

มะเร็งกล้ามเนื้อลายของอวัยวะสืบพันธุ์ภายนอกสตรี (รายงานผู้ป่วย)

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รายงานผู้ป่วยหญิงอายุ 13 ปี เป็นมะเร็งกล้ามเนื้อลายชนิดโบทริออยคอลล เกิดขึ้นที่อวัยวะสืบพันธุ์ภายนอก โดยมีประวัติมีก้อนที่ด้านซ้ายของอวัยวะสืบพันธุ์ ภายนอกเป็นเวลา 1 ปีก่อนรับไว้ ได้ให้การรักษาทดด้วยการผ่าตัดเอาก้อนนี้ออก กับอวัยวะสืบพันธุ์ภายนอกและต่อมน้ำเหลืองของบริเวณขาหนีบกับต่อมน้ำเหลือง ในช่องเชิงกรานออกทั้งหมด และให้การรักษาทดด้วย melphalan 2 เดือนหลัง ผ่าตัดเริ่มมีอาการแทรกซ้อนของมะเร็งเกิดขึ้น และถึงแก่กรรม 11 เดือนหลังจากการวินิจฉัยโรคได้

Rhabdomyosarcoma may be histologically divided into four categories: pleomorphic, alveolar, embryonal and botryoidal types.⁽⁸⁾ Botryoidal rhabdomyosarcoma is the only rhabdomyosarcoma which occurs predominantly in infants and children, and is rarely found in the vagina.^(2,9) It is even rarer to

find such a lesion at the vulva.^(8,5) Only 30 cases of sarcoma of the vulva had previously been reported.⁽⁶⁾

A case of botryoidal rhabdomyosarcoma of the vulva in a Thai girl is being reported since it is the only such lesion found in 79,490 specimens over the last fifteen years (1965 to 1979)

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Case Report

A Thai girl, single, aged 13 was admitted to the Department of Obstetrics and Gynaecology, Chulalongkorn Hospital on the 26th March, 1979 because of a tender mass at the left vulva. She did notice the mass about the size of her thumb a year prior to admission. This mass gradually enlarged and recently got so big that her walking was difficult. A week before her admission, she had pain and tenderness over the mass with the presence of discharge, resulting in a limping gait.

She had her menarche at the age of twelve and her periods had been regular, occurring once every 30 days and lasting 3 to 5 days. Her last menstrual period was on the 5th February, 1979.

On examination she was apparently well developed and in a good health apart from the enlarged left inguinal lymph nodes of 3 centimetres in diameter and a nodular, ulcerated and fairly highly vascularized tumour mass of 8×12 centimetres on the left side of the vulva. The mass with purulent discharge from the ulcer was tender and inflamed. Vaginal and rectovaginal examinations revealed essentially normal findings apart from the mass which was confined mostly in the left labium which extended upward to the mons and downward to the perineum and was partly up to the lower part of the vaginal wall. (Figure 1) The

vaginal epithelium was free from tumour invasion but the left vaginal wall was displaced to the right by pressure from the tumous mass.

As histological examination of the tumour biopsy revealed botryoidal rhabdomyosarcoma of the vulva, it was thought that radical vulvectomy followed by combined chemotherapy would be the appropriate management. The operation was then performed and the wound required skin graft. A month after her fairly satisfactory recovery from radical vulvectomy, she had four consecutive monthly courses of alkylating agent (melphalan at the dosage of 1 milligram per kilogram of body weight). Despite melphalan therapy, she had a rather fixed mass of 5 centimetres in diameter at the back of the vagina extending up to the level of sacrum in two months after surgical treatment. Radiotherapy was not given as her general health did not permit. Five months later she had mucous bloody stool and enlarged right submandibular lymph nodes and eventually developed sigmoid colon obstruction and fistula which required palliative colostomy. She finally expired 11 months after the diagnosis was made.

Pathological report

The radical vulvectomy specimen is consisted of a large mass of 6×7×11 centimetres occupying most of the left labium majorum, perineum and is partly in the lower vaginal wall. There are few enlarged left inguinal lymph nodes of 3 to 4 centimetres in diameter. The skin overlying the mass is ulcerated and



Figure : I

Photograph of the left vulva tumour demonstrates the huge, nodular, infected and ulcerated mass.

