СЛУЧАЙ ИЗ ПРАКТИКИ

THE USEFULNESS OF ULTRASOUND FOR THE IN-SITU DETECTION OF NASOGENIAN RHABDOMYOSARCOMA IN LACTATION AGE

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Rhabdomyosarcoma is a relatively common tumour in childhood and adolescence with a tendency for muscle cell-like differentiation. In many cases, it presents as a mass that displaces other tissues. Ultrasound is very accessible around the world, the only source of soft tissue imaging in others, and it is already the predilected study for paratesticular rhabdomyosarcoma; therefore, it should be seen as an instrument for improving time to diagnosis in other types of rhabdomyosarcoma, which can enhance the survival rate among children with this type of cancer, currently reported up to 74%. There has been hardly any image evidence in the literature of embryonal rhabdomyosarcomas detected by ultrasound. This report presents the US and CT findings of a nasogenian rhabdomyosarcoma with a brief review of the literature.

Keywords: rhabdomyosarcoma, embryonal rhabdomyosarcoma, paediatrics, doppler ultrasound, lactation age.

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ВОЗМОЖНОСТИ УЛЬТРАЗВУКОВОГО ИССЛЕДОВАНИЯ В ДИАГНОСТИКЕ НАЗОГЕННОЙ РАБДОМИОСАРКОМЫ У ДЕТЕЙ ЛАКТАЦИОННОГО ПЕРИОДА

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Рабдомиосаркома – часто встречающаяся опухоль в детском и подростковом возрасте с тенденцией к дифференцировке, подобной мышечным клеткам. Во многих случаях это образование смещает другие ткани. Ультразвуковое исследование (УЗИ) является широко доступным во всем мире, позволяет визуализировать мягкие ткани, а также является методом выбора при исследовании паратестикулярной рабдомиосаркомы, поэтому его следует рассматривать как предпочтительный метод диагностики других типов рабдомиосаркомы, что поможет увеличить выживаемость среди детей с этим типом рака, которая, как сообщается, в настоящее время достигает 74%. В литературе почти не встречаются УЗ-изображения эмбриональных рабдомиосарком. В этой статьей представлены результаты УЗИ и КТ назогенной рабдомиосаркомы и краткий обзор литературы.

Ключевые слова: рабдомиосаркома, эмбриональная рабдомиосаркома, педиатрия, ультразвуковая допплерография, лактационный возраст.

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Embryonal rhabdomyosarcoma (RMS) is the most common soft tissue neoplasm in childhood. It accounts for 7% of all malignancies; the incidence is higher in boys than in girls and approximately 350 cases per year are reported in the United States [1-3]. Most RMS are sporadic, although it seems to have an underlying genetic risk factor; the most common genetic syndromes associated are Li-Fraumeni syndrome, type 1 neurofibromatosis, Costello and Beckwith-Wiedemann [4]. However, overall survival has increased considerably from 55% to 63% (IRS I), to 71% (IRS II) and currently up to 80% (IRS-III/IV) due to the efforts of the Intergroup Rhabdomyosarcoma Study (IRS) protocols, utilizing multimodality therapy [2]. Small tumours and gross total tumour removal have favourable prognostic factors; therefore, early identification by accessible imaging methods, such as ultrasound, contributes to staging, correct and timely therapeutic decisions and monitoring of these patients.

This case aims to evidence the beneficial use of ultrasound on a pediatric patient diagnosed with embryonal parameningeal rhabdomyosarcoma in the nasogenian region in an early stage and to contribute to previously unseen images in the literature by this method.

Clinical case.

A 3-month-old male whose mother has identified increased volume in the left nasogenian region without other symptoms. He was brought to the pediatric service, and a rounded lesion attached to deep skin layers and non-painful and intact skin were documented. A soft tissue ultrasound was performed, and a hypoechoic ovoid neoplastic lesion with regular and well-defined margins was observed in the subcutaneous tissue of the left nasogenian region. It showed limited vascularity during the colour Doppler analysis. The size was 20.6 x 15.8 x 11.3 mm, and the centre of the tumour was 6.2 mm from the skin. The volume effect over adjacent tissues and the left maxillary bone contact was documented (Fig. 1).

