

Case report

Cervical rhabdomyosarcoma in an endocervical polyp of a 50 year old patient with intermenstrual bleeding

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1. Introduction

Rhabdomyosarcoma is a rare mesenchymal tumor that is usually diagnosed in children or young adults. It accounts for less than 1% of overall adult cancers and 2–5% of all soft tissue sarcomas (Shim et al., 2011). Although it is most commonly diagnosed in the head and neck, it can be seen arising in the genitourinary system, most commonly, the vagina (Sanders et al., 2008). It appears to have worse prognosis when diagnosed in adults, but this may be due to a lack of standardized treatment protocols for this population as recommendations are often based on studies in the pediatric population (Narin et al., 2016).

2. Case summary

A 50 year old Afro-Caribbean premenopausal female presented to her gynecologist with complaints of malodorous vaginal discharge, intermenstrual bleeding, and passing fleshy tissue per vagina for one month. She had no significant past medical nor family history, and denied loss of appetite, bloating, abdominal pain, or weight loss. On physical exam her cervix appeared grossly normal and the uterus was mobile and enlarged to 16 weeks size with palpable fibroids. During her workup, an endocervical polyp was visualized on hysteroscopy. Pathology review of endometrial curettings from a dilation and curettage (D&C) showed polypoid fragments of the endometrium, myometrium and cervical tissue with foci of primitive rhabdoid differentiation, suspicious for rhabdomyosarcoma. Immunohistochemistry of the primitive cells was positive for p53, myogenin, desmin, and SALL4 with high Ki-67 proliferation index of 40–80%.

She subsequently underwent surgical staging including a hysterectomy, bilateral salpingo-oophorectomy, and pelvic and para-aortic lymph node dissection. Final pathology confirmed embryonal rhabdomyosarcoma arising in a 1.5 cm polyp in the anterior endocervix. The tumor was composed of nodules of hyaline cartilage, small blue cells in sheaths, and cells with rhabdoid morphology (Fig. 1). The endometrium, ovaries, fallopian tubes, and ovaries were unremarkable. However, there was focal lymphovascular space and perineural invasion present. Immunohistochemistry of the final tumor specimen was

positive for desmin (100%), myogenin (40–50%) and CD56 (100%) with high Ki-67 proliferation index of 50–60% (Fig. 2). It was negative for AE1/AE3, synaptophysin, chromogranin, TTF-1 and CD10.

Systemic therapy with Vincristine, Dactinomycin and Cyclophosphamide (VAC) for forty weeks was recommended based on the data published by the Children's Oncology Group (COG) (Hawkins et al., 2018). Based on this data, concurrent external beam radiation to 5040 cGy was given concurrently during weeks 6–10. The patient finished her treatment without any delays. She experienced grade 1 diarrhea and constipation that resolved with changes in diet and grade 1 mucositis that resolved with Nystatin oral suspension. Computed tomography scans of the chest, abdomen, and pelvis after completion of chemotherapy showed no evidence of disease. Six months after finishing treatment the patient is without any signs or symptoms of recurrence.

3. Discussion

We present the case of a 50 year old with cervical rhabdomyosarcoma diagnosed in an endocervical polyp. This case highlights the difference in presentation between our patient and the pediatric population. In younger patients the tumor most commonly presents as a cervical mass or polyp, often described as “bunch of grapes” (Hosseini et al., 2016). However, visual inspection of the cervix in our patient revealed no abnormalities. The diagnosis was only suspected after D&C and then confirmed on final pathology specimen after hysterectomy was performed. As this diagnosis is rare in the adult population, immunohistochemistry stains can aid in diagnosis. In a pathologic review of 1052 cases from COG, myogenin was the most specific and sensitive marker for rhabdomyosarcoma [Morotti et al., 2006]. In the same publication, alveolar rhabdomyosarcoma stained diffusely for myogenin, embryonal rhabdomyosarcoma stained only focally, while other sarcomas were negative for the marker.

A small number of cervical rhabdomyosarcomas in adults have been reported in the available literature, and treatment recommendations are not uniform and are often based on data published from the COG (Narin et al., 2016). Studies published by this group are conducted in

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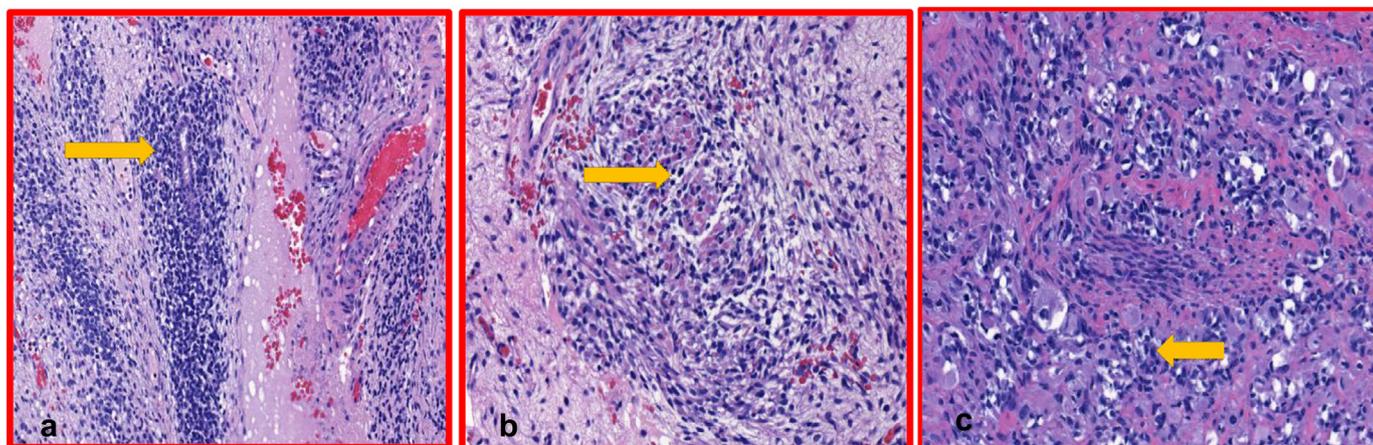