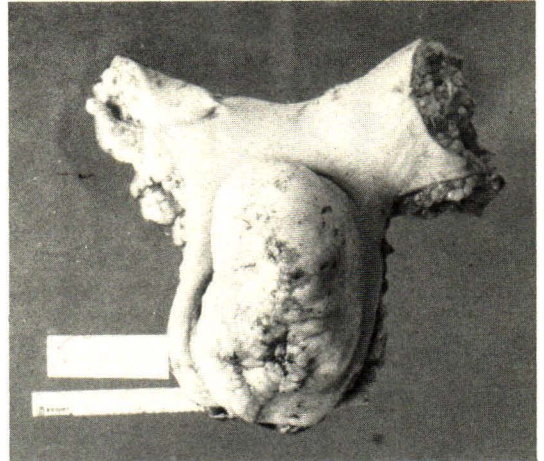


Figure : 2

Photograph of the radical vulvectomy specimen. Note the mass with bilateral inguinal lymph nodes and subcutaneous tissue en bloc.



Figure : 3

Photograph of the bisected tumour shows yellowish-white firm tumour with areas of necrosis in the subcutaneous tissue.

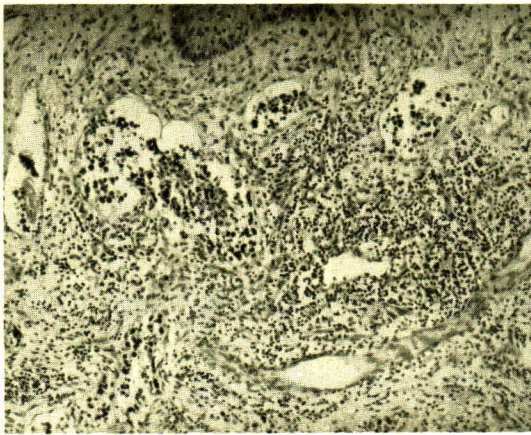


Figure : 4

Photomicrograph of the tumour shows malignant cells in the subepithelial zone. (H. and E. X 100)

Figure : 5

Photomicrograph of the tumour shows a tad-pole cell in the centre of the picture surrounded by elongated spindle and round cells with large hyperchromatic nuclei (H. and E. X 400)

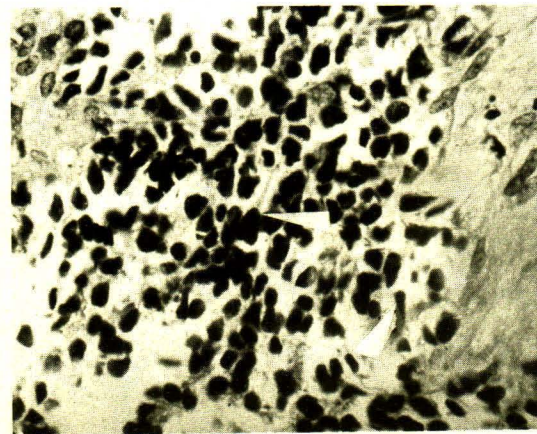
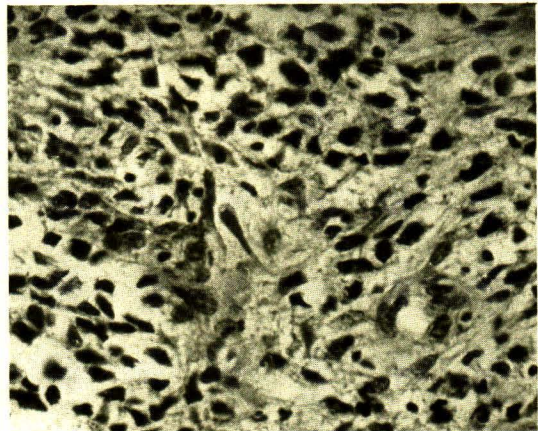


