

VASCULAR ANOMALIES IN CHILDREN – 17 YEARS EXPERIENCE

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Abstract

Objective: The aim of this study is to assess our results of the treatment in children with vascular anomalies.

Methods: We retrospectively reviewed the charts of 704 patients with 843 vascular anomalies treated in the Department of Pediatric Surgery between January 1996 and June 2012.

Results: Sex ratio was females/males 1.5:1, 69% were less than one year of age at the time of admission. The average time of hospital stay was 2 days. 83.5% of vascular anomalies were hemangiomas and 83,6 % of these affected the skin and subcutaneous tissue. 64% of hemangiomas were tuberous and most were located on the head and neck. 11% affected multiple sites and/or were associated with other vascular anomalies.

Conclusion: This audit allows us to improve the diagnosis, methods of treatment and improving our clinic's recording. Our results stress on the necessity for implementation of educational programs among the people living in the countryside, so that they would become aware of the importance of addressing the physician for their children's health concerns.

Key words: vascular anomalies, vascular tumors, vascular malformations, hemangiomas.

Introduction

In 1996, the International Society for the Study of Vascular Anomalies (ISSVA) accepted the classification of vascular anomalies into vascular tumors (the most common

of which is infantile hemangioma) and vascular malformations, based on clinical, radiological and pathological characteristics [1,2]. The differentiation was founded on the idea that the suffix "oma" (as in "angioma") implies a tumor that proliferates, which does not apply to vascular malformation, as they are non-proliferative [3]. According to contemporary statistical data, vascular anomalies are on the rising, affecting on average 10% of all infants; also, they are believed to account for about 45% of the tumors of skin and soft tissues [4]. It is believed that hemangiomas are the result of excess angiogenesis while vascular malformations are caused by errors in vessel remodeling [5]. The most frequent localizations of vascular anomalies are the teguments from the cervico- facial area; however, they can affect any organ system, be it parenchymatous or cavitory [4].

Material and methods

This study includes 704 patients (1 day to 18 years of age) with 843 vascular anomalies, who were admitted and treated in our clinic during the January 1996 – June 2012 time frame (Fig. 1). Patient data was obtained from hospital admission records, clinical observation forms, surgical records, imagistic studies and histopathologic exams. Since 2008 electronic records were also used. The analysis employed looked at demographic and anamnestic data, therapeutic methods, clinical evolution, treatment results and complications.

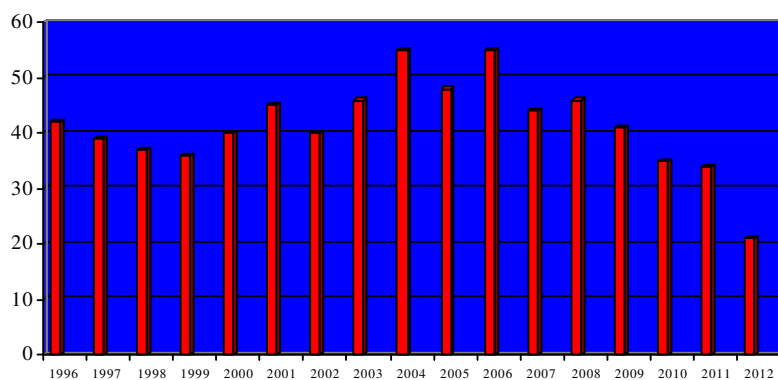


Fig. 1. Number of patients by year of admission.

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The Emergency Children's Hospital "Louis Țurcanu" serves the Western part of Romania, specifically the Timiș, Caraș Severin, Hunedoara and Arad counties. However, patients in this study came from other parts of the country as well, especially the Mehedinți, Gorj, Bihor, Botoșani, Suceava, Vaslui and Vrancea counties.

Results

Of the 704 patients included in the study, 427 are girls and 277 are boys, with an approximate female:male ratio of

1.5:1. 487 (69%) patients were less than one year of age at the time of admission. Table 1 and Figure 2 summarize patient distribution by age. The majority of patients (65%) came from the urban environment. 69% were less than one year of age at the time of admission. The number of days of admission varied between 1–28 days, with an average of 2 days. Only 6 patients stayed in the hospital for more than 15 days.

