

## PERINEAL LOCALIZATION OF RHABDOMYOSARCOMA IN CHILDREN

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**R**habdomyosarcoma is one of the most common tumors in the pediatric age, but perineal localization is very rare, with a poor prognosis due to late diagnosis. Magnetic resonance (MR) is a non-invasive technique that is very useful in the evaluation of local infiltration and can facilitate the diagnosis. In this article, we present a case of perineal rhabdomyosarcoma diagnosed with pelvic MR, with simultaneous presence of multiple synchronous metastases.

Keywords: alveolar rhabdomyosarcoma, adolescent, perineum, magnetic resonance imaging, neoplasm metastases.

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## ПЕРИНЕАЛЬНАЯ ЛОКАЛИЗАЦИЯ РАБДОМИОСАРКОМЫ У ДЕТЕЙ

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**Р**абдомиосаркома является одной из самых распространенных опухолей детского возраста, однако локализация её в промежности встречается очень редко, с плохим прогнозом из-за отсутствия ранней диагностики. Магнитно-резонансная томография (МРТ) – это неинвазивный метод, который очень полезен при оценке местной инфильтрации и может существенно облегчить диагностику. В этой статье мы представляем случай рабдомиосаркомы промежности, выявленной с помощью МРТ малого таза с одновременным наличием множественных метастазов.

Ключевые слова: альвеолярная рабдомиосаркома, подросток, промежность, магнитно-резонансная томография, метастазы.

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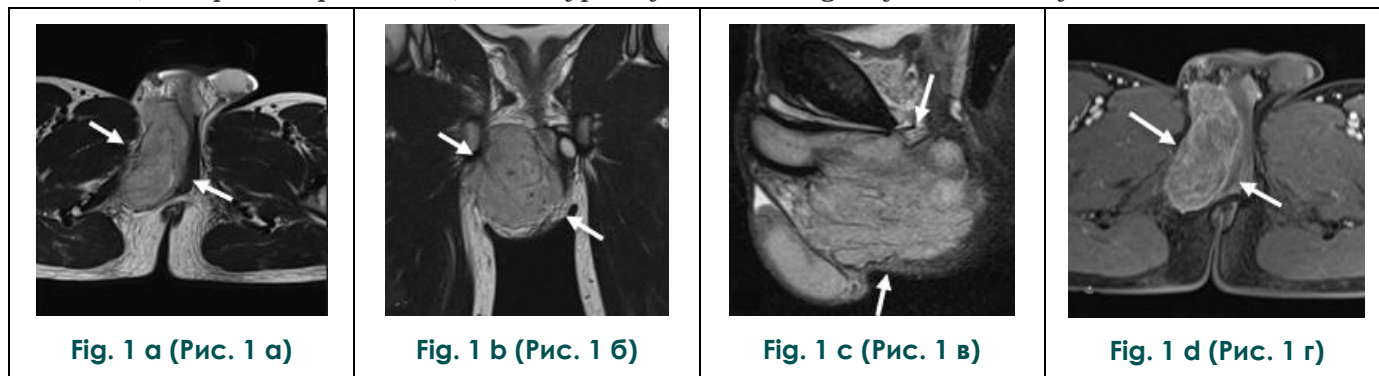
**I**ntroduction.

Rhabdomyosarcoma (RMS) is one of the most common tumors in the pediatric age, representing 5% of childhood tumors and about 50% of all childhood sarcomas [1, 2].

RMS has traditionally been classified into embryonal and alveolar RMS, typically seen in childhood, and pleomorphic RMS, which typically

**Case report.**

A 16-year-old boy was admitted to our pediatric emergency department due to perineal pain and a palpable stiff mass in the perineum. Physical examination showed a perineal mass, associated with right inguinal swelling, suspected of being an abscess or a massive lymphadenopathy. Urine and blood test analysis were performed, showing only mild leukocytosis.