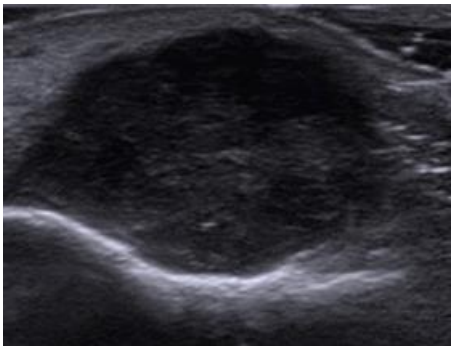


Fig. 1 a (Рис. 1 а)

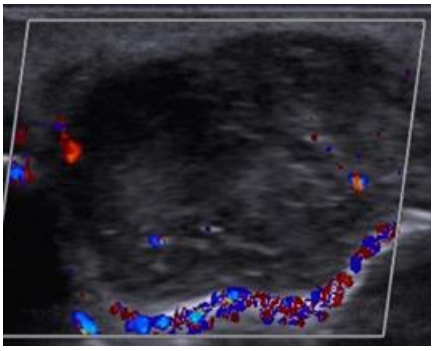


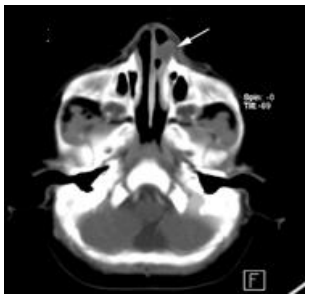
Fig. 1 b (Рис. 1 б)

Fig. 1. **US. A – transverse grayscale, B – colour Doppler Ultrasound.**

Ovoid mass in the left nasogenian subcutaneous tissue with soft tissue echotexture, isoechoic to muscle with well-defined margins and volume effect. Little vascularity is shown – aliasing effect of left maxillary bone tissue.

Рис. 1. **УЗИ. А – серая шкала, Б – цветовое Допплеровское картирование.**

Овальное образование в подкожной клетчатке носоглотки слева, мягкотканной структуры, с четкими контурами, изоэхогенное мышце. Визуализируется незначительная васкуляризация – эффект алиасинга левой верхнечелюстной кости.



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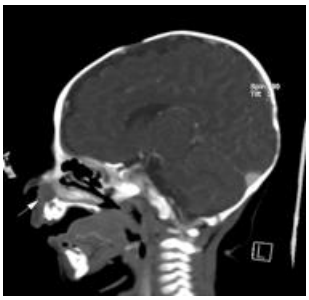


Fig. 2 b (Рис. 2 б)

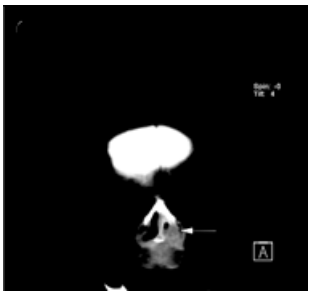


Fig. 2 c (Рис. 2 в)

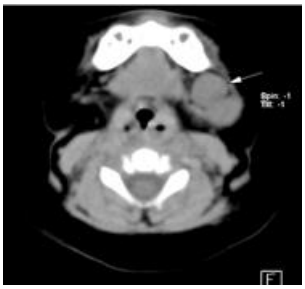
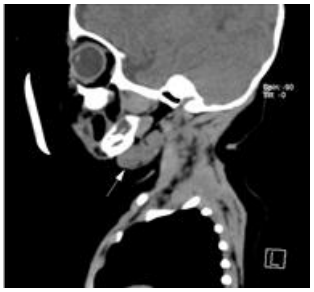


Fig. 2 d (Рис. 2 г)



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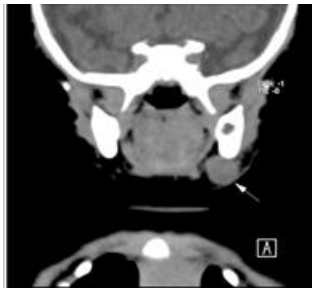


Fig. 2 f (Рис. 2 е)

Fig. 2. **Multiplanar simple and contrasted brain CT.**

A, D – axial views, B, E – sagittal views, C, F – coronal views. A, B, C – Axial, sagittal and coronal planes showed the location of the nasogenian rhabdomyosarcoma (white arrows). D, E, F – Axial, sagittal and coronal planes depict the location of retromandibular adenopathy.

Рис. 2. **КТ черепа, нативное изображение и с КУ.**

А, Г – аксиальная плоскость, Б, Д –сагиттальная плоскость, В, Е – корональная плоскость. А, Б, В – показано расположение назогенной рабдомиосаркомы (белые стрелки). Г, Д, Е – показано расположение ретромандибулярной аденопатии.

