

BACKGROUND: Ewing sarcoma (EWS) / primary neuroectodermic tumor (PNET) is the second more frequent bone neoplasia in Pediatric Oncology. This family of tumors is very aggressive, requiring an intensive treatment, with surgery, radiotherapy and chemotherapy, to be controlled. This treatment is associated with immediate and long term side effects. EWS/PNET is morphologically characterized as "blue and small round tumor cells". Immunohistochemical is essential for the diagnosis. Positive CD99 isn't pathognomonic of this family of tumors. However, it is expressed in the great majority of cases. Molecular biology analysis reveals the presence of t(11;22) (q24;q12) – *EWS* gene in 85% of the cases. The fusion of *EWS* gene 22q12 with *FLI1* 11q24 results in EWS-*FLI1* transcript. Sometimes, clinical presentation of these tumors is atypical, especially in children under 12 months, in whom the primary site frequently occurs in head, soft tissues and bones. In this way, the differential diagnosis can be done specially with granulocytic sarcoma. There is not any case in the literature describing the coexistence of these two diseases, PNET and myeloid leukemia. **HYPOTHESIS:** to determine the coexistence of two different types of cancer vs a single disease with aspects common to two different ones. **METHODS:** A 14-month WB was admitted to the service with irritability and pain while walking. Bone XR showed an isolated femoral lesion. A biopsy of this site was done, and the diagnostic of PNET was established. Work-up with PET CT, bone scintigraphy and bone marrow reveal several other bone lesions and marrow infiltration. Bone marrow studies reveal characteristics compatible with M7 AML, by morphology and immunophenotype (presence of CD13, CD33, CD34, CD38, CD41, CD56, CD 61, and CD117). Somatic karyotype was normal (46XY). The diagnosis of PNET had been done on morphological and immunophenotypical basis, revealing: negative LCA, VII polyclonal Factor, polyclonal myeloperoxidase, CD61, CD34, CD117 (*KIT* gene) and positive CD56 and CD99. *EWSR1* gene was negative by FISH. Initial chemotherapy was started as for PNET. TOPOTECAN and CYCLOPHOSPHAMIDE were given, with prompt symptomatic response and rapid disappearance of the signs of disease, including full marrow remission after this 1st cycle of chemotherapy. **RESULTS:** The enormous challenge regarding the correct diagnosis of this child is well representative of the difficulties determined by the presence of characteristics that are common to more than one type of neoplasia. The characteristics of the tumor biopsy and the clinical evolution presented by this child favors the diagnosis of PNET but the similarity of M7 AML marrow findings ought to be emphasized. This patient is being considered, therefore, as one that harbors a PNET whose characteristics of marrow infiltration resemble the ones presented by M7 AML.