**Fig. 1.** MRI, a, d – axial views, b – coronal view, c – sagittal view.

Perineal rhabdomyosarcoma. Pelvic MR shows a perineal mass (white arrows) which appears mildly hyperintense in axial (a) and coronal (b) T2w images, hyperintense in sagittal T2w STIR (c) and with non-homogeneous contrast enhancement on axial T1w GE FS image (d). The mass infiltrates the right ischio-pubicus ramus (a, d), the right ischio-cavernosus, puborectal and external sphincter muscles (b), the right corpus cavernosum, extending up to corpus spongiosum (c).

**Рис. 1.** МРТ, а, г – аксиальная плоскость, б – корональная плоскость, в – сагиттальная плоскость.

Рабдомиосаркома промежности. При МРТ малого таза выявлено образование промежности (белые стрелки), которое выглядит слегка гиперинтенсивным в аксиальной (а) и корональной (б) плоскостях на Т2w изображениях, гиперинтенсивным в сагиттальной плоскости на Т2w STIR изображениях (в) и с неоднородным контрастным усилением в аксиальной плоскости на Т1w GE FS изображениях (г). Образование инфильтрирует правую ишио-лобковую ветвь (а, г), правую ишио-кавернозную мышцу, лобково-прямокишечную мышцу и мышцы наружного сфинктера (б), а также правое пещеристое тело с распространением до губчатого тела (в).

affects adult age [2]. Head and neck are the most common sites of RMS, accounted in about 40% of cases, followed by pelvic and genitourinary region, in 20% of cases, whilst perineal localization is very rare [3].

Perineal rhabdomyosarcoma (PRMS) accounts for 2% of all RMSs, with a poor prognosis and a 5-year survival between 20% and 49% [4]. Although in RMSs the embryonal subtype prevails, the alveolar subtype is the most common of PRMSs, with a slight predominance in adolescents [1, 3, 5].

Magnetic resonance (MR) is a non-invasive technique that is very useful in the evaluation of local infiltration and can facilitate the diagnosis [2]. It is preferred to other non-invasive techniques due to its accuracy and the absence of ionizing radiation.

We report a rare case of alveolar PRMS in a 16-year-old boy, who presented with perineal pain. MR revealed a 8.3-cm solid perineal mass with right corpus cavernosum, corpus spongiosum and local muscle infiltration.

A pelvic MR was performed using a 1.5 T scan (Siemens Avanto, Siemens Medical Solutions, Erlangen, Germany), with a superficial 16-channel body coil. Three-plane localizer, axial T1 weighted (w) Turbo Spin Echo (TSE), axial and coronal T2w TSE and axial and coronal T2w Short Time Inversion Recovery (STIR) images were obtained. Further, axial T1w Gradient Echo (GE) Fat Sat (FS) images were acquired before and after Gadolinium-based contrast administration (respectively 30, 60 and 120 sec).

An oval-shaped, well-defined solid mass was detected in the perineum. It measured 4.3 x 8.3 cm x 6.6 cm (LL x AP x CC) and appeared isohypointense on T1w TSE and mildly hyperintense on T2w TSE and T2w STIR images. After contrast administration, a rapid and non-homogeneous contrast enhancement was observed on T1 GE FS images (Fig. 1).

As for extension, the mass interrupted and invaded the right corpus cavernosum, extending up to the corpus spongiosum. Moreover, the mass was adherent to the right ischio-pubicus ramus,

invading the right ischio-cavernosus, puborectal and external anal sphincter muscles.

Multiple enlarged lymph nodes were found in the ipsilateral external iliac chain and in the inguinal region.