Head and neck computed tomography showed a hypodense mass (attenuation pattern of 35–39 UH), obliterating the ipsilateral nostril and contacts with the nasal septum mucosa in the Cottle area II (Fig. 2).

Lymph nodes were identified in the left Ib cervical level with a contrast agent enhancement. (Fig. 3).

The tumour was resected and biopsied, the histopathological analysis reported embryonal rhabdomyosarcoma, and the immunohistochemistry for smooth muscle actin, myogenin, myoglobin, and CD99 was a positive proliferation rate of 20% (Ki67)

The patient was scheduled for chemotherapy treatment and will continue ongoing follow-up oncological outpatient consultation.

Discussion

Historical data.

Rhabdomyosarcoma is the most common soft tissue neoplasm in childhood, and it can occur anywhere in the body; however, its most frequent location is on the head and neck. [3, 5]. It is divided according to its anatomical origin into parameningeal, orbital and other parts or non-parameningeal, the former being the most frequent and with the worst prognosis [2]. In 1972 the Intergroup Rhabdomyosarcoma Study (IRS) Committee was formed; three study groups of pediatric oncological patients published four clinical trials, which compared treatments by clinical group (I) to improve survival (II), treatment (III), and results according to surgical, radio and chemotherapy risk (IV).

Clinical relevance of this report.

This tumour location has not been documented by ultrasound, a benign imaging method for children. The tumour could be characterized with more images, and an early diagnosis would be obtained, avoiding ionizing radiation. To the best of our knowledge, there are no reports in PubMed evincing the detection of rhabdomyosarcoma in the nasopharynx region by ultrasound.

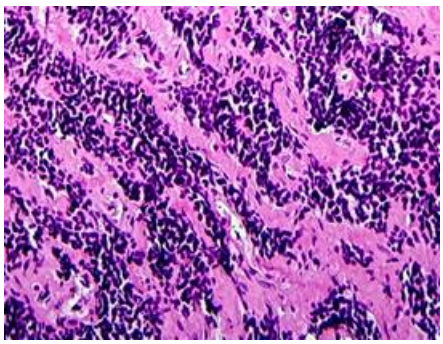


Fig. 3 a (Рис. 3 а)

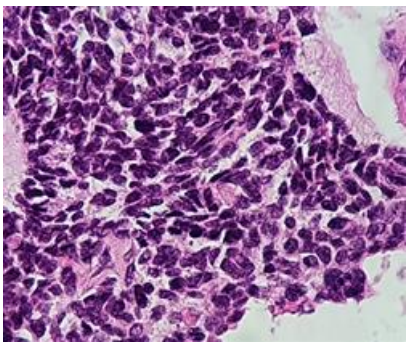
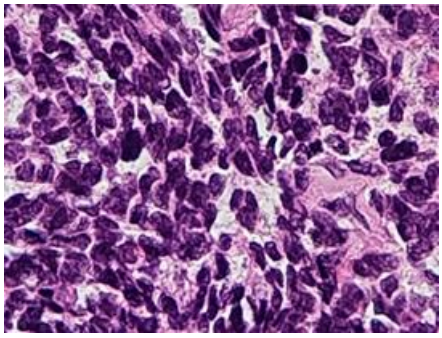


Fig. 3 b (Рис. 3 б)



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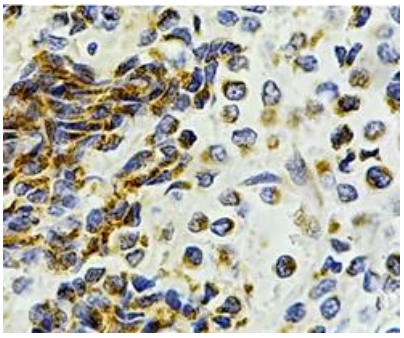

Fig. 3 d (Рис. 3 г)

Fig. 3. Microsection.

A – small round blue cell tumour (H&E; 4x) with neoplastic cells, B, C – nucleomegalia and hyperchromatism with scant cytoplasm (H&E; 10x and 40x). D – Neoplastic cells were myogenin positive, which supported a diagnosis of embryonal rhabdomyosarcoma (Myogenin; 40x).

Рис. 3. Микропрепарат.

А – мелкокруглоклеточная опухоль с окрашиванием в синий цвет (окраска гематоксилин-эозином; 4х), с неопластическими клетками. Б, В – Нуклеомегалия и гиперхроматизм со скудной цитоплазмой (окраска гематоксилин-эозином; 10х и 40х). Г – Неопластические клетки были миогенин-позитивными, что подтверждало диагноз эмбриональной рабдомиосаркомы (миогенин; 40х).

