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ORIGINAL PAPER

Vascular lesions of the ulnar upper extremity are predominantly lymphatic malformations

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ABSTRACT

Introduction: Lymphatic malformations are a common vascular anomaly in the paediatric population. Although the most frequent location is the cervical region, they can appear in any location where lymphatic tissue is present. We are expanding the research of the locoregional distribution patterns of these lesions in the upper extremity previously published by our colleagues in Hospital La Paz in Madrid, Spain.

Material and methods: We reviewed the medical records, radiological images, and clinical photographs of 12 patients with upper extremity lymphatic malformation that were assisted in the University Hospital of Łódź, Poland, and compared them with a Spanish group from La Paz University Hospital, Madrid. The locoregional distribution of the lesions, treatment applied, and clinical course were assessed. Statistical analysis of the collected data was performed with STATA version 15.0 (StataCorp)

Results: Eleven of the 12 patients (92%) presented an ulnar distribution pattern of the lymphatic malformation, while one (8%) presented a radial distribution pattern. The patient with radial distribution pattern had the lesion located proximally in contrary to the Spanish one with the radial lesion located distally. One of the patients (8%) presented some complication associated with the lymphatic malformation, such as pain/haemorrhage. Ultrasound-guided sclerotherapy was performed in 8 patients (67%), whereas 3 of the patients (33%) underwent a surgical excision. No differences were found in clinical presentation or evolution between patients with ulnar and radial distribution.

Conclusions: Upper extremity lymphatic malformations characteristically follow an ulnar distribution pattern, although the radial territory may also be affected. When diagnosing a patient with vascular lesion on the ulnar side, the differential diagnosis can start with lymphatic malformation, while on the radial side with other pathologies. A detailed characterisation of these patterns may contribute to a better and earlier diagnosis of the pathology.

KEY WORDS:

ulnar, lymphatic, pattern, extremities, paediatric.

INTRODUCTION

Lymphatic malformations (LMs) are a common pathology in the paediatric population. Most of them follow a facial, cervical, or upper extremity distribution, although they can be found in almost all organs with lymphatic tissue, including the visceral region. Diagnosis is mainly clinical and radiological, with ultrasound being the recommended initial examination and magnetic resonance imaging being the gold standard to assess the extent of the lesion and the coexistence of associated pathology. Most cases are asymptomatic or have limited

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symptoms, such as intralesional bleeding or infection. Some studies found a clinical worsening in the pubertal stage, suggesting that hormonal change may affect the evolution of these malformations. Although the clinical management of these lesions is variable and must be individualised for each patient, evolutionary surveillance, ultrasound-guided sclerotherapy, and surgical excision of the malformation are the most common therapeutic options. The location of the lesion is one of the most important factors determining the treatment. Since some locations, such as the anterior cervical and the tracheobronchial sites, may be life-threatening, they require early invasive procedures, including a tracheostomy. The lymphatic malformations are a well-known pathology. In recent years, there has been a growing need for new diagnostic and therapeutic approaches to the topic of vascular lesions in children. Basic research in this area also needs to be developed. This is evidenced by the interesting work carried out by Arredondo Montero *et al*. at La Paz Hospital, Madrid, Spain [1]. Their study sheds light on fundamental issues related to the occurrence of LMs contributing to our understanding of the biology of these lesions. Building on their insights, this study aims to test the assumptions outlined in a distinctly different patient population in consultation with the author of the above publication. In collaboration with the author, we expand their group with 12 Polish patients adding new information about the locoregional distribution patterns of LMs in the upper extremity from a separate, unique cohort.

While most studies are based on small groups, it is necessary to expand their scope to include an increasingly broad demographic and to obtain results from multiple centres. Our study delves into the unique characteristics of the Polish patient population in the context of locoregional LM patterns. In doing so, we aim to uncover potential differences that may exist in this diverse cohort.

By taking this approach, we hope to enrich existing knowledge of LMs and pave the way for more targeted and personalised interventions in clinical practice.