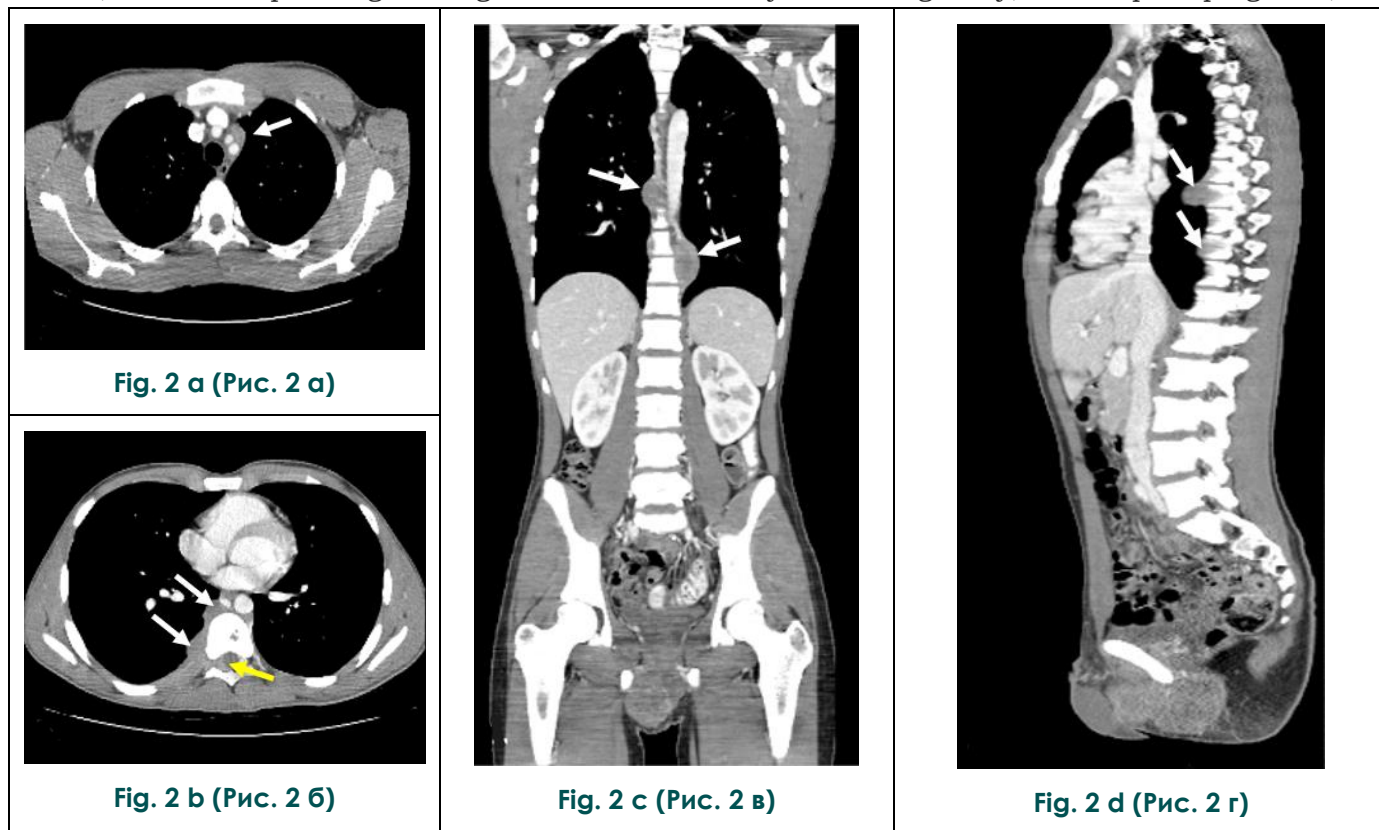
MR features raised the suspicion of perineal sarcoma.

A biopsy of the perineal mass was then performed, with a histopathological diagnosis of alve-

um, was administered, showing an initial good response. Six months later other secondary lesions appeared in the knee and in the soft tissue of the scalp with meningeal infiltration. The patient died 10 months after his diagnosis from complications related to his metastatic disease.

