

A Case Report on Malignant Rhabdoid Round Cell Tumor in Pelvis

ABSTRACT

The malignant neoplasm is called as malignant rhabdoid tumor is also known as renal tumor. Malignant tumor had highest rate of proliferation. Tiny, round, and generally undifferentiated cells make up malignant small round cell tumours. The round cell tumor is a group of malignant tumors composed of relatively small and undifferentiated cells with increased nuclear - cytoplasmic ratio. Abdominal and pelvic soft tissue malignant tumor are rare but significant type of cancers. Ewing's sarcoma, peripheral neuroectodermal tumour, rhabdomyosarcoma, synovial sarcoma, non-lymphoma, Hodgkin's retinoblastoma, neuroblastoma, and hepatoblastoma are all examples of these cancers. Mast cell tumour, histiocytoma, lymphoma, plasmacytoma, and transmissible venereal tumours are some of the different types of round cell tumours. Melanomas are the cytologic "great impostor," as they might look on cytology as round cell tumours despite being classed as mesenchymal cancers. Rhabdoid tumours have long been thought to be extremely malignant and have a bad prognosis. Children with this form of tumour have a six to eleven-month median survival duration. They're also less common. In 5% of small round cell tumor patients, can curable and it is best achieved by combining systemic chemotherapy with thorough cytoreductive surgery. Here we report a 9-year-old female child who was diagnosed with malignant rhabdoid round cell tumor in pelvis. She undergone excision of pelvic floor tumor and further managed with Chemotherapy.

Key words: Malignant rhabdoid tumor, Pelvis, Round cell tumor, Tumor.

1. INTRODUCTION

A tumour (sometimes called a neoplasm) is a growth of abnormal cells in the body. Cells that divide too quickly or do not die when they should cause this illness. Tumors can be divided into two types.

- Benign
- Malignant

Malignant tumours are characterised by uncontrollable cell growth that can spread locally and/or to distant areas. Malignant spreads from the periphery, sending invading processes outward. Malignant growth was faster and increased at a variable rate. This illness causes anaemia, weakness, and weight loss, among other things. The tumour grows too large for the blood supply to keep up, causing substantial tissue damage. A malignant tumour is the cause of death. Tumors that are malignant are cancerous tumours (ie, they invade other sites). They spread through the bloodstream or lymphatic system to distant places. The medical word for this form of spread is metastasis. Metastasis can develop in any part of the body, but it is most frequent in the lungs[1]. Malignant rhabdoid tumour, a highly malignant neoplasm, is best recognised as a kidney tumour,

however malignant rhabdoid tumour is also found in other organs, including the liver. Pelvic tumours are the most common type of tumour in the pelvis. A paranuclear eosinophilic inclusion body ultrastructurally consisting of cytoskeletal filaments and immunoreactive for cytokeratins and vimentin is frequently seen in malignant rhabdoid tumours, with medium-sized to large, so-called rhabdoid A paranuclear eosinophilic inclusion body, ultrastructurally comprised of cytoskeletal filaments and immunoreactive for cytokeratins, is frequently seen in cells[2]. Malignant tumours grow and spread to adjacent nerves and muscles, causing pain and soreness as a result. The best medical treatment for a pelvic tumour is surgery, which has the highest success rate. The annual incidence of renal tumours in children under the age of 15 is 0.19 per million, 0.89 per million for rhabdoid tumours, and 0.32 per million for cancers of other locations [3]. Diagnosis of pelvic examination from biopsy before referral for imaging. Oral and IV contrast material are routinely used in pelvic CT for the evaluation of malignancy. It's important to make sure that imaging reaches below the perineum to include the pelvic floor and groynesentirely. Factors affecting the survival of patients with malignant tumors are :

- Size of malignant tumor.
- Grade of tumor.
- Vascular invasion
- Lymph node involvement
- Cancer markers are tumour cell markers that differ depending on the type of cancer.

Treatment options for soft tissue malignant tumors are combined skills of an exceptional surgeon, pathologist, radiologist, radiotherapist, medical oncologist , and sometimes a plastic surgeon.

In 90% of cancer cases ,the goal of treatmet is removal of malignant tumor.

2.PRESENTATION OF CASE

A 9-year-oldfemale childpatient was admitted in a tertiary care hospital for the management malignant rhabdoid round cell tumor in pelvis. The subject is a known case of a stage 3 malignant tumor. She was prescribed with 4 cycles of chemotherapy and underwent treatment with Cyclophosphamide, Mesna, Doxorubicin, Dexamethasone, and Granisetron. And advised for pelvic surgery. She had medical and surgical history on non-rhabdomyosarcoma(RMS) soft tissue sarcoma of pelvis.

On general examination subject hadpallor .She was normal on systemic examination. The vital signsinclude blood pressure 98/50 mmHg and pulse rate 92 bpm . On laboratory investigation WBC was decreased $4.5 \times 10^9/L$, hematocrit was decreased 22.8% .

