A Successful Pregnancy Outcome in Treated Vulval Rhabdomyosarcoma

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Summary

A 14 year old girl presented in 1986 with a huge perineal swelling which was progressively increasing in size and associated with loss of weight and loss of appetite. Biopsy from the mass revealed rhabdomyosarcoma of the vulva. She was treated with chemotherapy and radium implant. She responded well to the regime. Fibrosis of the vulva and vagina caused difficulty in consummation. Once it was corrected, she conceived easily and proceeded to a normal pregnancy and delivery.

Key Words: Vulva rhabdomyosarcoma, Chemotherapy, Pregnancy

Introduction

Cases of vulva rhabdomyosarcoma responding well to chemotherapy and proceeding to a normal pregnancy and outcome is rare and so far there has been no cases reported in the local literature.

A Case Report

This is a case report of a patient who had vulva rhabdomyosarcoma presenting in childhood and who had successful pregnancy after completing chemotherapy and radiotherapy.

The 14 year old girl presented in October 1986 with a perineal swelling which progressively increased in size over 10 months. She also had a history of loss of weight and appetite.

Examination under anaesthesia revealed a huge irregular solid mass on the left vulva covering the urethral meatus, the introitus and extending to the mons pubis. Dilated veins were present on the surface with a necrotic ulcer on the anterior aspect, which was bleeding. The vaginal epithelium was intact and the inguinal lymph nodes were palpable bilaterally.

A biopsy was performed, histopathology of which showed a cellular mass of spindle and oval shaped cells arranged in streams (Fig. 1). Mitoses were present with numerous pyknotic nuclei. Large areas of haemorrhage and necrosis were present. No definite cellular differentiation was evident. Immunohistochemical stain was positive for vimentin but negative for desmin, actin and myoglobin. Therefore the diagnosis of undifferentiated sarcoma of the vulva with possibilities of rhabdomyosarcoma, leiomyosarcoma and fibrosarcoma was given.

Biopsy from lymph nodes showed reactive changes. The diagnosis was undifferentiated sarcoma of the vulva with possibilities of rhabdomyosarcoma, leiomyosarcoma and fibrosarcoma.
Fig. 1 Cellular tumour displayed spindle and oval shaped cells with numerous mitoses (x400)

She was given 10 courses of chemotherapy consisting of cisplatinum, bleomycin and methotrexate. Chemotherapy was then followed by external beam radiation and radium implant. The tumour shrunk. On the follow up in 1993, there was no evidence of recurrence. In 1995, after 18 months of marriage, she complained of non consummation as she had a fibrosed vulva and a stenosed vaginal introitus. Vaginoplasty was offered to release the fibrosis and the introitus widened. The procedure was repeated. Daily use of the vaginal dilators was advised. She was able to have satisfactory sexual intercourse one month later. She became pregnant 3 months later. Her LNMP then was the 1st January 1997. A detailed ultrasound done at 20 weeks showed a grossly normal fetus.

She was delivered by Caesarean section at 38 weeks to a female weighing 2.7kg. Intraoperatively, the ovaries, tubes and uterus were normal. She was discharged on the fourth post-operative day and remained well at follow-up.

Discussion

Rhabdomyosarcoma is the most common soft tissue sarcoma of children. Over the years its management had shifted from extensive mutilating surgery to a more organ conserving surgery, multi-agent chemotherapy as well as local radiation. The latter treatment reduces morbidity and assures a better quality of life as well as retaining fertility. However, to date very few patients having successful pregnancy outcome had been reported in the local literature.

Apart from our patient, another similar case was reported by Nasir J in February 1997. In this instance, a 14 years old girl with rhabdomyosarcoma of the arm was treated with cyclophosphamide, cisplatin and vincristine. She later developed chemotherapy-induced ovarian failure and was started on cyclical HRT. Seven years later, she had amenorrhoea which was associated with an 18 weeks pregnancy. Her pregnancy progressed well and a healthy baby was delivered.

Chemotherapy and radiation had been shown to reduce fertility in children treated for cancer. It had also been shown to increase the risk of adverse pregnancy outcomes. However, offsprings of these patients have little risk of childhood cancer and birth defects.

In our patient, her fertility status did not seem to be affected as chemotherapy was completed at 15 years of age and she then had regular menses. Local radiation given as treatment had caused extensive fibrosis and difficulty in sexual intercourse. Once this was corrected she was able to conceive almost immediately. Her pregnancy progressed uneventfully. She was delivered by a Caesarean-section due to her previous vaginoplasty.

This case illustrates that vulva rhabdomyosarcoma in a young patient could be treated fully without jeopardising fertility. In this patient pregnancy was spontaneously conceived but the vaginal post-radiation fibrosis necessitated a caesarean section for delivery. Both the mother and daughter were healthy. In this instance, the patient was counselled to expect a good outcome of pregnancy and was not discouraged from conceiving.
References

