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VAGINAL YOLK SAC TUMOR IN AN INFANT-MANAGEMENT DILEMMA.

Oncology

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ABSTRACT

Malignant Germ Cell Tumors are extremely rare in pediatric age group. Yolk sac tumor of vagina is a highly malignant, rare form of GCT and is seen exclusively in children less than 3 years. There are no treatment guidelines for this malignancy as the literature is limited to few case reports. Treatment options are primary chemotherapy, induction chemotherapy with conservative surgery or radical surgery. A 12 month old female infant was referred to our center for intermittent vaginal bleeding. The clinical examination revealed a grape like material per vaginally. She had elevated serum AFP levels (7781ng/ml), and MRI pelvis suggested mass lesion involving uterus and upper vagina. Histopathological examination suggestive of yolk sac tumor, confirmed by IHC (PLAP- strong positive, AFP, AE1, B-HCG positive). Patient was treated with chemotherapy (JEB). There was complete clinical and radiological response and decreased serum AFP levels. After DFS of 4 months patient developed recurrent lesion and had raised serum AFP levels. As parents didn't consent for surgery she was started on second line chemotherapy (VeIP). The size of recurrent lesion and serum AFP levels decreased but patient developed lung metastasis.

KEYWORDS

infantile malignancy, germ cell tumor

CASE REPORT:

A 12 month old female infant with no other congenital anomalies presented with complaints of painless intermittent vaginal bleeding for 25 days. Per abdominal examination was normal and per vaginally grape like tissue was found which was sent for biopsy. MRI pelvis showed 3.5×5.6 cm mass lesion involving uterus and upper vagina, homogenously hypointense on T1W1, hyperintense on T2/STIR with flow voids on T2W1. There was no distant metastasis at the time of presentation. Lesion was indenting anteriorly on urinary bladder with focal capsular breach at left lateral part of uterus which was abutting to left pelvic side wall. Amongst the tumor markers S.AFP was 7781ng/ml. Biopsy was suggestive of Yolk Sac tumor which was confirmed with IHC showing tumor cells which were strong positive for PLAP marker and focally positive for AFP,AE1 and B-HCG markers.

Patient was treated with 3 cycles of JEB protocol (Carboplatin 600mg/m^2, Etoposide 120mg/m2, Bleomycin 15units) every 21 days which she tolerated well. S.AFP level declined to 4.68ng/ml, with complete clinical and radiological response on follow up imaging done after 4 weeks. Patient was kept on close follow up with monthly S.AFP and abdominopelvic ultra-sonographic evaluation.

After remaining disease free for 4 months she again developed per vaginal hemorrhagic spotting with elevated S.AFP 311ng/ml. Follow up MRI showed 20×15 mm recurrent lesion in upper vagina. Patient was advised for radical surgery (total abdominal hysterectomy), which her parents deferred.

The patient was treated with 3 cycles of second line chemotherapy of VeIP protocol (Vinblastine, Ifosfamide, and Cisplatin). Patient responded well to chemotherapy with complete clinical response. Follow up MRI (after 4 weeks) showed reduced size of primary lesion (10×9 mm) with declined S.AFP level of 140ng/ml. Although patient remained clinically asymptomatic, follow up CECT scan done after 2 months, showed unchanged primary lesion in size with multiple subcentimetric bilateral lung metastatic deposits. Patient was advised to start extra cycles of VeIP chemotherapy as it has showed response.

DISCUSSION

Pediatric malignant germ cell tumors (GCT) are very rare cancers accounting for less than 3% of pediatric malignancy. Endodermal Sinus Tumor (EST) is the most common GCT seen in pediatric age group.[1,2] In children, the tumors are frequently located in the sacrococcygeal region, testis, and ovary, and in the areas adjacent to these locations, the thorax, the vagina, and other sites.[1,2] The extra gonadal location of YST could be the result of germ cells "lost" during their migration from the yolk sac along the dorsal wall of the embryo to the gonadal folds.[3] EST (Yolk Sac Tumor) of vagina is extremely rare tumor, reported exclusively in less than 3 years of age group worldwide.[1,2,4]

Yolk sac tumor of vagina often present with vaginal bleeding with occasion protrusion of polypoidal mass per vaginum. Vaginal examination under anesthesia is recommended. Sarcoma Botyroids- a variant of embryonal rhabdomyosarcoma is the most common clinical differential.[4]

Formerly yolk sac tumor of vagina was confused with clear cell carcinoma of vagina, but experienced histopathological examination showing characteristic Schiller-Duval bodies is diagnostic of Yolk sac tumor.[2,4,5] Some authors classify the histological patterns of yolk sac tumors as follows: festoon, reticular, solid, and polyvesicular, most tumors exhibit more than one of these patterns.[2]

The usefulness of elevated S.AFP as a tumor marker in diagnosing yolk sac tumor vagina is well established.[6] S.AFP is used for monitoring disease response and elevated S.AFP is also suggestive of tumor recurrence. [4,5]In our case, patient was diagnosed by histopathological examination, confirmed with IHC and elevated S.AFP tumor marker. During treatment S.AFP was used as a tumor marker to see disease response.

Current treatment strategy includes chemotherapy alone or chemotherapy with conservative surgery. Overall treatment has evolved over the last 3 decades from radical surgery to conservative surgery with chemotherapy to chemotherapy alone.[7] Anderson et al

reviewed the cases of yolk sac tumor of vagina and showed improved disease free survival at 2 years with primary chemotherapy.[8] Maligne Keimzelltumoren Study Group report showed survival benefit of platin based neoadjuvant chemotherapy with conservative surgery over radical surgery and suggested induction chemotherapy followed by conservative surgery and adjuvant chemotherapy as an effective approach.[9] Upfront surgery followed by chemotherapy is also discussed to have a role as a treatment strategy.[7] Recent case series by Yuan et al suggested nonsurgical treatment with chemotherapy until complete response followed by an additional cycles of consolidated treatment.[10] Our patient was treated with chemotherapy first which resulted in initial complete response, but later on she developed recurrent and later on metastatic disease; suggesting the role of conservative surgery to control primary disease. Because of paucity of literature definitive treatment protocol for vaginal yolk sac tumor is snot well defined.

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