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Case Report

Embryonal Rhabdomyosarcoma of the Cervix, Unusual Presentation in Menopausal Patient: A Case Report

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Abstract

Rhabdomyosarcoma (RMS) is a malignant neoplasm originating from undifferentiated myogenic progenitor cells, commonly diagnosed in the pediatric population, its incidence in adults is exceedingly rare. It mainly affects the cerebral and cervical regions, followed by the genitourinary tract.

the majority of genital tract rhabdomyosarcomas occur in the vagina, followed by the cervix and the uterine corpus, RMS of the cervix account for less than 1% of all cervix cancer, and approximately half of them are embryonic type, most commonly present in pre-pubertal or in reproductive age women in the second and third decade of life, rarely in postmenopausal patients.

The treatment ranges from radical surgery to conservative surgery, and neoadjuvant chemotherapy.

Keywords: *Embryonal Rhabdomyosarcoma; MRI; Uterine Cervix*

Introduction

Rhabdomyosarcoma (RMS) of the cervix is a rare disease entity accounting for less than 1% of all cervix cancer, particularly in the adult population, it's a malignant neoplasm that originates from undifferentiated myogenic progenitor cells, histologically subdivided into the embryonal, alveolar, spindle cell/sclerosing and pleomorphic types.

While embryonal is the most common histology seen, it commonly presents in pre-pubertal women or reproductive-age women in the second and third decades of life.

Given the rarity of the disease and its histologic variants, standard treatment guidelines are not only scarce but difficult to devise, the treatment is based on the association of surgery and pre or post-operative chemotherapy.

We report a case of a 62-year-old woman diagnosed with cervical embryonal rhabdomyosarcoma.

Case Report

We describe the case of a 62-year-old woman who presented with an eight-month history of abdominal pain and post-menopausal bleeding, a gynecological examination revealed the presence of a grape-like cluster cervical mass.

The diagnosis of RMS was confirmed histologically and supplemented by pelvic MRI, showing a locally advanced neoplastic process in the cervix with regional lymph node extension.

Patient history

A 62-year-old Moroccan woman, without a previous medical history, multiparous [4], presented with an 8-month history of abdominal pain and irregular post-menopausal bleeding.

Clinical finding

Pelvic examination demonstrated a 7 cm multi-lobulated mass with a 'grape-like' appearance, smooth, and focally hemorrhagic, the Rectal exam found a renitent mass protruding in the anterior face of the rectum, and the rest of the bodily examination was normal.

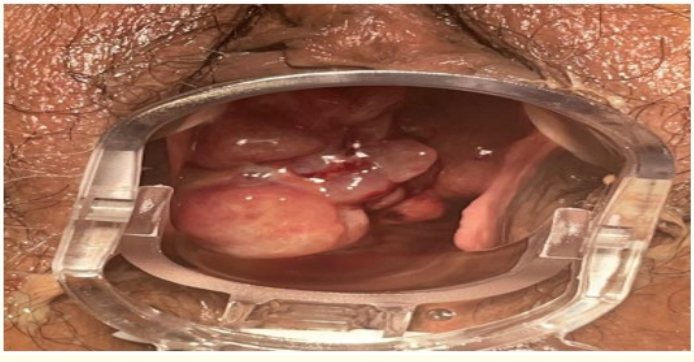

Figure 1: *Speculum exam shows a cervical multi-lobulated mass with a grape-like appearance.*

Laboratory tests revealed anemia with a hemoglobin level of 9 g/dL (normal range: 11.9 to 15.1 g/dL).

The biopsied tissue, counterstained with hematoxylin and eosin, showed infiltration of the endocervical mucosa by poorly differentiated, round-nucleated, spindle-shaped cells within myxoid fibrous tissue, indicative of rhabdomyoblastic differentiation.

