leiomyosarcoma should be suspected for any rapidly enlarging uterine mass, especially when it occurs in a postmenopausal woman. However, rapid tumor growth can occur in benign tumors, as in our case. Pregnancy and hormonal therapy are well documented as causes of enlargement of benign leiomyomas. Infarction and cystic change in a benign fibroid can result in rapid enlargement. Moreover, benign degenerative changes can sometimes give a heterogenous appearance on magnetic resonance imaging. However, the absence of invasion or extrauterine extension may point to a benign condition [9].

3.2. Hydropic leiomyomas: typical features and clinical outcome

These tumors typically show excessive amounts of edema fluid separating fascicles in a delicate filigree pattern. Thick tumor fascicles are generally not found. In addition, the presence of floating blood vessels in the extracellular material, as in our case, suggests hydropic change. The presence of hyalinization is another reassuring feature [10]. Because of the striking microscopic similarity with the more aggressive myxoid smooth muscle tumors [11], performing alcian blue special stain was fundamental in excluding this possibility. Another microscopic feature in our case was the presence of red blood cells, inflammatory cells and hemosiderin-laden macrophages floating in the extracellular material.

Initially described by Clement et al., the behavior of leiomyomas with perinodular hydropic degeneration was not different from that of typical leiomyomas [1]. However, Coad et al. reported the first case showing extrauterine extension, which was initially alarming because it was suspected to be an aggressive tumor [2]. A similar behavior was reported by Jashnani et al [3]. Furthermore, Kim et al. described cotyledonoid dissecting and intramural dissecting leiomyomas with perinodular hydropic degeneration [4]. This was also described by Ceyhan et al. They documented the occurrence of benign satellite nodules resembling intravenous leiomyomatosis or even vascular invasion [5]. Although leiomyomas with perinodular hydropic degeneration may undergo extrauterine extension, the tumor in the present case was relatively well circumscribed and was limited within the myometrium. However, the rapid enlargement and the relatively large size in the present case represent another problematic issue regarding the behavior of these tumors.

In conclusion, the pathologist should be aware of perinodular hydropic degeneration in uterine leiomyomas to avoid overdiagnosis, especially in view of the alarming clinical and histological features. The tumor in our case demonstrated rapid growth, and was morphologically similar to a myxoid smooth muscle tumor. Alcian blue is a simple stain that can help establish the diagnosis.

Contributors

Ali Al Khader contributed to conceptualization, data curation, investigation, methodology, supervision, validation, visualization, writing the original draft, and review and editing.

Esra Nsour contributed to investigation, methodology, validation, writing the original draft, and review and editing.

Abd-Naser Abdallat contributed to investigation, methodology, validation, writing the original draft, and review and editing.

All authors saw and approved the final manuscript.

Conflict of interest

The authors declare that they have no conflict of interest.

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Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.
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