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Case Report

Bulky Pelvic Hodgkin Lymphoma in a Prepubertal Girl

Abstract

Pure infra-diaphragmatic presentation is rare in pediatric Hodgkin lymphoma (HL). Isolated pelvic presentation is restricted to case reports in adults. We describe a unique presentation of HL as a primary pelvic disease in an 11-year-old prepubertal girl. She was treated based on the Euronet-Paediatric Hodgkin's Lymphoma Group protocol (EuroNet-PHL-C1) recommendations and assigned to treatment Group 2 (Stage IIB). The patient had bulky disease and a suboptimal response on interim 18-fluorodeoxyglucose positron emission tomography. The child was administered chemotherapy alone. Pelvic radiotherapy was consciously avoided due to the high risk of gonadal failure. She remains disease-free for 18 months now. Treatment of pediatric HL necessitates a delicate balance between achieving cure as well as avoiding serious late effects of therapy.

Keywords: *Hypogonadism, infradiaphragmatic, late effects, positron emission tomography scan, radiation*

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Introduction

Primary sub-diaphragmatic pediatric Hodgkin lymphoma (HL) comprises merely 3% of all pediatric cases.[1] Isolated HL in the pelvis is rarely reported.[2,3] We report a prepubertal girl with a pelvic mass, which uniquely turned out to be HL. Besides chemotherapy, the management involved debating pros and cons of administering radiotherapy to the sensitive site.

Case Report

An 11-year-old girl presented with fever for 4 months. On examination, a left-sided suprapubic mass was palpated. Ultrasonography confirmed a mass in the pelvis. A magnetic resonance imaging revealed a large mass located in the pelvis, along with enlarged lymph nodes [Figure 1]. The initial clinical possibility was of a gonadal germ cell tumor. Serum alpha-fetoprotein and beta-human chorionic gonadotrophin were normal. A core biopsy was consistent with a diagnosis of HL, nodular sclerosis (NS) type. Immunohistochemistry was confirmatory; the large atypical cells were positive for CD15/CD30 and negative for CD20. An 18-fluorodeoxyglucose-positron emission tomography-computed tomography (FDG-PET-CT) confirmed FDG avidity of the mass and adjoining nodes. The bone marrow was not infiltrated.

The patient was assigned to stage IIB and treatment - Group-2 of the Euronet-Paediatric Hodgkin's Lymphoma Group protocol (EuroNet-PHL-C1).[4] Two cycles of O-vincristine, E-etoposide, P-prednisone, A-adriamycin were administered. An interim FDG-PET-CT revealed >50% reduction of the mass, with a Deauville score (DS) of three, with complete resolution of the nodes. As per the guidelines of EuroNet-PHL-C1 trial, with a local partial remission and a DS of three, radiotherapy was indicated following two cycles of C-cyclophosphamide, O-vincristine, P-prednisone, DAC-dacarbazine (COPDAC). However, there was a major concern for infertility with radiotherapy to the pelvis. The multi-disciplinary team, along with the parents debated the options. The emerging international consensus was of DS of 1–3 being considered as a complete metabolic response at early interim PET.[5] Indeed, in the succeeding Euronet-C2 trial, the threshold of radiotherapy was to be raised to DS of 4–5, instead of 3–5.[6] The team opted to avoid radiotherapy and administer two additional (total: four) cycles of COPDAC, instead. An ultrasonography performed following six cycles of chemotherapy revealed a small cystic residual mass in the region of the left

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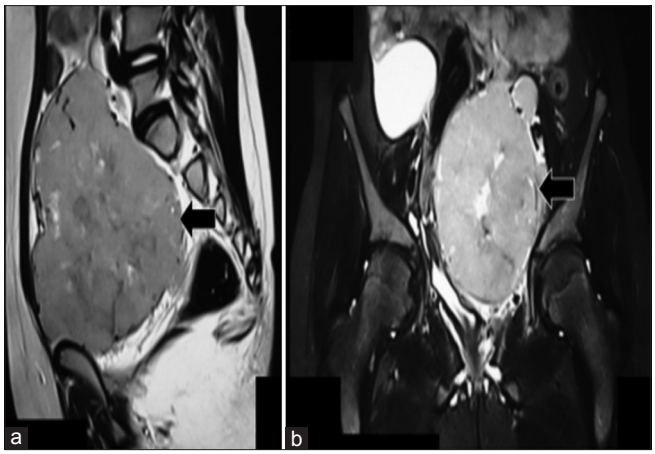

Figure 1: Magnetic resonance imaging (a - sagittal view, b - coronal view) demonstrated a large pelvic mass (arrows in a and b). The left sided ovary was not distinctly identified. The uterus and urinary bladder were displaced and compressed by the mass

adenexa. A diagnostic laparoscopy performed to visualize and biopsy the mass demonstrated trivial bogginess of soft tissue in the pelvic wall with no visible mass. She has been well and in follow-up for 18 months.

Discussion

Primary infradiaphragmatic HL has been reported in 4%–13% of patients in studies encompassing all age groups.[7,8] Published literature on isolated pelvic disease is restricted to adults.[2,3] Infradiaphragmatic HL demonstrates distinct characteristics when compared to supradiaphragmatic disease such as older age, multiple involved sites, increased frequency of lymphocyte predominant histology, and a reduced frequency of NS variants.[7-10] Survival rates are not observed to be different, and recommendations for treatment do not differ.[4,7-9] Bulky disease with an inadequate response on interim FDG-PET is ideally radiated, albeit with increased risk of gonadal failure, premature menopause, and infertility in pelvic HL.[11] According to the Lugano classification for the management of lymphomas, DS-3 on interim FDG-PET indicates a favorable prognosis in most patients; interpretation must take into account timing of assessment, clinical context, and treatment.[5] We consciously avoided radiotherapy and administered two additional cycles of chemotherapy to avoid gonadal failure. A longer follow-up is necessary for ensuring a relapse-free period.

Conclusions

Pelvic HL is a rarity in children. The decision for radiotherapy is arduous due to the risk of infertility.

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Conflicts of interest

There are no conflicts of interest.

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