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Pediatric rhabdomyosarcoma at presentation: Can cross-sectional imaging findings predict pathologic tumor subtype?

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ABSTRACT

Objective: The purpose of the study is to determine whether there are cross-sectional imaging features of pediatric rhabdomyosarcoma that are specific to the different pathologic subtypes of the tumor.
Materials and methods: Medical records of 14 pediatric patients who were diagnosed with rhabdomyosarcoma were reviewed retrospectively. Patient demographics, including age and sex, as well as final pathologic report were obtained. The initial CT, MRI, or both obtained at presentation, prior to the diagnosis being established, were reviewed by two radiologists. We recorded tumor features including site, size, margins, local extension, and presence of metastases. Presence of calcification, hemorrhage, or necrosis as well as attenuation and heterogeneity of the tumor were also recorded.
Results: Ten of our fourteen patients were formally diagnosed with the embryonal subtype of rhabdomyosarcoma, while three were found to have the alveolar subtype, and one subtype was poorly differentiated. There was no significant difference in the attenuation and in the heterogeneity of the tumor between the embryonal and the alveolar subtype on CT.
Conclusion: Imaging features at presentation, such as attenuation and heterogeneity, could not correlate to the pathologic subtype of pediatric rhabdomyosarcoma.

1. Introduction

Rhabdomyosarcoma is a malignant tumor that represents 5% of all childhood cancers [1]. In adults, rhabdomyosarcoma is rare, accounting for only 3% of the soft-tissue sarcomas [2]. The current classification for rhabdomyosarcoma includes embryonal (with the less common subtype botryoid and variant spindle cell) which have an intermediate to superior prognosis, alveolar (with a poorer prognosis), undifferentiated sarcoma (also with a poorer prognosis), and sarcoma not otherwise specified [3]. Although rhabdomyosarcoma shares certain imaging characteristics with other soft-tissue sarcomas, it has been reported that there are rather unique features at presentation which should make the radiologist consider the diagnosis. In a previous study, it was shown that tumor heterogeneity is more prominent in alveolar and pleomorphic subtypes of adult rhabdomyosarcoma. These subtypes were also noted to have extremely high signal on T2 and STIR MRI [4]. The purpose of this study is to evaluate whether similar findings are present in pediatric cases of rhabdomyosarcoma.

2. Methods

We retrospectively reviewed the medical records of 14 pediatric patients diagnosed with rhabdomyosarcoma. Patient demographics, including age and sex, as well as final pathologic reports were obtained. Two radiologists reviewed the initial CT and MRI studies obtained at presentation, prior to the diagnosis. Tumor features including site, size, margins, local extension, and presence of metastases were recorded. Presence of calcification, hemorrhage, or necrosis as well as attenuation and heterogeneity of the tumor were also recorded.

The CT studies were all performed with contrast. On review of these studies, the tumor attenuation was compared to axial muscle attenuation by placing regions of interest in the tumor and in nearby axial muscle. Increased tumor attenuation in comparison to the axial muscle was reported as high attenuation, decreased attenuation was reported as low attenuation, and similar attenuation was reported as iso-attenuation.

In those patients who underwent MRI at presentation, the signal intensities in T1 and T2 unenhanced and gadolinium-enhanced T1 sequences were evaluated by visually evaluating the tumor and comparing to the signal intensities of axial muscle in that sequence. Increased tumor signal was reported as high intensity, similar was reported as iso-intense, and lower was reported as low intensity.

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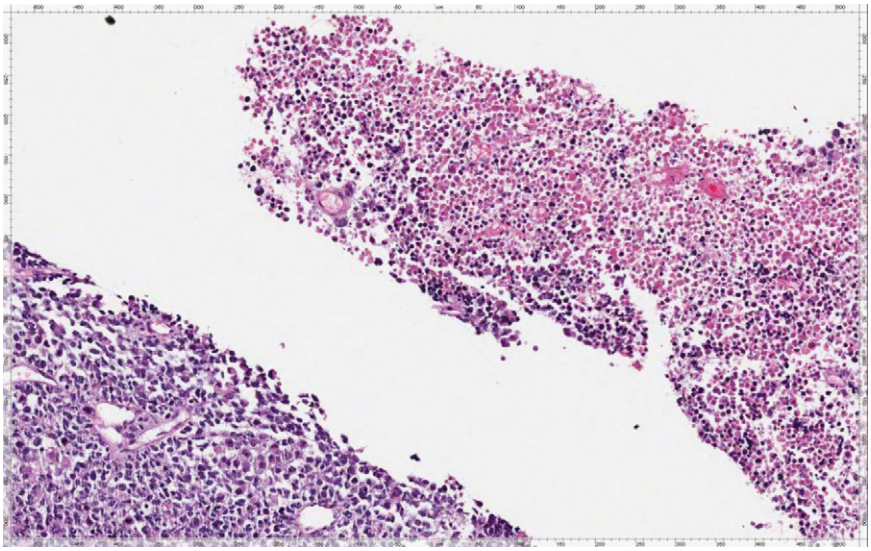

Fig. 1. Poorly differentiated rhabdomyosarcoma (below left) with nest of necrosis (above right). H&E × 200.