MATERIAL AND METHODS

We collected data on 12 Polish patients who were treated for malformations of the lymphatic system in the upper limb at the Department of Paediatric Surgery at the Central Clinical Hospital in Łódź 2020–2023. The usual methodological and ethical principles for scientific publications were followed. Photographs, clinical data, sociodemographic information, and radiological images were collected from the patients' medical records, and the information was anonymised in accordance with the applicable regulations (Figures 4, 5). Informed consent for publication was obtained from the parents of all patients whose images were included in this publication. Statistical analysis of the collected data was performed with STATA, version 15.0 (StataCorp). We used mean and standard deviation for quantitative variables and percentages for categorical variables. We compared sociodemographic and clinical variables between patients with ulnar and patients with radial distribution of the lymphatic malformation. We used Fisher's exact test to compare categorical variables. Statistical significance was defined as a p-value below 0.05 (2-sided).

RESULTS

Twelve Polish patients were attended for a lymphatic malformation in their upper extremity 2020–2023. The main clinical and sociodemographic characteristics of these patients are shown in Table 1. There was no impairment of limb function. Eleven of the 12 patients in this series (92%) presented an ulnar distribution pattern of the lymphatic malformation, while one (8%) presented a radial distribution pattern. The patient with radial distribution pattern had the lesion proximally located (arm and proximal radius). Similarly, in all the patients with ulnar distribution the pattern of the lesion was proximal – mostly close to the ulnar joint. None of the patients had a relevant medical-surgical history. One of the patients (8%) presented some complications associated with the lymphatic malformation, such as pain/haemorrhage. Ultrasound-guided sclerotherapy was performed in 8 patients (67%), whereas 3 of the patients (33%) underwent a surgical excision of the lesion. One of the patients was included in the sirolimus treatment program because of early rapid progression of the lesion resulting in inhibiting growth of the lesion. The family of 3 patients (33%) refused interventional treatment. One patient (8%) who had been treated with surgery presented clinical recurrence and required surgical reintervention. None of the patients presented relevant complications during the follow-up of 2–4 years. We did not find statistically significant differences in any clinical or sociodemographic characteristic between patients with ulnar or radial distribution pattern of the malformation. We compared the results with the work of the Spanish group and analysed the 2 groups together. We present the results in Figure 1. The following conclusions and differences were highlighted.

There was a significant occurrence of LMs on the ulnar side in both groups, which was also confirmed by the combined percentage of both populations. Lymphatic malformations were more frequently qualified for primary surgery in the Spanish centre, with a complication rate of less than 25%, and with a lower complication rate among the Polish population. In the Polish population, qualification for sclerotisation prevailed, at 67% to 17% (mean 41.67%). Reported recurrences were one patient in the Polish population (8%) and 2 patients in the Spanish population (17%) (mean 12.5%) (Figure 2).

Only the comparison of age and malformation occurrence site of the total population showed significance with a p-value below 0.05. It is possible that this was caused by age deviation caused by individual older patients included in the group (Figure 3).

TABLE 1. The main clinical and sociodemographic characteristics of the patients – Polish group

Patient No	Age	Sex	Lymphatic malformations localisation	Malformation type	Radiological pattern	Treatment	Observation years/relapse
1	5 years	Female	Left forearm – ulnar	Lymphatic	Microcystic	Excision	3 years/relapse after 2 years
2	2 weeks	Male	Right forearm – ulnar	Lymphatic	Microcystic	Not yet treated	–
3	2 month	Male	Left arm and forearm – ulnar	Lymphatic	Macrocystic/mixed	Sclerotisation, excision	2.5 years
4	11 years	Male	Left arm and ulnar	Veno-lymphatic	Veno-lymphatic	Sclerotization	10 years
5	14 years	Female	Left arm and ulnar	Veno-lymphatic	Veno-lymphatic	Sclerotization	2 years
6	16 years	Female	Right ulnar	Lymphatic	Macrocystic	Sclerotisation	4 years
7	14 years	Male	Right ulnar	Lymphatic	Macrocystic	Sclerotisation	2 years
8	1 weeks	Male	Left arm and forearm – radial	Lymphatic	Mixed	Sclerotisation, excision	2.5 years
9	1.5 years	Female	Right ulnar	Lymphatic	Mixed	Not yet treated	–
10	2.5 years	Female	Right ulnar	Lymphatic	Macrocystic	Sclerotisation	1.5 years
11	2 month	Female	Left and arm – ulnar	Lymphatic	Mixed	Sclerotisation, rapamycin	2 years
12	5 month	Male	Right forearm - ulnar	Lymphatic	Macrocystic	Not yet treated	–

