Case report

Botryoid rhabdomyosarcoma of the uterine cervix: Report case

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ABSTRACT

Rhabdomyosarcoma of the cervix is among the rare histological types of cervical cancers, which are usually seen in young girls or women in the general active period. The usual circumstances of diagnosis are dominated by recurrent metrorrhagia. At the initial stage, the lesions may be asymptomatic or take on the appearance of a benign polyp. In the case reported here, the clinical symptomatology was recurrent metrorrhagia with a cervical implanted polyp occurring 28 months after resection of a benign polyp.

RMS is one of the malignant tumors of mesenchymal origin. Typically, it is a malignant tumor proliferation of cells with morphological and/or phenotypic striated muscle differentiation. The characteristic cells of this tumor are rhabdomyoblasts. Rhabdomyosarcomas are classified into three histological subtypes: embryonal, alveolar, and anaplastic. Within embryonal RMS, it is possible to distinguish between botryoid, leiomyomatous and anaplastic forms. Botryoid and leiomyomatous forms are classically described as having a more favorable prognosis.

Treatment is based on a multidisciplinary approach that includes indications for conservative surgery, chemotherapy, radiotherapy and brachytherapy.

1. Introduction

Rhabdomyosarcoma is a separate pathological entity according to the 2002 WHO classification and is distinct from other soft tissue sarcomas [1].

Botryoid rhabdomyosarcoma of the uterine cervix is a rare tumor affecting electively young girls or women during their genital activity.

We report the observation of an 18 year old girl followed for a botryoid rhabdomyosarcoma of the cervix. The work has been reported with respect to the SCARE 2020 criteria [2].

2. Patient and observation

Miss A.M., 18 years old, having menstruated at the age of 13 with a regular cycle, presented with a history of a polyp delivered through the bistoury vagina, the histopathological result of which was a simple remodeled polyp with no signs of malignancy.

The patient consulted 28 months later for recurrent metrorrhagia.

The gynecological examination performed in the operating room showed a uterus of normal size on vaginal touch and clean and soft vaginal walls with a polyp at the external os of the uterine cervix, with a sentinel implantation for which a resection was performed.

The histological study of the specimen showed a spindle cell mesenchymal proliferation of poorly differentiated appearance, suggestive of an eroded bot. Poorly differentiated suggesting an eroded botryoid sarcoma.

Botryoid rhabdomyosarcoma of the cervix (Fig. 1) was confirmed by immunohistochemical study, given the positivity of the cells for desmin and vimentin as well as for myosin.

Pelvic magnetic resonance imaging (Fig. 2) showed edema of the posterior lip of the ectocervix without nodule or contrast.

The patient had an abnormal lymph node associated with an effusion at the cul-de-sac of Douglas.

After a thoracic-abdominal-pelvic CT scan, the tumor came back localized.

The patient is still under chemotherapy after agreement of the multidisciplinary consultation meeting.

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early diagnosis. Often, it is a mucosal polyp that recurs very quickly after take on the appearance of a benign polyp. This explains the usual lack of removal [5].

Metrorrhagia. At the initial stage, the lesions may be asymptomatic or their genital activity.

Cervical location is the least frequent. The average age of diagnosis is in the second decade of life, thus affecting young girls and women during their genital activity.

It represents about 0.2 % of all malignant tumors of the uterus [4]. Cervical location is the least frequent. The average age of diagnosis is in the second decade of life, thus affecting young girls and women during their genital activity.

The usual circumstances of diagnosis are dominated by recurrent metrorrhagia. At the initial stage, the lesions may be asymptomatic or take on the appearance of a benign polyp. This explains the usual lack of early diagnosis. Often, it is a mucosal polyp that recurs very quickly after removal [5].

At the gynecological examination, when it is a botryoid form, typically, a mass externalized at the vulva in the form of a cluster is observed. This can then be easily biopsied. If vaginal bleeding occurs, vaginoscopy should be performed and any suspicious vegetation should be biopsied [6].

In the case reported here, the clinical symptomatology was recurrent metrorrhagia with a cervical implanted polyp occurring 28 months after resection of a benign polyp.

4. Pathologically

RMS is one of the malignant tumors of mesenchymal origin. Typically, it is a malignant tumor proliferation of cells with morphological and/or phenotypic striated muscle differentiation. The characteristic cells of this tumor are rhabdomyoblasts, slightly elongated cells with intracellular cross streaks and eosinophilic cytoplasm. These specific striations, the elongated or fusiform appearance of the cell with multiple nuclei are signs of myoblastic maturity found in 50–60 % of embryonic subtypes and 30 % of alveolar subtypes [6].

According to the IntergroupRhabdomyosarcomaStudy (IRS), the International Society of Pediatric Oncology (SIOP), and the National Cancer Institute (NCI), rhabdomyosarcomas are classified into three histological subtypes: embryonal, alveolar, and anaplastic.

Within embryonal RMS, it is possible to distinguish between botryoid, leiomyomatous and anaplastic forms. Botryoid and leiomyomatous forms are classically described as having a more favorable prognosis.

Cyto-pathologically, RMS can be recognized by the expression of myosin and MyoD antigen. The markers used for characterization are usually myoglobin, desmin and actin [7].

In our observation, the study of tissue tumor markers showed positivity to desmin, vimentin and myosin.

RMS of the uterine cervix is a tumor with massive locoregional extension.

The radiological examinations performed at the time of discovery of the tumor will thus serve to assess the local and distant extension of the tumor in order to establish the stage. These examinations, repeated during treatment to monitor the response, will make it possible to consider the type of surgery (conservative or radical) and to monitor the distant evolution [8].

In rhabdomyosarcomas of gynecological origin, MRI is the preferred examination in this region. It has indeed shown its robustness for pelvic examination, but also for all soft tissue tumors [9].

Treatment is based on a multidisciplinary approach that includes indications for conservative surgery, chemotherapy, radiotherapy and brachytherapy [10].

All therapeutic management should be discussed at a multidisciplinary consultation meeting dedicated to sarcoma. This type of tumor has an excellent response to chemotherapy and radiotherapy, with response rates often exceeding 80 % for most patients. The efficacy of chemotherapy has led to a change in aggressive and sometimes disruptive surgical strategies, in contrast to other types of adult sarcoma. Indications for surgery should be given for localized tumors that can be completely resected without disruption. In other cases, biopsy alone should be proposed. Neoadjuvant chemotherapy regimens allow for complete conservative surgery in the majority of cases. Complete surgery is a major prognostic factor, with significantly increased overall survival in patients with complete resection [11].

However, there is currently no codified therapeutic consensus, due to the extreme rarity of these tumors and therefore the absence of randomized studies that have established the superiority of one protocol over another. The RMS Intergroup recommends chemotherapy (VAC) followed by conservative surgery whenever possible and then chemotherapy for 6 to 12 courses [12].

The prognosis for this condition has improved dramatically in recent years, with 5-year overall survival increasing from 25–30 % to almost 70 % in children [9].

5. Finding

Botryoid rhabdomyosarcoma of the uterine cervix is a rare tumor occurring essentially in young girls, which is the case in our patient, and
its therapeutic modalities are not well codified given the rarity of the reported cases. The place of chemotherapy is validated by most authors, contrary to radiotherapy whose effectiveness is still unproven.

Thanks to neo-adjuvant chemotherapy, surgery has become increasingly conservative.

Oncology reports

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Ethical approval

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Author contribution

El Qasseh Rajaa: Corresponding author writing the paper
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The authors declare having no conflicts of interest for this article.

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