3. Results

Eleven of our 14 patients had CT and eight had MRI, with five having both studies at presentation. Ten patients were eventually diagnosed with embryonal rhabdomyosarcoma, and three patients with the alveolar subtype. The other patient was initially diagnosed with poorly differentiated rhabdomyosarcoma (Fig. 1). Repeat biopsy 3 months later revealed maturation of the tumor and abundant rhabdomyoblasts.

Demographic details for each of the cases are shown in Table 1. Table 2 demonstrates the patterns studied for each of the pathologic subtypes. In seven of the ten patients with the embryonal subtype, the tumor was defined as low attenuation in comparison to axial muscle, such as case 4 (Fig. 2). In only one patient with embryonal rhabdomyosarcoma was the tumor found to have high attenuation when compared to axial muscle (Fig. 3). Two of the alveolar subtype cases demonstrated low tumor attenuation with the other noted to have similar attenuation to axial muscle (Fig. 4). In all eleven CT studies, the tumor appeared moderately to markedly heterogeneous.

In all eight patients who underwent MRI, the tumor demonstrated low and homogeneous signal in the T1-weighted sequence. In the T2-weighted and contrast-enhanced T1-weighted sequences, the tumor demonstrated high and heterogeneous signal regardless of the pathologic subtype (Fig. 5).

4. Discussion

There are three known pathologic subtypes of rhabdomyosarcoma: embryonal which is the most common, alveolar, and pleomorphic which occurs almost exclusively in adults.

The embryonal subtype resembles closely various stages in the embryogenesis of normal skeletal muscle. The embryonal subtype

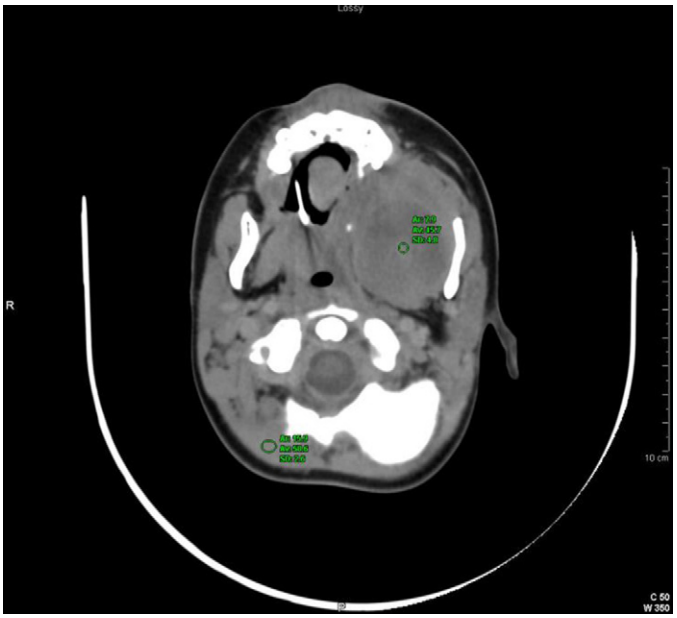

Fig. 2. Embryonal rhabdomyosarcoma of case 4. Tumor located in the left masticator space. Tumor attenuation is less than that of the muscle (45 HU and 58 HU respectively). Tumor is well defined. Bones around tumor are being remodeled without gross invasion. The tumor has markedly heterogeneous enhancement.

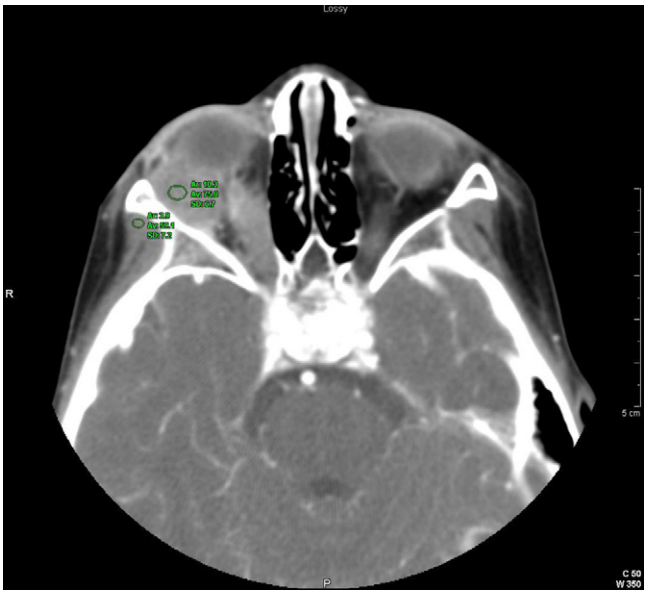

Fig. 3. Embryonal tumor (case 7) originating in the right inferior rectus muscle demonstrates hyperattenuation (HU 75) as compared to nearby muscle (HU 59).