EMBRYONAL RHABDOMYOSARCOMA WITH PSEUDOALVEOLAR PATTERN

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BACKGROUND: Embryonal rhabdomyosarcoma is the most common subtype of rhabdomyosarcoma in childhood. Both PAX3- and PAX7-FOXO1 chimeric genes have been identified in alveolar rhabdomyosarcoma with RT-PCR, and this identification is useful to determine the subtypes of rhabdomyosarcoma.
CASE: The patient was 1-year old female and had a soft tissue tumor measuring 34 x 22 mm in size in the back region. A computed tomography (CT) revealed a heterogeneously enhanced mass in the right paravertebral muscle. A magnetic resonance image (MRI) demonstrated an ill-demarcated lesion, which exhibited low to iso signal intensity on T1-weighted image and high signal intensity on T2-weighted image with fat suppression. Wide resection was performed. However complete resection was difficult because of the tumor invasion into the intervertebral foramen. Grossly, the tumor was solid, and white in color on the cut surface. The tumor was composed of proliferation of atypical spindle-shaped or polygonal cells arranged in nests separated by fibrous septa, mimicking alveolar rhabdomyosarcoma. PAX-FOXO1 chimeric gene could not be detected with RT-PCR, and FOXO1 split also could not be found with FISH. Adjuvant chemotherapy was used postoperatively combined with radiotherapy. Because of the right pulmonary metastasis five months after the operation, partial resection was performed. The tumor was predominantly composed of proliferation of atypical polygonal cells arranged in nests and sheets, indicative of embryonal rhabdomyosarcoma. Eight months later, liver metastasis was found and follow-up with adjuvant chemotherapy is in progress.
CONCLUSION: It was difficult to make a diagnosis of embryonal rhabdomyosarcoma in the primary tumor, because the tumor revealed pseudoalveolar pattern with artifitial damages and fibrocollagenous stroma. The genetic analysis was useful adjunct to make a correct diagnosis in this case.