Histology.

It is a blue cell tumour of undifferentiated musculoskeletal tissue and is divided into embryonic, alveolar and pleomorphic subtypes; the embryonic type is more frequent with a prevalence of 60% and occurs in children; in general, it has a better prognosis than the alveolar variant, which occurs in adolescents, who have a lower survival rate [6, 7].

Classification.

According to the Intergroup Rhabdomyosarcoma Study, the classification is based on the clinical group, stage, and risk at presurgical (Table 1) and postsurgical staging classification state (Table 2) [6, 7].

tion, vascularity, dimensions and relationships with adjacent structures; it may also be helpful to perform an ultrasound-guided biopsy of the lesion [4, 8].

Treatment.

Rhabdomyosarcoma treatment is multidisciplinary; surgical resection can be carried out in patients with localized tumours; however, the backbone is chemotherapy; radiotherapy is helpful in some cases [9].

Clinical presentation.

Clinical manifestation can be highly variable and depends on the primary tumour location and the presence or absence of metastases. Rhabdomyosarcoma often presents with a

Table №1. IRSG presurgical staging classification [6].

Stage	Sites	Tumour	Size (diameter)	Node	Metastases
I	Orbit, head and neck (excluding parameningeal), genitourinary	Confined/Extension to surrounding tissue	≤ 5 / >5 cm	Not involved/regional involved/unknown	No
II	Bladder/prostate, extremity, cranial, parameningeal, other	Confined/Extension to surrounding tissue	≤ 5 cm	Not involved/ unknown	No
III	Bladder/prostate, extremity, cranial parameningeal, other	Confined/Extension to surrounding tissue	≤ 5 cm	Regional involved	No
IV	All	Confined/Extension to surrounding tissue	≤ 5 cm	Not involved/regional involved	Yes

Imaging findings.

Although confirmation is made by histopathological analysis, imaging studies are decisive for the diagnosis and applicable in staging and surgical planning. CT and MRI are essential in diagnosing and following RMS; they are also used to evaluate tumour response to treatment and recurrences [4]. However, they do not allow the characterization of superficial lesions, which are predominantly small. On the other hand, Doppler ultrasound is non-invasive; it does not require contrast material, radiation, or general anaesthesia. It is even valuable for early diagnosis due to its easy access, cost-benefit relation, portability and predominant imaging modality in the pediatric population [4]. Finally, ultrasound allows us to obtain valuable information on the composi-

visible or palpable mass or compression or invasion of adjacent structures [10]. According to the site of involvement, this type of cancer has an unspecific presentation with a variable prognosis (Table 3). Due to its slow growth rate, alveolar subtypes can be extended by diagnosis and have poor prognostic [4].

Complications.

Relapse usually occurs within the first three years. Most relapses in localized rhabdomyosarcoma are at the primary tumour site (75%) with or without nodal and distant metastases, with a poor prognosis, although some children remain curable [10]. In addition, its location at the base of the skull, close to vital nerves and vessels, often results in a more complex surgical resection [10]. Metastases are more frequent in lungs and bones [4].

Table №2. IRSG Postsurgical Grouping Classification [6].

Group 1	**Localized disease, completely excised, no microscopic residual**
A	Confined to the site of origin
B	Infiltrating beyond the site of origin
Group 2	Total gross resection
A	With evidence of microscopic local residual
B	Regional disease with involved lymph nodes
C	Microscopic local and nodal residual
Group 3	Incomplete resection
Group 4	Distant metastases

Table №3. Primary site involvement and survival [2, 4

Site	5-year survival	Predilected subtype
Genitourinary	85%	Embryonal
Extremities	74%	Alveolar
Head and neck	80%	Embryonal
Parameningeal (nasopharynx, paranasal sinus, middle ear-mastoid)	74%	
Superficial head and neck (scalp, external ear, parotid, face, buccal mucosa, pharynx, tonsil, larynx, neck)	78%	
Orbit (orbit, periorbital, eyelid)	95%	

Differential diagnosis.

Differential diagnoses should include neuroblastoma, Ewing's sarcoma, and lymphoma [4].

Conclusion.

We present the case of an infant diagnosed with an embryonal variant of a parameningeal rhabdomyosarcoma of nasogenian location that was identified early by ultrasound, which made it possible to treat the lesion and acquire images that had not yet been seen in the literature on rhabdomyosarcoma in this location.