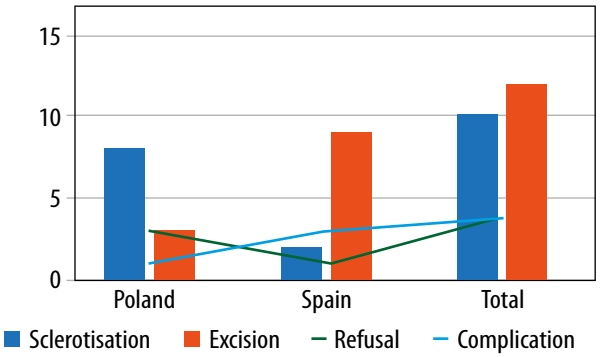

FIGURE 1. Comparison of treatment in the Polish and Spanish patient groups – number of patients

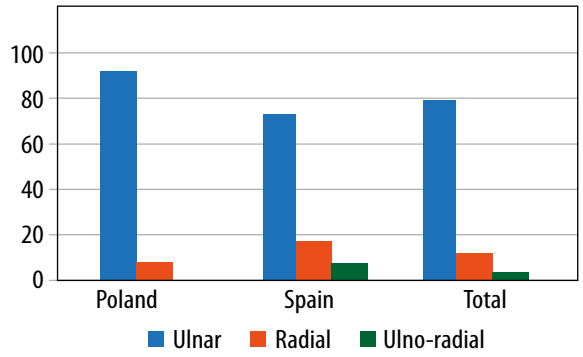

FIGURE 2. Upper limb distribution pattern of lymphatic malformations (%)

DISCUSSION

The locoregional distribution patterns of lymphatic vessels and malformations have gathered significant interest due to their clinical relevance and diagnostic implications.

Articles studying the anatomy and function of the lymphatic system are continually being written worldwide and successfully emphasise the importance of this issue [1–4].

The diagnosis and treatment paths of LMs is in the process of being standardised. At this point, it varies widely depending on the specific case and department. Many lesions are diagnosed on the basis of characteristic history, location, and USG. Improving knowledge of the anatomy and frequency of lesions can significantly facilitate diagnosis and improve treatment outcomes of patients affected by this disease.

These anomalies, especially those confined to upper extremities, are very rare. Studies with larger sample sizes would help to validate previous findings and contribute to a better knowledge of LMs in the upper extremity in diagnostic-therapeutic terms.

Our study expands on a small sample size from La Paz Hospital in Spain, bearing in mind that there may be

a great between-individual variability in upper extremity lymphatic network conformation [1].

Despite the expanded group of patients, the small sample size remains an important limitation of our study. Therefore, interpretation of the results must be made with caution. Lymphatic lesions are highly variable. With this in mind, our results still have the potential to improve clinical practice and guide future research in this area.

By analysing data from a cohort of patients treated for LMs in the upper limb, we aimed to uncover distribution patterns that may help in therapeutic decision-making. The Polish cohort of 12 patients were compared and combined with the Spanish group of 12 patients [1], confirming a notable occurrence of ulnar distribution patterns among upper extremity lymphatic malformations. Specifically, 92% of Polish patients exhibited ulnar distribution, with proximal lesions predominantly observed near the ulnar joint and proximal part of the forearm. One patient (8%) presented a radial distribution pattern. The patient with radial distribution pattern had the lesion located proximally, in contrary to the Spanish one (8%) with the radial lesion located distally. Considering both groups with a total of 24 patients, the ulnar distribution pattern was predominant in 84% of patients. This observation aligns with previous studies indicating a predilection for ulnar involvement in LMs of the upper extremity [1]. While it would be beneficial to further expand the group of our cohort, we can already confirm that when diagnosing lesions of the ulnar side of the upper limb the LMs will be a common finding, usually easily confirmed by ultrasound examination.