Table 1: Patient distribution by age.

| Age | No. Patients |
|---------|--------------|
| 0-2 mo | 44 |
| 2-6 mo | 236 |
| 6-12 mo | 207 |
| 1-3 yrs | 137 |
| 3-7 yrs | 28 |
| >7 yrs | 52 |

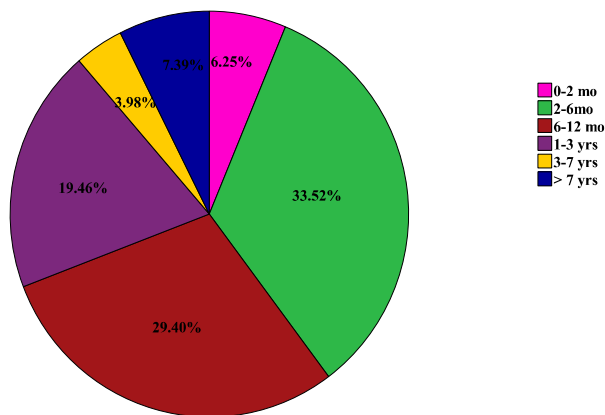


Fig. 2. Patient distribution by age.

The prevalence of vascular anomalies was as follows: hemangiomas (83.5%), lymphangiomas (4%) and "Others" (12.64%) (Fig.3). The "Others" category includes 89 patients with vascular malformations (mostly Klippel-

Trenaunay Syndrome, Sturge-Weber Syndrome and peripheral arteriovenous malformations) but also vascular tumors (Kaposiform hemangioendothelioma, congenital hemangiomas and glomangiomas).

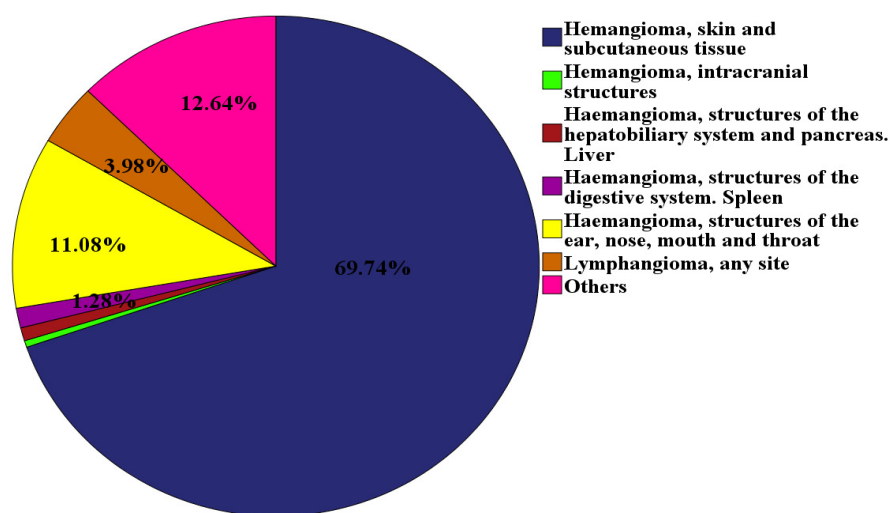


Fig. 3. Distribution of patients with vascular anomalies by diagnostic.

By far, the greatest percentage of hemangiomas affected the head and neck area (51%), followed by the trunk (15%). 11% of patients had multiple hemangiomas,

with or without other associated vascular anomalies (capillary malformations in most cases) (Fig. 4). Female: male distribution of hemangiomas was in a ratio of 2:1.

