A Case Report on Malignant Rhabdoid Round Cell Tumor in Pelvis

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Authors’ contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/JPRI/2021/v33i59A34287

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/80151

Received 10 October 2021
Accepted 14 December 2021
Published 16 December 2021

ABSTRACT

The malignant neoplasm is called as malignant rhabdoid tumor or renal tumor. The Malignant tumor had the highest rate of proliferation. Tiny, round, and generally undifferentiated cells make up malignant small round cell tumours. A round cell tumor is a group of malignant tumors composed of relatively small and undifferentiated cells with an increased nuclear - cytoplasmic ratio. Soft tissue malignant tumours of the abdomen and pelvis are a rare but serious kind of cancer. Examples of these tumours include Ewing's sarcoma, peripheral neuroectodermal tumour, rhabdomyosarcoma, synovial sarcoma, non-lymphoma, Hodgkin's retinoblastoma, neuroblastoma, and hepatoblastoma. 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 be 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 a malignant rhabdoid round cell tumor in the pelvis. She has undergone excision of pelvic floor tumor and further managed with Chemotherapy.

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Keywords: Malignant rhabdoid tumor; pelvis; metastasis; chemotherapy.

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 (i.e., 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 111tumour, 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 para nuclear 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. 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 treatment is the removal of the malignant tumor.

2. PRESENTATION OF CASE

A 9-year-old female child patient was admitted to a tertiary care hospital for the management malignant rhabdoid round cell tumor in the pelvis. The subject is a known case of a stage 3 malignant tumor. She was prescribed 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 of non-rhabdomyosarcoma (RMS) soft tissue sarcoma of the pelvis.

On general examination subject had pallor. She was normal on systemic examination. The vital signs include blood pressure 98/50 mmHg and pulse rate 92 bpm. On laboratory investigation WBC was decreased 4.5 x 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 the 2nd day she underwent a 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 the left intracapsular region with hyperpigmentation. During admission subject was found to be anemic had Hb level of about 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 the 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 grow uncontrollably and spread locally. A malignant rhabdoid tumor in the pelvis must be recognized from a variety of other sarcomas that might affect this area. All though cells with granularity the most attention are given to the cytoplasm and filamentous inclusion. The majority of cells are devoid of cytoplasm [5]. Desmoplastic small round-cell tumor (DSRCT) is an uncommon, aggressive malignant tumour that most commonly occurs in the abdominal-pelvic region [6]. In a study, malignant rhabdoid 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 extra renal extracranial rhabdoid tumor [7]. Even though malignant tumour is usually resistant to a variety of 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 a Children’s Hospital Los Angeles between 1983 and 2003, with only one in the pelvis. An 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 [10].

Malignant rhabdoid round cell tumor in the 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 unusual neoplasia that has also been detected in a variety of anatomic regions, but their histogenesis is unexplained [11].

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 patients with a 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. The 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. Muscle differentiation and staining are both lacking in the skeleton's ultrastructure. 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, etopside, [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. A conservative management for the condition patient underwent pelvic floor tumor and was advised four cycles of chemotherapy with cyclophosphamide, mesna, doxorubicin, dexamethasone, and granisetron and was discharged.

4. CONCLUSION

Here we concluded that the malignant rhabdoid tumor in the pelvis is rare and occurs mainly in females due to the accumulation of menstrual blood for the reason of uterus becoming enlarged in a girl or young woman. The malignant rhabdoid tumor was a renal tumor of infancy and childhood. The malignant rhabdoid tumor develops a pelvic mass and occurs 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. A Round cell tumor is a highly aggressive malignant tumor made up of small and undifferentiated cells with an 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.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES