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Case Report

Retroperitoneal Leiomyosarcoma in an Adolescent: A Case Report

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Abstract

Cancer in adolescents differs from that in adults. The various types of cancer occurring in this age group tend to progress rapidly, necessitating specific, long-term treatment. We report a case of retroperitoneal leiomyosarcoma in a 17-year-old adolescent with no significant medical history, which followed a fulminant course. Cancer in adolescents frequently requires specialized care particularly psychological support given the significant physical, affective, and emotional upheavals involved.

Key Words: adolescence; retroperitoneal leiomyosarcoma; surgery; poor prognosis

Introduction

Retroperitoneal leiomyosarcomas in adolescents are rare malignant tumors. Leiomyosarcomas arise from smooth muscle cells. Twenty percent of leiomyosarcomas are located in the retroperitoneum. They can grow to a very large size while remaining clinically silent, which explains why they are often at an advanced stage by the time medical attention is sought. The diagnosis is frequently suspected based on CT scans and confirmed through histology. Surgery is the treatment of choice. Adjuvant therapy (radiotherapy and/or chemotherapy) may be beneficial in facilitating the resectability of the tumor. The prognosis is typically poor. The purpose of this report is to describe the case of a 17-year-old adolescent with a massive retroperitoneal (retro vesical) leiomyosarcoma.

Case Report:

A 17-year-old adolescent with no significant medical history presented with abdominal pain and a sensation of heaviness in the hypogastric region, accompanied by weight loss (of unspecified amount). A computed tomography (CT) scan revealed a retroperitoneal tumor in the hypogastric area, characterized by soft-tissue density and areas of necrosis. Magnetic resonance imaging (MRI) was not performed. Intraoperative exploration revealed a large, 25-centimeter-diameter retroperitoneal tumor that was firm and adherent to the bladder, sigmoid colon, and iliac vessels (rendering it unresectable). A biopsy was performed for histological analysis, which confirmed a grade III leiomyosarcoma. The progression of the cancer was extremely rapid, occurring amidst a highly stressful and distressing situation for the adolescent and his family: - By postoperative day 1, the mass had reached the level of the umbilicus; - By day 5, it extended beyond the epigastric region, by postoperative day 10, the patient passed away due to respiratory distress.

Discussion

Adolescence is a transitional phase of life between childhood and adulthood. The onset of cancer during this period of physical and emotional upheaval is a traumatic event for the adolescent and those around them [1]. Although rare, cancer is the second leading cause of death after accidents in this age group [2].

The most common types of cancer among adolescents are [3] Lymphomas (22.9 %) Carcinomas and malignant epithelial tumors (malignant melanomas, thyroid carcinomas) (1 9.5 %). Bone and extra-osseous sarcomas (17.6 %). Germ cell tumors (12.7 %). Leukemias (11.9 %) Central nervous system tumors (10.6 %).

Retroperitoneal sarcomas are rare tumors (accounting for 1% of all neoplasms). Leiomyosarcomas represent 20–30 % of these, with 20% occurring in the retroperitoneal space. In this location, leiomyosarcomas can reach a large size before any symptoms manifest meaning patients often do not seek medical attention which explains why they are frequently diagnosed at an advanced stage [4]. There are no specific symptoms associated with leiomyosarcomas. Typically, retroperitoneal leiomyosarcomas present with vague symptoms. The most common findings include abdominal or lumbar pain (40% of cases), weight loss (15- 40% of cases), and an increase in abdominal girth (13 % of cases), or they may be discovered incidentally [5].

The exact cause of the development of leiomyosarcomas is unknown, but certain risk factors have been identified, including prior pelvic irradiation, use of tamoxifen, age, and specific genetic factors [6]. Metastases are rare and tend to occur late. Loco regional recurrence is also uncommon. In the case of our adolescent patient, the presenting complaints were abdominal pain, a sensation of heaviness, and significant weight loss. When a diagnosis is suspected, diagnostic imaging is indicated. The condition is frequently first suspected based on computed tomography (CT) scans, which provide information regarding the size, location, and involvement of surrounding structures. A CT scan typically reveals a heterogeneous, irregular mass, lacking both fatty components and calcifications. A thoraco-abdominopelvic

CT scan with the administration of a contrast agent is essential for the diagnostic process and for developing a therapeutic plan [7].

Magnetic resonance imaging (MRI) appears to have greater sensitivity than CT scanning or ultrasound for distinguishing between a leiomyoma and a leiomyosarcoma. MRI is indicated for patients with an allergy to iodinated contrast agents, as an adjunct to a non-contrast CT scan, or in cases where CT analysis is challenging. It allows for a more precise assessment of muscular, bony, or vascular involvement [7]. PET scans are useful for detecting recurrences; given their superior sensitivity and specificity compared to CT scans in this context, they play a role in the ongoing monitoring of a recurring tumor [8].

A definitive diagnosis can only be established through the histology of a biopsy (spindle shaped cells with hyperchromatic nuclei). This approach helps to rule out several differential diagnoses, including benign tumors, extragonadal germ cell tumors, and lymphomas [9].

In our case, the diagnosis was suspected based on a CT scan and confirmed by histological analysis of a biopsy (grade III leiomyosarcoma). Curative-intent surgical excision is the only treatment. A macroscopically complete resection improves survival, but the rate of local recurrence is generally high [10].

Radio chemotherapy may play a beneficial role in improving resectability. Immunohistochemistry is important. Survival is closely linked to the histological grade of the tumor. In our case, the tumor was unresectable; only a biopsy was performed. No adjuvant treatment was offered. The tumor's progression was fulminant: By the first postoperative day, the mass had reached the umbilicus; by the fifth day, it extended beyond the epigastric region; and the patient died on the tenth postoperative day due to respiratory distress. The prognosis for retroperitoneal leiomyosarcomas is generally poor. The quality of the resection is the most significant prognostic factor in multivariate analysis (R2: 21-month median survival versus R0/R1: 69-month median survival). An R2 resection is an independent predictor of the risk of disease-specific mortality [11].

Recurrences are very frequent, even after an R0 oncological resection. The grade is a powerful, independent prognostic factor for recurrence-free survival [12]. Local recurrence and its treatment are the primary factors contributing to disease-specific mortality in retroperitoneal sarcoma [13]. Given their poor prognosis and the fact that they are frequently of a higher grade, recurrences often require a multidisciplinary management approach [14]. Psychological support should be provided to patients with cancer and their families.

Conclusion

Leiomyosarcomas are tumors with a very poor prognosis. They require a multidisciplinary approach with a particular focus on psychological support for the adolescent and their family, who may be in distress due to the cancer. Cancer in adolescents is a public health issue. It is essential to identify the underlying causes, improve care, and establish a short- and long-term follow-up program for adolescents affected by cancer.

Declaration of interests

authors declare no conflicts of interest.

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