Fig 1. The tumor is composed of nodules of hyaline cartilage and (a) cambium layer around entrapped cervical glands. There are areas of small blue cells in sheaths representing rhabdomyoblasts (b) and other areas showing cells with rhabdoid morphology (c). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

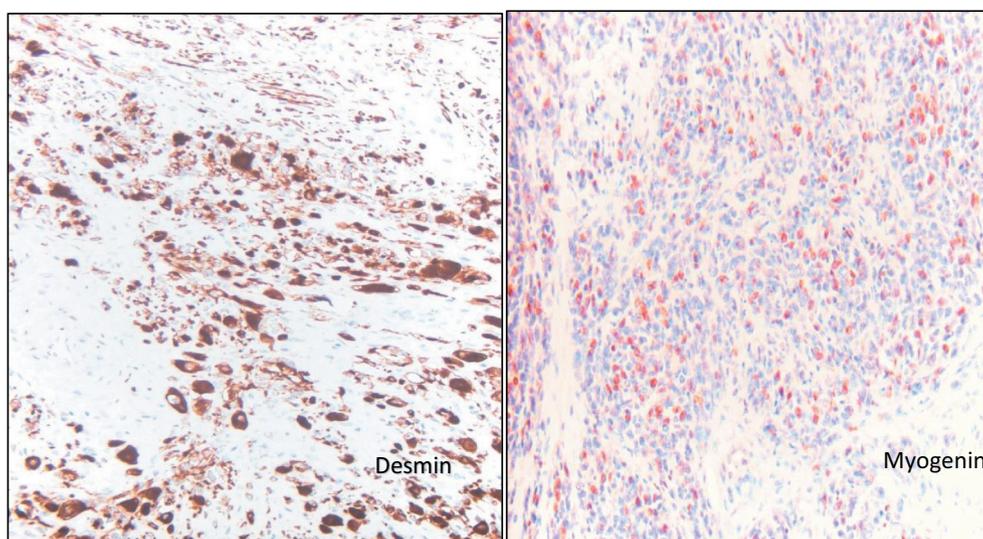


Fig. 2. Immunohistochemistry slides showing tumor cells staining positive for desmin and myogenin.

the pediatric population and once diagnosed, patients are designated as having low, intermediate or high risk disease. In general, patients with local disease that is completely resected are considered low risk while patients with local disease and microscopic residual after resection are considered intermediate risk. Patients in the high risk category have gross residual disease after biopsy or debulking of the tumor. Treatment recommendations are based on stage, site of disease and residual tumor in cases where surgery is performed (Bradley et al., n.d.).

Treatment modalities for cervical rhabdomyosarcoma range from fertility sparing surgery to radical hysterectomy with or without adjuvant chemotherapy and radiation. In the pediatric population, standard treatment is considered chemotherapy and preservation of fertility. In adults, surgery has proven beneficial and intensive chemotherapy tolerance is more of a concern. Our adjuvant treatment recommendations were based on the COG guidelines. Based on results of study ARST0531, this group recommends chemotherapy with VAC for 40 weeks and radiation therapy concurrently that may start on week 4 of treatment (Hawkins et al., 2018). In the aforementioned study, the use of vincristine and irinotecan (VI) in half of the cycles did not improve patient outcome.

The prognosis of rhabdomyosarcoma in both children and adults varies with stage, location, and histology. Outcomes in the pediatric population have improved over time with the addition of chemotherapy

to treatment. Systemic therapy has also allowed for less radical surgery and preservation of fertility in this patient population. Treatment modalities for rhabdomyosarcoma in adults still vary widely, in part due to lack of standard guidelines. A SEER database study showed that the five year overall survival in adults with rhabdomyosarcoma was 27% as compared to 61% in children (Sultan et al., 2009). Analysis of the same data showed that adults with rhabdomyosarcoma were less likely to receive chemotherapy than children with the same diagnosis. Another series of 171 adults with rhabdomyosarcoma showed that five year overall survival was 40%, however this increased to 61% for patients receiving treatment based on updated pediatric guidelines (Ferrari et al., 2003).

This case highlights the importance of a thorough investigation as adult patients with rhabdomyosarcoma might not present with the typical cervical mass that is often described in the pediatric population. Also, more data on treatment and survival of adults with cervical rhabdomyosarcoma are needed.

Conflict of interest statement

No authors have any conflict of interest to declare.

Author contribution

Blerina Salman: clinical evaluation, data gathering, paper writing.
 Amir Momeni-Boroujeni: pathology analysis and paper writing.
 Margaux Kanis: paper writing and analysis.
 Yi-Chun Lee: paper editing.

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