Comparison with data from other medical centres provides valuable insights into potential variations in clinical management and outcomes across different patient populations.

The significant occurrence of ulnar involvement in both cohorts underscores the reproducibility of our findings across diverse patient populations. Notably, while the Spanish cohort exhibited a higher rate of primary surgical interventions, ultrasound-guided sclerotherapy prevailed in the Polish cohort, suggesting potential variations in treatment approaches between centres.

Variations in treatment approach and complication rates underscore the need for individualised management strategies tailored to the specific needs of each patient [5, 6]. In recent years, increasingly performed genetic research and discovered gene relations, as well as the introduction of modern systemic therapies, have contributed greatly to progress in the treatment of vascular malformations. For several years, our department has been running a sirolimus treatment program. One of the patients included in our study was started on rapamycin treatment. This decision was made due to the rapid progression of the disease in the first weeks of follow-up despite sclerotherapy. After the drug was introduced empirically, disease stabilisation was achieved. There is no impairment of limb function for this and other cases. The patient remains under observation, and further procedures are planned, including genetic testing [7].

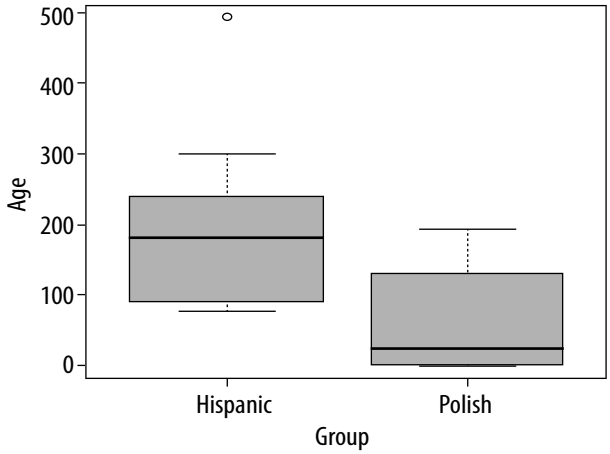

FIGURE 3. Comparison of age and malformation occurrence site of total population

Despite the similarities in the 2 groups regarding the distribution and type of malformations in the study, the differences between chosen treatment methods are noteworthy. Although our work does not focus on the treatment of lymphatic malformations, it is worth noting that the treatment of Polish patients included in the study began relatively recently, while in the Spanish group the patients were admitted years earlier. This may indicate that standards of management are becoming more established and that there is a shift towards minimally invasive procedures despite similar success rates [6, 8]. These trends would probably be more pronounced on a larger group of subjects.

Complications after procedures vary, of course, although few are directly related to the type of procedure performed. These range from rare complications like bleeding, mainly occurring in excisions, to less troublesome but more frequent complications such as swelling and pain. It is difficult to assess retrospectively the overall severity and frequency of the mentioned complications with such a small and heterogeneous group of patients [8].

While we analysed the data from the 2 centres, it was also important to consider the age differences of the 2 groups. Deviations in the Spanish group in the form of single, much older patients may distort the results in terms of recurrence or treatment modality and outcome.

CONCLUSIONS

Clinically, our findings have important implications for diagnostic approaches and patient management strategies. We believe that extending the number of patients studied and the international and multicentre nature of the study deepens existing knowledge of the distribution of lymphatic malformations. Recognition of ulnar distribution patterns should prompt consideration of LMs