**Discussion.**

Perineal rhabdomyosarcoma (PRMS) is a very rare malignancy, with a poor prognosis, es-



**Fig. 2.** MSCT, a, b – axial view, c – coronal view, d – sagittal view.

Distant metastases from perineal rhabdomyosarcoma. Whole-body Computed Tomography with contrast administration shows multiple localization (white arrows) in anterior (a), posterior mediastinum and paravertebral regions (b, c, d), with invasion of the spinal canal (b, yellow arrow).

**Рис. 2.** МСКТ, а, б – аксиальная реконструкция, с – корональная реконструкция, д – сагиттальная реконструкция.

Рабдомиосаркома промежности. При МРТ малого таза выявлено образование промежности (белые стрелки), которое выглядит слегка гиперинтенсивным в аксиальной (а) и корональной (б) плоскостях на T2w изображениях, гиперинтенсивным в сагиттальной плоскости на T2w STIR изображениях (в) и с неоднородным контрастным усилением в аксиальной плоскости на T1w GE FS изображениях (г). Образование инфильтрирует правую ишио-лобковую ветвь (а, г), правую ишио-кавернозную мышцу, лобково-прямокишечную мышцу и мышцы наружного сфинктера (б), а также правое пещеристое тело с распространением до губчатого тела (в).

olar rhabdomyosarcoma (Alk+, Desmin+, and Myogenin+).

A whole-body Computed Tomography (CT) was then performed for staging, showing multiple lesions in the anterior and posterior mediastinum and in the paravertebral regions with an invasion of the spinal canal (Fig. 2).

Due to the dissemination of the disease, the patient was not eligible for surgery. Chemotherapy, followed by local radiotherapy in the perine-

essentially due to late diagnosis and non-specific symptoms which often pass misdiagnosed due to the rarity of the disease. Differential diagnosis is extensive and should include a duplication cyst, a perianal abscess, a vascular malformation, and a malignancy; even though the perineal abscess has a higher prevalence in adult age, it is encountered as the most common cause of perineal mass even in childhood [5]. Clinical and laboratory findings of perineal abscesses are usually nonspecific and

a differential diagnosis is difficult: local pain and swelling are often the only non-specific symptoms, whereas fever is variable (20-90%), and elevation of white blood cell count is observed in less than 2/3 of cases [5].

Ultrasonography is the first non-invasive technique used, because it is able to discriminate some perineal masses, detecting morphology and gross texture of lesions; intralesional vascularization can be assessed with Color-Doppler evaluation, but it is often inaccurate [6].

CT has a low spatial and tissue discrimination and it is usually required for staging and follow-up, checking for pathological lymph nodes and secondary lesions [7, 8]. Moreover, in children, use of ionizing radiation should be avoided if not strictly required.

Pelvic MR is a non-invasive multimodality imaging technique with a high spatial resolution and a precise tissue characterization, able to discriminate benign from malignant processes and to orientate toward the hypothesis of a specific lesion. According to literature, RMS is usually isohypointense on T1w images, moderately hyperintense on T2w images, hyperintense on T2w STIR images and shows a heterogeneous enhancement after contrast administration on T1w images, as observed in our case [1, 3, 8]. No peculiar MR fea-

tures are described in literature for each RMS subtype.

On diagnosis, about 10-20% of RMSs present metastases, and the most common sites of metastases are bones, lungs and distant lymph nodes [8]. In our case, secondary localizations were reported at staging CT in anterior and posterior mediastinum and both in local and distant lymph nodes.

PRMS has a poor prognosis due to its late detection with an advanced-stage diagnosis due to the large size of the tumor and the involvement of local lymph nodes.

#### Conclusion.

PRMS is a rare tumor, with a poor prognosis and a high mortality that should be considered in the differential diagnosis of perineal masses in children and adolescents. MR is a non-invasive multimodality imaging technique useful in the diagnosis and in the local staging of the tumor and in guiding choice of therapies.

#### Ethics statement.

This study received approval form Ethical Committee of Foundation IRCCS Ca' Granda Maggiore Policlinico Hospital and patient provided written informed consent for publication of his case details and accompanying images.

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