Figure : 6

Photomicrograph of the tumour, again shows large hyperchromatic nuclei with scanty acidophilic cytoplasm of spindle and round cells and tad pole cells (marker) (H. and E. X 400)

inflamed. Scattered small white nodules are noted on normal skin (Figure 2). Sectioning reveals an illdefined yellowish white firm tumour situating mainly in the subcutaneous tissue. Areas of necrosis are also present. (Figure 3)

On microscopic examination, sections of the vulva show a highly cellular and pleomorphic tumour of the sub-epithelial zone. (Figure 4) The tumour cells are mostly admixture of elongated spindle and round cells with some tad-pole cells. (Figure 5) These cells contain hyperchromatic nuclei with scanty acidophilic cytoplasm. (Figure 6) The subepithelial zone is markedly edematous. There are tumour thrombi in many small blood vessels. The skin is partly infiltrated by tumour cells leading to ulceration of the epidermis. Sections of inguinal lymph nodes also show metastatic tumour which is similar to the main tumour.

Discussion

Generally, malignant lesions of the vulva are diagnosed late as patients often delay in coming to see physicians. This patient is no exception and she came into the hospital after having severe pain and tenderness of the previously slow growing tumour starting from the size of her thumb but became huge and was infected on admission. Early diagnosis of vulvar malignancy in childhood may be possibly dealt with by public education, particularly to their parents and early consultation to physicians. Clinically the site and size of this huge, nodular, highly vascularized

tumour at the vulva with inguinal lymph nodes involvement are all evidences suggesting a malignant lesion although enlargement of the left inguinal lymph nodes may be resulted from the inflamed ulcerated mass at the left vulva. The severity of this malignant tumour may be clinically divided into four stages, i.e.. stage I: The tumour is confined to its organ of origin, Stage II: The lesion locally and proliferatively invades a structure adjacent to the organ of origin, Stage III: There is involvement of the regional lymph nodes and stage IV is that of distant metastasis.⁽¹¹⁾ The diagnosis of stage III rhabdomyosarcoma of the vulva in this patient was clinically and histologically^(10,11) made without delay. The infected ulcer on the mass was promptly and appropriately treated prior to operation. Surgical removal of the mass was mandatory in this patient, as it was inflamed and huge, making it difficult for her to walk. Pelvic exenteration has such psychological impact and limits the quality of life postoperatively that surgeons should take these points into judicious consideration before making decision and should discuss the problem with the parents prior to the operation

Radical vulvectomy followed by combined chemotherapy was thought at the time to be the appropriate treatment as opposed to simple excision of the mass and combined chemotherapy. The surgical intervention was performed with wide excision of the skin and total removal

of inguinal and all pelvic lymph nodes (Figure 2). The radical vulvectomy wound required skin graft and she made a fairly satisfactory recovery. Unfortunately, for this particular patient, the combined chemotherapy of choice with Vincristine, Actinomycin D and Cyclophosphamide (VAC) was not given as originally planned, due to unforeseen circumstances.

Radiotherapy was used by Kumar et al 1976 in combination with surgery and combined chemotherapy in two children and one infant with good result.⁽⁷⁾ However, radiotherapy was unable to be given to this patient as she had bone marrow depression at the time.

Ortega J.A. (1979) reported a therapeutic approach to childhood pelvic rhabdomyosarcoma with initial combined chemotherapy and less surgical intervention without changing the survival rate.⁽¹¹⁾ The use of combined chemotherapy (VAC) to rhabdomyosarcoma of the vagina or of the vulva has been reported with good outcome.^(4,12,13) The

combined chemotherapy in Ortega's report was consisted of vincristine (1.5 mg/m²) on day 1,8, actinomycin D (20 µgm/kg on day 1,2,3,4, and cyclophosphamide (10 mg/kg) on day 1,4,8. These were given intravenously and repeated every four weeks, whenever bone marrow recovery permitted. The total period was 24 months. This intensive combined chemotherapy, though not always available in the developing country as happened to the reported patient, appears to be one of the promising means to achieve cure for this very rare and fatal rhabdomyosarcoma of the vulva in childhood.

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