Conflicts of Interest.

The authors declare that there is no conflict of interest regarding the publication of this article.

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References:

1. Yoneyama K, Nakagawa M, Hara A. Benign phyllodes tumor with hemorrhagic cyst in a 14-year-old girl: A case report. Int J Surg Case Rep. 2020; 67: 114-116. doi:10.1016/j.ijscr.2020.01.037

2. Fernández-Ferreira R, Arroyave-Ramírez A, Motola-Kuba D, Alvarado-Luna G, Mackinney-Novelo I, Segura-Rivera R. Giant Benign Mammary Phyllodes Tumor: Report of a Case and Review of the Literature. Case Rep Oncol. 2021; 14 (1): 123-133. doi:10.1159/000510741

3. Fede Â BS, Pereira Souza R, Doi M, De Brot M, Aparecida Bueno de Toledo Osorio C, Rocha Melo Gondim G, Casali-da-Rocha JC, Jbili R, Bitencourt AGV, Alves de Souza J, Caparica Bitton R, Baroni Alves Makdissi F, Moraes Sanches S. Malignant Phyllodes Tumor of the Breast: A Practice Review. Clin Pract. 2021; 11 (2): 205-215. doi:10.3390/clinpract11020030

4. Abdul Hamid S, Rahmat K, Ramli MT, Fadzli F, Jamaris S, See MH, Teoh KH, Taib NAM. Radiopathological characteristics and outcomes of phyllodes tumor of the breast in Malaysian women. Medicine (Baltimore). 2018; 97 (31): e11412. doi:10.1097/md.0000000000011412

5. Macdonald OK, Lee CM, Tward JD, Chappel CD, Gaffney DK. Malignant phyllodes tumor of the female breast: association of primary therapy with cause-specific survival from the Surveillance, Epidemiology, and End Results (SEER) program. Cancer. 2006; 107 (9): 2127-2133. doi:10.1002/cncr.22228

6. Foucar CE, Hardy A, Siziopikou KP, Wang L, Parini V, Hansen N, Jeruss JS. A mother and daughter with phyllodes tumors of the breast. Clin Breast Cancer. 2012; 12 (5): 373-377. doi:10.1016/j.clbc.2012.07.011

7. Mishra SP, Tiwary SK, Mishra M, Khanna AK. Phyllodes tumor of breast: a review article. ISRN Surg. 2013; 2013: 361469. doi:10.1155/2013/361469

8. Zhang Y, Kleer CG. Phyllodes Tumor of the Breast: Histopathologic Features, Differential Diagnosis, and Molecular/Genetic Updates. Arch Pathol Lab Med. 2016; 140 (7): 665-671. doi:10.5858/arpa.2016-0042-RA

9. Donegan WL. Sarcoma of the breast. Major Probl Clin Surg. 1979; 5: 504-542

10. Limaiem F, Kashyap S. Phyllodes Tumor Of The Breast. In: StatPearls. StatPearls Publishing. Copyright. 2021, StatPearls Publishing LLC., Treasure Island (FL),

11. Jing P, Wei B, Yang X. Phyllodes tumor of the breast with nipple discharge: A case report. Medicine (Baltimore). 2018; 97 (52): e13767. doi:10.1097/md.0000000000013767

12. Kalambo M, Adrada BE, Adeyefa MM, Krishnamurthy S, Hess K, Carkaci S, Whitman GJ. Phyllodes Tumor of the Breast: Ultrasound-Pathology Correlation. AJR Am J Roentgenol. 2018; 210 (4): W173-w179. doi:10.2214/ajr.17.18554

13. Tan H, Zhang S, Liu H, Peng W, Li R, Gu Y, Wang X, Mao J, Shen X. Imaging findings in phyllodes tumors of the breast. Eur J Radiol. 2012; 81 (1): e62-69. doi:10.1016/j.ejrad.2011.01.085

14. Mitus JW, Blecharz P, Jakubowicz J, Reinfuss M, Walasek T, Wysocki W. Phyllodes tumors of the breast. The treatment results for 340 patients from a single cancer centre. Breast. 2019; 43: 85-90. doi:10.1016/j.breast.2018.11.009

15. Gullett NP, Rizzo M, Johnstone PA. National surgical patterns of care for primary surgery and axillary staging of phyllodes tumors. Breast J. 2009; 15 (1): 41-44. doi:10.1111/j.1524-4741.2008.00669.x .