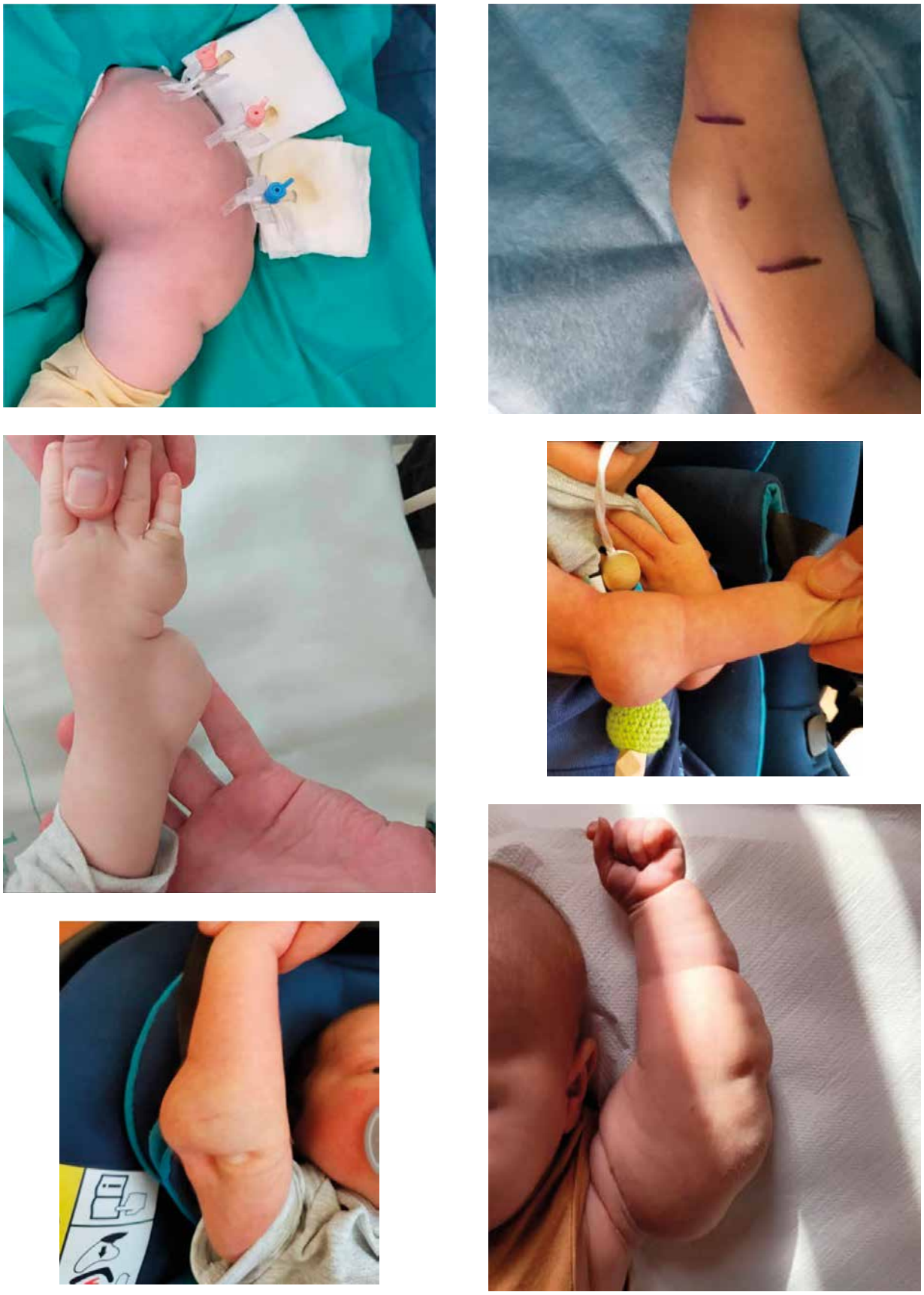

FIGURE 4. Clinical photographs of patients affected by lymphatic malformations of the upper extremity with ulnar distribution pattern

