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Rhabdomyosarcoma in adrenal region of a child with hypertension and fever: A case report and literature review☆

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Abstract Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children but rarely involves the adrenal. We describe a case of alveolar rhabdomyosarcoma (ARMS) of the right adrenal in a 5-year-old boy with a characteristic history of hypertension and recurrent fever. On surveillance imaging, a right adrenal mass was incidentally detected, and a right adrenalectomy was subsequently performed. After the surgery, the fever disappeared simultaneously, and the blood pressure gradually returned to normal level. This is the first reported case in children.

Rhabdomyosarcoma (RMS) is a rare mesenchymal malignancy that typically affects children and adolescents. RMS accounts for approximately 3% to 7% of all malignancies in children and adolescents, with an incidence of approximately 4.4–4.5 cases per million children/adolescents per year [1]. Theoretically, RMS can affect any body part. However, only one RMS case in the adrenal region has been described in the literature [2]. Here we report a case of alveolar RMS (ARMS) in the right adrenal region of a pediatric patient with a characteristic history of hypertension and fever, and review the literature on this extremely rare disease.

A five year old boy presented on admission to our hospital with a 3-month history of recurrent fever (usually about 38–39 °C, occasionally >39.5 °C) unresponsive to antibiotic treatment and moderate hypertension (occasionally >125 mmHg), accompanied by a slight cough. The general physical examination was unremarkable except for hepatosplenomegaly. The laboratory examination revealed an elevated WBC count of 20.6×10^9/L (accompanied by an abnormal white blood cell differential) and serum C-reactive protein (CRP) level of 168 mg/dl. The other laboratory studies, including basal endocrine data of adrenal (such as 17-OH corticosteroids, 17-ketosteroid and vanillylmandelic acid) were within normal limits. However, ultrasonography revealed a hypoechoic mass in the right adrenal region (Fig. 1A). On multiphasic contrast-enhanced CT images, the mass was slightly heterogeneous hypodense (attenuation value of 42 HU, Hounsfield unit) on pre-enhanced images, with moderate enhancement at the venous phase and

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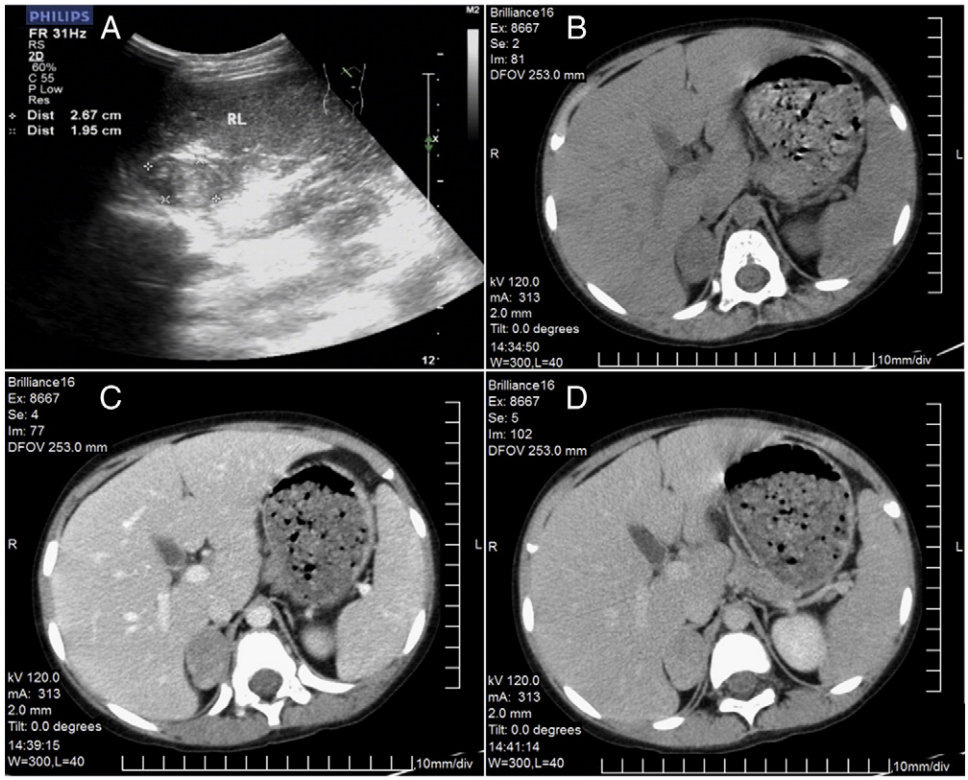

Fig. 1 Diagnostic images of the right adrenal tumor. (A) Ultrasonography (US); (B) Pre-enhanced CT image; (C) Venous phase CT image; (D) Delayed phase CT image.

continued enhancement at the delayed phase (attenuation values of 85 HU and 92 HU, respectively). According to the imaging and clinical manifestations, a typical adrenal adenoma should not be the first consideration of diagnosis and a malignancy couldn't be ruled out. The patient subsequently underwent adrenalectomy, and an encapsulated and well-demarcated tumor that measured 4 cm in diameter was confirmed to arise from the right adrenal. The specimen was fish-like in appearance and tan-red-whitish in color, with no foci of hemorrhage or necrosis. Histologically, the tumor was composed of small round and oval cells with typical gland cellular and false glands-like architecture. Immunostains were positive for Myogenin, actin(α-sarcomeric) and vimentin, whereas negative for CgA, EMA, Syn, CD34, S100, HHF35, and F8 (Fig. 2). Adrenal alveolar-type rhabdomyosarcoma (ARMS) was diagnosed. The patient underwent an uneventful postoperative course, and was discharged without postoperative chemotherapy or radiotherapy. After the operation, the fever disappeared simultaneously, and the blood pressure gradually returned to normal levels spontaneously within 3 months. No recurrence or metastases were detected during a 22-month follow-up up to now.

RMS is the most common soft tissue sarcoma (STS) in the pediatric population, accounting for more than 50% of all pediatric STSs. Various environmental exposures or clinical factors are reported to be associated with an increased risk of RMS, and genetic changes may also play an important role in the development of RMS [3]. The cell of origin of RMS still remains unclear and controversial. RMS was initially considered to be originating from myogenic cells. However, some studies suggest that RMS can arise from non-muscle cells, such as mesenchymal stem cell (MSC) [4], which may explain its virtual occurrence in any body part, including the retroperitoneal spaces or adrenal glands, as shown in our case.

Based on the histological and biological features, RMS is classified into three subtypes, with embryonal (ERMS) and alveolar (ARMS) subtypes making up the vast majority of cases [5,6]. Compared to ERMS, ARMS is predominantly found in the trunk and extremities and is more prevalent among older children and adolescents with over 50% of the cases being older than 10, and is clinically more aggressive. The overall incidence of RMS and ERMS remained stable over the last 30 years; however, there was a statistically significant increase in the incidence of ARMS [7]. As RMS is resistant to chemotherapy and radiotherapy, surgical resection remains the standard treatment especially for localized tumors, and the prognosis is associated with the quality of surgery [8].

It is extremely difficult to preoperatively diagnose an adrenal incidentaloma as RMS. However, some CT imaging features may be helpful to distinguish between an adenoma and a non-adenoma including the malignancy. Lipid-rich adenomas are characterized by an attenuation value of less than 10 HU at unenhanced CT. Lipid-poor adenomas can be