Sarcoma Botryoid of Vagina and Cervix in a three year old girl

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Abstract

A three year old with three months history of abdominal lump, blood mixed vaginal discharge and occasional urinary retention was operated as a case of sarcoma botryoid of vagina and cervix. This was proven histologically. With surgery it was possible to remove a huge mass arising from cervix and vagina measuring 10X10 cm. It was almost mimicking central cervical myoma. Initial surgery was observed to be beneficial in this case as it showed immediate good results by providing reduction of total tumour mass. She was planned for combination chemotherapy with VAC; however she developed abdominopelvic recurrence very soon.

Keywords: young girl; vaginal growth; rhabdomyosarcoma; sarcomabytryoids.

Introduction

Sarcoma Botryoid is one of the rare tumours involving vagina in girls younger than 4 and at the same time is more virulent than its counterpart the cervix which is seen at the age of four-sixteen.[1] This rare malignancy has been described in a 5 months child where radical hysterectomy had been performed.[2] However in present day people have comeup with newer therapeutic options like combination chemotherapy, surgery or radiation in combination so as to revert to a simpler surgical procedure like total abdominal hysterectomy in an endevour for better 5 year survival rate also thereby reducing the surgical morbidity.

Case

A girl (IPN 288233) 3 years of age from Dhalkebar was admitted on 12th Nov 2000 with the history of blood stained discharge from a growth per vagina for the last three months. Earlier she was admitted in some other hospital due to the bleeding from the same growth which had just appeared for ten days. A biopsy was taken and she was discharged. Soon after there was urinary retention and was relieved by continuous catheterisation by some local doctors and was referred to our hospital finally. The biopsy report was not collected by the patient's party.

She looked pale (Hb 6.4 gm), abdominal examination revealed a firm nontender suprapubic mass approximately 12X10 cm. A bluish growth was seen protruding out of the introitus with evil discharge. An ultrasound of abdomen reported a mass in the lower part of abdomen measuring 8X5.8 cm with no ascitis.

On 23rd November 2000 (057.7.29) EUA was done and biopsy taken from the fragile projectile growth. The biopsy (S2373/057) however was inconclusive. Provisional diagnosis of botryoid sarcoma was kept because of strong resemblance of this mass to botryoid sarcoma described in text books which was also supported by CT abdomen pelvis, Cystogram and IVU showed the mass also producing hydroureter and hydronephrosis.

After 200 ml of B +ve blood transfusion, and improving Hb to 10.5 gm%; on Nov. 21st 2000 (057.8.6) abdomen was opened with low transverse incision. A small infantile uterus was seen sitting over a capsulated rounded mass of about 10 cm which on first impression looked like a central cervical myoma. The mass was continuous from the cervix involving vagina and projecting out of it.

At surgery, while separating the mass from the posterior part of bladder and urethra was injured. Hence a suprapubic catheter was kept.

The mass with uterus and ovaries were removed along with whole length of vagina. Cut section of the mass showed grape like projection and the histology (S2765/057) confirmed the diagnosis of botryoid sarcoma. Postoperatively she was covered with gentamycin, ampicloxacllin and metronidazole. Except of the left leg swelling which may be due to lymphatic obstruction, her recovery was good. She was discharged on Dec. 11th

2000 (26-8-057) with an advice for admission for chemotherapy in two weeks, however there was pelvic recurrence (enlarged matted lymph node) with mounting serum urea and creatinine with very little urinary output. The patient party refused for chemotherapy as they were uncertain of future prognosis.

Discussion

This is the first case of Sarcoma botryoids in this hospital however we had no difficulty in anticipating the diagnosis since it is so well described in the text.[1]

Histologicallly the tumour has a continuous condensed band of tumour cell below the surface epithelium with smaller pleomorphic tumour cells some having rhabdomyoblastic differentiation embedded in myxoid stroma. The dense cambium cell layer in the subepithelial location is characteristic of this malignancy.

Preoperative chemotherapy in form of VAC (Viincristine, actinomycin, cyclophos-phamide) has been reported to diminish the tumour volume and size to such an extent that radical surgery at the outset has been unnecessary. In 1970's multimodality therapy with radical surgery, radiotherapy and comination chemotherapy was the main treatment which has been more recently modified to local excision with or without chemotherapy thus preserving fertility.[3-8]

Fig. 1: Sarcoma botryoid, protruding from vagina.

As primary surgery followed by chemotherapy has been a favored option, we offered initial surgery in this particular case in consideration of this large tumour size which keeps every doubt possible whether this girl would have tolerated chemotherapy that would have been continued invariably for longer period. An inadequate chemotherapy would have been the consequence of non-compliance.

Seeing this little girl leave the hospital without a trace of evil discharge forced us to believe that surgery in the first place is by no question better in this circumstance which provided such an excellent immediate effect. At the same time we learnt primary recurrences in the pelvis has still to be feared of. There is 26% incidence of lymph node metastasis and pelvis is the most common site for primary recurrene.

Fig. 2: Per operative picture showing grape-like structure.

Conclusion

Botryoid Sarcoma is one of the virulent tumours involving vagina and cervix in young children. Equal success has been obtained with less radical surgery and adjuvant chemotherapy with or without radiotherapy.

The chemotherapy using VAC has received lot of attention however local excision of growth possible only in the initial stage of disease can be combined with chemotherapy for better 5 year survival as well as preservation of future fertility.

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