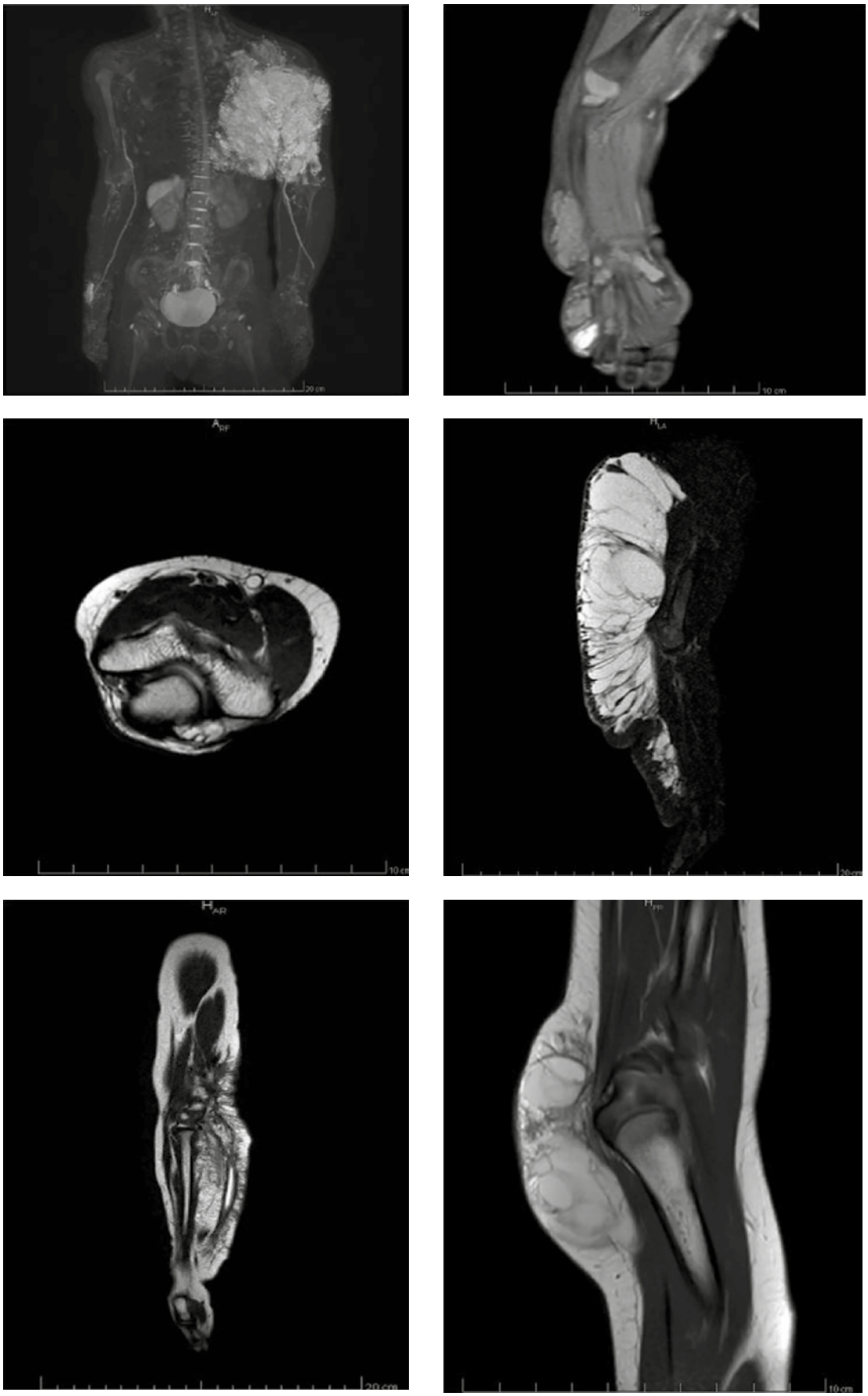

FIGURE 5. Radiological imagining of patients affected by lymphatic malformations of the upper extremity with ulnar distribution pattern (magnetic resonance imaging)

in the differential diagnosis of upper extremity tumours in paediatric patients. This underscores the importance of early diagnosis and the use of complementary imaging modalities such as ultrasound to confirm suspicions and guide appropriate treatment decisions. Lymphatic lesions should not be considered first in the differential diagnosis of radially distributed vascular lesions. Occasional malformations of both radial and ulnar portions are extremely rare, occurring in a total of less than 5% of the patient population [3, 5].

We underscore the importance of individualised management approaches and the need for further research to elucidate the underlying mechanisms driving these distribution patterns.

DISCLOSURES

1. Institutional review board statement: Not applicable.
2. Assistance with the article: None.
3. Financial support and sponsorship: None.
4. Conflicts of interest: None.

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