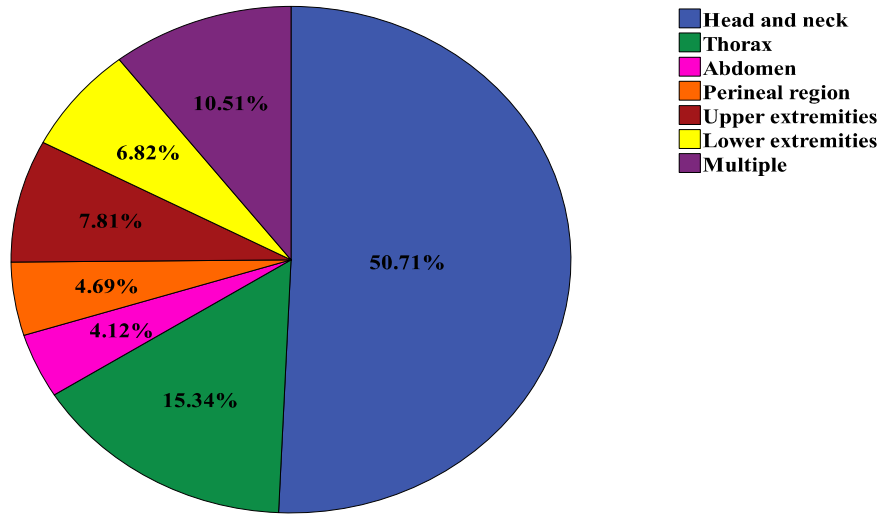


Fig. 4. Hemangioma distribution by anatomical location.

Of the 587 patients with hemangiomas, 376 (63.6%) were tuberous, 83 (14.2%) cavernous, 79 (13.6%) unspecified and 49 (8.4%) suffered complications such as bleeding, infections, and/or ulceration. Out of the 704

patients treated solely in our clinic, 88.15% required therapeutic intervention and in most cases, lesion excision was performed (Fig 5).

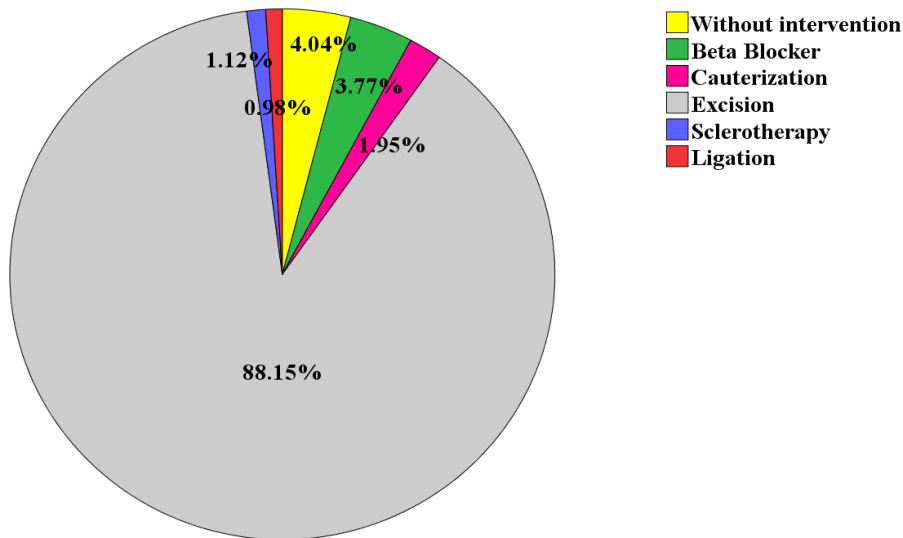


Fig. 5. Types of interventions employed.

In 4.4% of patients, especially those with giant hemangiomas, up to 5 surgical interventions were required for complete excision. In 11 cases of large-sized hemangiomas, reconstructive surgery with skin grafting was performed. In 8 cases of hemangiomas located in regions

that made it difficult for surgical treatment to be performed, sclerosing agents were injected (Bleomycin or Aethoxysklerol), together with peri and intralesional ligation. A single lymphangioma case required intralesional injection with Picibanil (OK-432); the posttherapeutic result

in this case was very good. In May 2010, our clinic introduced treatment with beta blocker (propranolol) for patients with infantile hemangiomas; 3.77% of the patients with vascular anomalies have so far benefited from such treatment, with very good results.