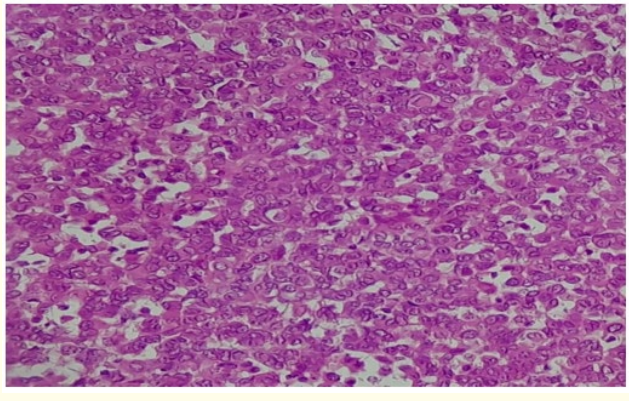

Figure 2: *Infiltration of the endocervical mucosa by poorly differentiated round-nucleated spindle-shaped morphology in a myxoid fibrous tissue related to rhabdomyoblastic differentiation.*

Immunohistochemical analysis revealed that tumor cells were immunoreactive to desmin. Nuclear staining for myogenin. However, the tumor cells were not immunoreactive to cytokeratin. These findings are consistent with an embryonal RMS.

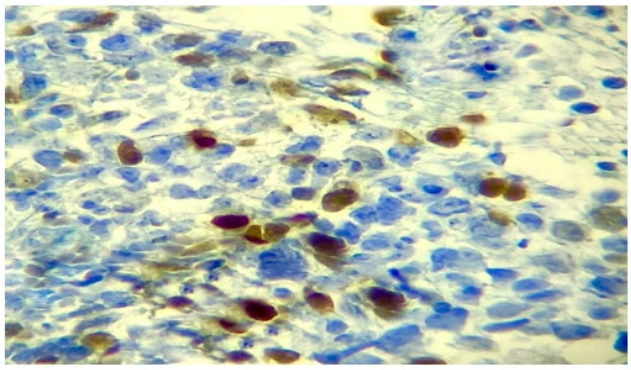

Figure 3: *Tumor cells express myogenin antibody.*

A pelvic MRI is performed to establish a loco-regional extension assessment, showing a voluminous process at the cervix, with lobulated contours, heterogeneous T2 high signal intensity, high signal intensity on diffusion, strongly enhanced after injection of gadolinium, extending into the upper two-thirds of the vagina, invading the endometrium at the top, and then respect the parametrium laterally.

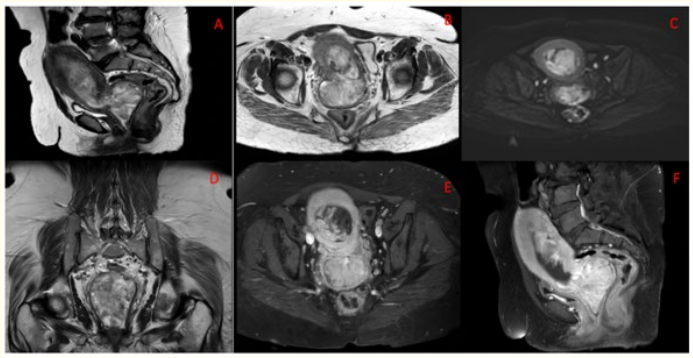

Figure 4: *MRI of the pelvis sagittal T2 (A), axial T2WI (B), diffusion (C), coronal T2WI (D), axial and sagittal T1WI after gadolinium injection (E, F), showing a tumor in the cervical region with endometrial and vaginal protruding.*

Therapeutic intervention

Surgical treatment was proposed to the patient, the operation consisted of a total open hysterectomy with adnexectomy.

An initial assessment of the abdominal cavity was carried out, showing no peritoneal carcinosis or extrauterine tumor location, which confirmed the decision to abstain from pelvic lymph node dissection.

Histological examination of the surgical specimen confirmed the initial diagnosis with untouched parameters.

Upon follow-up, the case was discussed in a multidisciplinary gynecological oncology consultation meeting to consider adjuvant chemotherapy.

