EXTRA-RENAL RHABDOID TUMOR INVOLVING THE LUNG, RIBS AND SPINE IN A 12-YEAR-OLD CHILD*

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The results of complex chemotherapy and surgical treatment of extra-renal rhabdoid tumor with involvement of the lung, ribs and spine in a 12-year-old child are presented. Long-term results with no evidence of continued growth of the tumor and metastasis were followed for up to 3 years from the start of treatment and 2 years 7 months after radical surgery with total (360°) two-level spinal reconstruction.

Key Words: rhabdoid tumor, extra-renal rhabdoid tumor, treatment, spinal reconstruction, long-term results, children, spinal tumor, pediatric oncology.

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Malignant extra-renal rhabdoid tumor (ERRT) is a rare aggressive poorly differentiated type of soft tissue sarcomas, occurring primarily in children, which has morphological presentation similar to Wilms' tumor (nephroblastoma), but localized outside kidney tissue. Grossly, the tumor is an irregularly shaped whitish-gray, multifocal, rather soft and large bundle, containing areas of necrosis and hemorrhage. Microscopically, the tumor had solid sheets and massive trabeculae of relatively large polygonal, spindle-shaped and/or rounded cells with relatively light, moderately polymorphic nuclei with large nucleoli; occasionally, there are some giant multinucleated cells with abundant oxyphilic cytoplasm containing rounded hyaline or glassy inclusions, which sometimes displace the nuclei; mitotic figures are frequently seen. Stroma is hyalinized and may be chondroid in appearance. Immunohistochemistry reveals markers of vimentin, cytokeratin, epithelial membrane antigen (EMA) and CAM 5.2. The tumor has to be differentiated from epithelioid sarcoma, embryonal and alveolar rhabdomyosarcoma, melanoma, poorly differentiated renal cell cancer, malignant mesothelioma, Wilms' tumor, and primitive neuroectodermal tumor [2, 3, 7].

In the late 1990s – early 2000s key publications on the ERRT described particular features of imaging, histological and immunohistochemical verification of various tumor localizations: central nervous system, intestines, liver, orbit, thorax, soft tissue, neck et al. [4, 7, 9, 10]. In the last decade, however, greater emphasis has been placed on clinical, pathological and genetic characteristics of the tumor [6, 8], as well as on existing treatment options [1, 5]. According to the European Rhabdoid Register, even though the use of complex therapy, including targeted chemotherapy, radiation therapy and surgical treatment, increased the average overall survival rate of ERRT patients almost three fold over the last 10 years, it still amounts to only 33 months. [1].

The extreme rarity of the disease allows us to present our own clinical case. Patient P.A., 12 years old, was initially hospitalized to one of St. Petersburg pediatric hospitals with a diagnosis of "acute right-sided pneumonia". He was admitted 2.5 weeks after the onset of the disease, which was accompanied by high fever (40 °C), shortness of breath, and pain in the chest. Antibiotic therapy with Zinnat was ineffective.

Chest radiography (January 17, 2012) revealed total shadowing on the right (Fig. 1a). Pleural puncture produced 1400 mL of hemorrhagic effusion. CT of the chest (January 19, 2012) revealed mass lesion of the posterior mediastinum with destruction of Th_9–Th_{10} vertebral bodies and arches, as well as heads and necks of the adjacent ribs (Fig. 1b–d).

The patient was hospitalized to the department of pediatric hematology of the City Clinical Hospital No 31 in critical condition, caused, in addition to the initial complaints (chest pain, shortness of breath, high fever), by the presence of dry cough, dryness and peeling of the skin, loss of about 20% of body weight (weight on admission about 30 kg). Dense, immobile, soft tissue lesion about 8 cm in diameter without precise contours and painful on palpation was identified in the right paravertebral region of the posterior surface of the thoracic cage at the level of Th_9–Th_{10} vertebrae. Bone scan with Tc^{99} revealed foci of moderate hyperfixation of the radiopharmaceutical on the right: posteriorly along ribs IX, X and paravertebrally in the area of ribs VII, VIII (136–143 %); in the left half of Th_9–Th_{10}