Discussions

The separation of vascular anomalies into vascular tumors and vascular malformations has been of key importance in easing the ability of clinicians to diagnose and treat these entities [6]. Vascular tumors are believed to mostly arise sporadically, but an Autosomal Dominant (AD) mode of inheritance has been reported by Blei et al, the genetic defect affecting the 5q 31–33 chromosome [7]. Infantile hemangiomas usually appear within a few days after birth, exhibit an accelerated rate of growth during the infant's first year of life and then undergo regression by the age of 7 [8]. The incidence of infantile hemangiomas during the first 3 days of life ranges from 1.1% - 2.6% and it rises to 8.7% - 12.7% during the first month – one year interval [9]. In our study a number of 280 (47,7%) patients with infantile hemangiomas were diagnosed before six months of life; our results are consistent with literature data. Most hemangiomas (51%) affected the head and neck area; the thorax was involved in 15% of cases, with multiple areas being affected in 10.5% of cases like in Haggstrom observation.[10]

Vascular malformations are almost always present at birth and differ from hemangiomas based on clinical appearance, histopathologic features, and biologic behavior [11]. In rare cases, a patient can present both vascular tumors and vascular malformations. Literature has reported on the copresence of a hemangioma and a capillary malformation of the port-wine stain type at either the same site or in close proximity [6].

In other instances, however, if these two lesions are located at distant sites, their association is coincidental [6]. Due to insufficient reporting, we are unable to determine what percentage of our patients fit either one of the situations above; however, we know that 11% of had multiple hemangiomas, with or without other associated vascular anomalies (capillary malformations in most cases).

While literature presents a female:male ratio for infantile hemangiomas anywhere from 3-9:1, the ratio of patients in our clinic was lower (2:1). The reason for female predominance is unknown but it is believed to be linked to hormonal differences [12]. As most cases referred to our surgery clinic were of a more severe nature, the type of treatment most commonly employed for vascular anomalies

was surgery, with very good results. Annual distribution of patients suggests the decreasing number of cases in our clinic, could be explained by increasingly administration of beta blocker. Though propranolol treatment was administered on a relatively small sample size (28 patients) its very good results in treating infantile hemangiomas make the drug a likely, less invasive choice in the future management of these lesions. That pattern of evolution is specific for a surgery department were almost all (88,15%) cases were surgically treated due to their complications-bleeding, infection and ulceration.

70% of our patients presented hemangioma on the skin of subcutaneous tissue, evidence not described by previous authors. Although the incidence of vascular malformations is known to be reduced in surgical wards, we find 89 cases having complex syndromes, even if they are generally reported from medical departments.

According to the National Institute of Statistics, as of January 1, 2010, the urban/rural distribution of population in Romania was 55% and 45% respectively [13]. However, only 35% of the patient population came from the rural environment. As the rate of vascular anomalies is uniform throughout the population, the significant discrepancy in the patients from the urban versus rural environment is suggestive of the fact that people living in the countryside are less willing to address the physician in matters of their children's health. We would like to therefore emphasize the importance of implementation of educational programs, such that awareness is increased on the significance of health checkups in growing children.

While initially mostly presenting an aesthetic problem, vascular anomalies can lead to problems such as: ulceration, hemorrhage, functional impairment, visual/auditory obstruction, and respiratory/cardiac insufficiency or Kasabach-Merritt syndrome.

Conclusions

It is essential for the clinician to differentiate between vascular tumors and vascular malformations, in order to implement a better management. The lower percentage of patients from the rural environment is suggestive of the fact that parents from the countryside are not as likely to bring their children to the doctor as parents living in urban areas. Hence, increased awareness on the importance of regular checkups is needed. Given the excellent results that propranolol has brought in treating hemangiomas, we feel confident to make increasing use of this form of treatment. If medical treatment fails, patients will benefit from surgical procedures.

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