Discussion

Rhabdomyosarcoma (RMS) is a malignant mesenchymal tumor arising from the embryonal muscle cells, commonly diagnosed in the pediatric population [1], its occurrence in adults is exceedingly rare, it mainly affects the cerebral and cervical region, followed by the genitourinary tract, the vagina is the most common site of the genital tract rhabdomyosarcoma (RMS), only 0.4 - 1% of primary are found on the cervix, generally found in the second and third decade of life [2]. It is exceptionally uncommon in post-menopausal patients, however, only a few cases have been published in patients over the age of 50 [3].

Most patients present the appearance of recurrent metrorrhagia associated or not with leucorrhoea, abdominal and lower pelvic pain, or pollakiuria [4,5].

The examination shows an appearance that depends essentially on the stage of discovery, a benign polyp relapsed after excision, which may cause a delay in diagnosis [6] or a classic polyploid "grape-like" mass in advanced phases [7], as is the case in our patient.

The immunohistochemical profile of the tissue is nowadays a must in diagnosing sarcomas, there are four subtypes of rhabdomyosarcoma: embryonal, alveolar, spindle cell/sclerosing and pleomorphic [8], 58% of all rhabdomyosarcomas are of the embryonal subtype [9].

Histologically, the embryonal subtype is characterized by the appearance of clusters of spindle and round pleomorphic cells with hyperchromatic nuclei, rhabdomyoblasts, and a myxoid stroma [8,10,11].

Rhabdomyosarcoma is an aggressive tumor with massive locoregional extension, rare metastasis, and frequent recurrences [12], conventional radiology is very useful in the assessment of the primary tumor, delineating the tumor's extension to adjacent organs, and identifying metastasis [13], to establish an adequate therapeutic strategy.

The pelvic MRI is the gold standard for detecting locoregional extension, RMS has a low to intermediate signal intensity on T1-weighted images and an intermediate to high signal intensity on T2-weighted images, the high T2 signal is secondary to excessive myxoid stroma creating a multicentric cystic appearance, the mass showed heterogeneous enhancement on post gadolinium sequences, positron emission tomography-Computed tomography (PET-CT) and CT are used to search for metastases, which will condition the therapeutic attitude [14].

Since cervical sarcomas is a rare entity and histologically diverse, standardized management guidelines for the disease are lacking, according to an expert consensus from the Children's Oncology Group for treating RMS of the cervix, uterine small, localized tumors, without metastasis preferably should be treated with local fertility-sparing treatment based on only partial surgical excision of the lesion, with optional adjuvant chemotherapy [15].

The most important principle for surgical treatment in RMS is the complete resection of the primary tumor with the surrounding margin of normal tissue. Most of the ERMS (or all RMS) of the genitourinary tract in adults is diagnosed at an advanced stage, with widespread disease and metastasis to the regional and distal lymphatic nodes.

In such cases, an aggressive multimodal treatment combines radiotherapy and chemotherapy with a total hysterectomy with regional lymphatic node resection [16].

The prognosis of patients with rhabdomyosarcoma depends on the age, the histologic subtype, size, site of origin, disease extent at presentation, and residual disease after treatment.

The international rhabdomyosarcoma study group's staging criteria include tumor size greater or less than 5 cm, extension to surrounding tissue, lymph node, and distant metastasis. In cases of early-stage disease, the embryonal subtype, younger age, and presence of exophytic features collectively suggest a more promising prognosis [17].

Conclusion

Embryonal rhabdomyosarcoma of the cervix in a menopausal patient is an uncommon disease presenting with vaginal bleeding or a protruding mass. Accurate diagnosis can be established by performing appropriate immunohistochemical stains.

Pelvic MRI or CT scans are highly valuable for evaluating the primary tumor, mapping its proximity to neighboring organs, and detecting metastases.

Due to the rarity of rhabdomyosarcoma in older woman, firm management guidelines are non-existent, they are devised from experience with comparable cases having analogous tumor pathology.

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