As surgical prophylaxis on day 1 subject was treated with tablet Co- trimoxazole 80/40 mg . On 2nd day she underwent surgical procedure After surgery she was treated with combination drug of injection piperacillin tazobactam 2.5 g , then injection Amikacin 225 mg. After post operative the child had tenderness over left intracapsular region with hyperpigmentation During admission subject was found to be anemic had Hb level

was 7.6 g/dL was treated with tablet Ferrous Fumarate 50 mg , tablet Folic acid 5 mg , syrup Ascorbic acid 5 ml. After 3 days of surgery. For further management the patient was advised with cycle of chemotherapy and discharged.

3.DISCUSSION

Rhabdoid tumor of the kidney is the most aggressive and deadly renal tumor in children , accounting for 2 % of all renal tumors [4]. Malignant tumors grows uncontrollably and spread locally. A malignant rhabdoid tumor in the pelvis must be recognised from a variety of other sarcomas that might affect this area . Despite the fact that cells with granular the most attention is given to the cytoplasm and filamentous inclusion. The majority of cells are devoid of cytoplasm [5].Desmoplastic small round-cell tumour (DSRCT)is an uncommon, aggressive malignant tumour that most commonly occurs in the abdominal-pelvic region[6]. In a study , malignant rhabdoid was tumor was diagnosed with 53 patients , from that 53 patients 32 cases was diagnosed with malignant rhabdoid tumor of the kidney and 21 cases was diagnosed with extrarenal extracranial rhabdoid tumor[7].Even though malignant tumour is usually resistant to a variety multimodality treatment, more rigorous and unique treatment options would be required in the future[8].Only 14 occurrences of extracranial malignant rhabdoid tumour (MRT) were reported in a research undertaken by Childrens Hospital Los Angeles between 1983 and 2003, with only one in the pelvis. Extrarenal extracranial rhabdoid tumour (EERT) is most common in children, with a reported frequency of 0.15 per million in children under the age of 15, and it is uncommon in adult[9].A polyphenotypic immunoprofile and ultrastructural diversity are essential features of Rhabdoid Tumor[11]

Malignant rhabdoid round cell tumor in pelvis are rare cancer occur in children. Rhabdoid tumours of the kidney in children are described by a mutation or deletion of the 22q11 gene. Adult rhabdoid tumours are an unusual neoplasia that have also been detected in a variety of anatomic regions, but their histogenesis is unexplained[10].

A malignant cell has accelerated cell cycle, genetic alterations, invasive growth, enhanced cell mobility, chemotaxis, changes in the cellular surface, production of lytic agents, and so on [12].

Pediatric patient with malignant rhabdoid tumor of the pelvic region present with pain in the pelvic area , frequent urination due to pressure on the bladder. Researchers said that about twenty percent of women experience a pelvic mass in their lifetime. Pelvic tumor is diagnosed with ultrasonography, abdominal /pelvic CT , MRI etc.

Pathogenesis of malignant rhabdoid tumor described as:

Malignant rhabdoid tumours lack the embryonal rhabdomyosarcoma's strap cells and extensive cytoplasmic spindling. Skeletal structure is deficient ultrastructurally muscle differentiation and lack of muscle staining. This differentiation is also aided by related antigens[5].

Paediatric neoplasms such as rhabdomyosarcoma and neuroblastoma are more common in children with neurofibromatosis type 1 than in children without the condition [13].

Classification of renal pelvic tumors as following: It includes four grade : Grade I includes papillary with normal mucosa (thickness and morphology) , grade II includes papillary with minimal pleomorphism , mitosis,

giant cells, and invasion , grade III includes flat , transitional cells (plant life) to epidermoid (keratin forming) with significant pleomorphism , mitosis , giant cells and invasion , grade IV extreme pleomorphism or marked undifferentiation. Actinomycin D, vincristine, cisplatin, cyclophosphamide, adiramycin, etoposide,[14] are among the chemotherapeutic drugs used to treat rhabdoid tumors, with no clear advantage of one over the other[15].

The case study focuses on a 9 -year- old female patient who had already been diagnosed with malignant rhabdoid tumor in pelvis. As a conservative management for the condition patient underwent pelvic floor tumor and advised four cycles of chemotherapy with cyclophosphamide, mesna, doxorubicin, dexamethasone and granisetron and discharged.

4.CONCLUSIONS

Here we concluded that the malignant rhabdoid tumor in pelvis is rare and occur mainly in females due to accumulation of menstrual blood for the reason of uterus become enlarged in a girl or young women. Malignant rhabdoid tumor was a renal tumor of infancy and childhood. Malignant rhabdoid tumor develops a pelvic mass and occur in adolescent. Rhabdoid tumour is a rare, fast-growing cancer that most commonly develops in the kidney or CNS (brain and spinal cord), although it can also develop in soft tissue in other parts of the body. Round cell tumor is highly aggressive malignant tumor made up of small and undifferentiated cells with increased nuclear cytoplasmic ratio.

CONSENT

Written informed consent was obtained from the patient.

ETHICAL APPROVAL

As per standards , the ethical approval was collected and preserved by the author.

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