Paediatric B-lymphoblastic Lymphoma of Bone: Presentation of Two Cases of Extremely Rare Entity

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Introduction. Primary lymphoma of the bone (PLB) is a rare tumour, which represents about 3% of all primary malignant bone tumors and 1% of all malignant lymphomas (Zhou et al., 2014). It is even rarer in children (Glotzbecker et al., 2006). Diffuse large B-cell lymphoma is the most common histological subtype, other variants are mentioned as casuistic (Power et al., 2008). There are only several cases of lymphoblastic PLB published. The exact diagnosis is important, since lymphoblastic PLB should be treated according to acute leukemia protocols with good prognosis (Messina et al., 2014).

Aim, Material and Methods. We present two cases of lymphoblastic PLB in paediatric patients, diagnosed and treated at the Children's Clinical University Hospital (Riga) Hematooncology Department. Patients' data were obtained from the Hospital IS, histological slides – from Pathology Bureau archive, cytological slides and flow cytometry files – from Clinical Laboratory archive.

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Results. Case 1. 14-year-old boy was admitted with bone lesions (vertebrae with fractures, cranium, pelvis, ribs, sternum, right hand, no extraosseous sites). Blood values were normal, with biochemistry of tumor lysis. Marrow histology revealed patchy infiltrates of large cells with multilobated nuclei and phenotype LCA–, CD20–, CD79a+, CD10+, TdT–, CD34+. Flow showed 5% cells of the same phenotype (in addition, CD19+, sIg–, cIgM–). Except for TdT, phenotype was that of B-precursors. The patient was treated by acute leukemia standard risk BFM. He relapsed while on therapy, regimen was intensified; he was transplanted and is currently in remission.

Case 2. 16-year-old boy was hospitalised with B-symptoms, generalised pain in legs and right hand; tender subcutaneous lesions had newly appeared. Blood values were normal except elevated LDH. MRI revealed lesions in both femurs and tibias, right shoulder and pelvic bones (predominantly left). Marrow aspirate and trephine biopsy from the left iliac revealed total infiltration by polymorphic medium-sized cells with phenotype CD45weak, $CD19^{+}$, $CD79a^{+}$, $CD20^{-}$, CD10weak, $CD34^{-}$, TdT^{+}, $bcl2^{+}$. The same infiltrate was found in the skin nodule. B-precursor neoplasm was diagnosed and BFM standard risk treatment commenced. MRD at day 15 was 2% with excellent radiological response, at day 33 MRD was < 10E-4. On day 50, while still on intensive chemotherapy, radiological relapse in pelvic bones was observed, though it was undetectable by bilateral histology and cytology with flow. The patient was switched to high-risk BFM and is currently under treatment.

Conclusions. The cases demonstrate that B-lymphoblastic PBL, besides being extremely rare, associates with considerable diagnostic difficulties. Morphologically, tumour cells in both cases were larger (particularly in case 1) and more pleomorphic than the majority of paediatric B-precursor cell leukemias. There was a considerable patchiness of the bone infiltrate with a high risk of sampling artifact (like in the relapse of case 2). Finally, tumour cells in both cases were of "incomplete" phenotype, lacking some typical blast antigens like CD34 or TdT. The problems led to multiple samples and a need for broad phenotyping panel to exclude mature B-cell neoplasms and tumours of non-hematological origin. Combined clinical, surgical, radiological and laboratory input was necessary in both cases. Contrary to the literature data, however scarce, both patients did not respond well to the initial therapy.