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Pseudohemangioma of the head - diagnostic difficulties in a pediatric patient with Ewing's sarcoma of the scalp: case report

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Abstract

Introduction: Ewing’s sarcoma (ES) is the second most common malignant bone tumor of childhood and adolescence. Typically, ES affects the long bones, pelvis and the trunk. Primary location in the scalp is rare.

Aim of study: The aim of our study is to focus on the case of a pediatric patient with Ewing's sarcoma located in the scalp, which adopted a cavernous angioma mask, and to assess the available knowledge on the diagnosis and treatment of the extracostal ES located in the scalp.

Material and methods: The research method was a case study. The research material was the patient's medical record, and the technique used was an analysis. An unsystematic review of Polish and English-language scientific literature was conducted. Electronic databases: PubMed, SCOPUS and Google Scholar were searched using the keywords: sarcoma, Ewing, hemangioma, pseudohemangioma, the head, the scalp.

Results: When the girl was 6 months old, a tubercle appeared on her head. It was asymptomatic, but gradually increased. Imaging studies suggested the diagnosis of cavernous hemangioma. The tumor did not respond to treatment and still was growing. A decision was made to include resection. The upward tumor underwent histopathology with immunohistochemical assessment and genetic testing. The result pointed to extraskeletal Ewing’s sarcoma.

Conclusions: Despite the few cases described in the literature, remember about atypical locations of primary Ewing sarcoma such as scalp. Making the right diagnosis was a challenge for the multi-specialist team of doctors. Histopathological examination supplemented by immunohistochemical assessment and genetic testing gave a definitive diagnosis and enabled proper treatment.

Key words: sarcoma; Ewing; hemangioma; pseudohemangioma; the head; the scalp

Legend:

Angio-CT – Angio-Computed Tomography
CK AE 1/3 – Cytoceratin AE1/AE3
CNS - Central Nervous System
CT - Computed Tomography
EES – Extraskeletal Ewing’s Sarcoma
ES – Ewing’s Sarcoma
ESFT – Ewing’s Sarcoma Family of Tumor
ESHN – Ewing’s Sarcoma of Head and Neck
ESR - Erythrocyte Sedimentation Rate
EWSR1 – Ewing’s Sarcoma Breakpoint Region 1
FISH - Fluorescence In Situ Hybridization
FLI1- gene of Friend Leukemia Integration 1 transcription factor
LCA - Leucocyte Common Antigen

MRI - Magnetic Resonance Imaging
NSE - Neuron-Specific Enolase
PAS - Periodic Acid-Schiff
PAS-D - Periodic Acid-Schiff Diastase
PET - Positron Emission Tomography
PNET - Peripheral Primitive Neuroectodermal Tumor
USG - Ultrasonography
VIDE - Vincristine, Ifosfamide, Doxorubicin, Etoposide

I. Introduction

I.I General information

Ewing's sarcoma (ES) is the second most common - after osteosarcoma - primary bone cancer in children and young adults [1, 2, 3, 4, 5, 6, 7, 8, 9, 10]. Most often it is diagnosed in patients up to the age of 20 and the age median is 15 years [2, 6, 7, 9, 11]. The incidence is comparable in both genders, although some authors emphasize that boys show a higher one than girls (1.5-1.6:1) [1, 2, 5, 6, 7, 9]. What is more, scientists noted a higher incidence in the white than the African race individuals [1, 5, 6, 10, 12, 13].

I.II Ewing Sarcoma Family of Tumors

Ewing's sarcoma and primitive neuroectodermal tumors (PNETs) belong to the Ewing's sarcoma family of tumors (ESFT), which is characterized by specific molecular features. The most common is the t(11; 22) (q24; q12) translocation which involves the EWSR and FLI1 genes [2, 3, 6, 7, 8, 9, 11, 12, 13, 14, 15]. Abnormalities in the EWSR1 gene region can be detected by fluorescent in situ hybridization (FISH), which is used in diagnostics [2, 7]. EWSR1-FLI1 as a fusion protein has the properties of an abnormal transcription factor and is involved in the oncogenesis process [3, 6, 7, 11, 12, 14]. In the microscopic image, small, round, primitive cells with blue staining and glycogen-rich cytoplasm are noteworthy. The formation of structures called Homer-Wright rosettes characteristic of neural differentiation is less expressed in ES than in the related PNETs [1, 4, 6, 7, 8, 9, 11, 12, 14, 15, 16].

I.III Location

Two groups of ES can be distinguished in literature: skeletal and extraskeletal (EES). Typically, Ewing's sarcoma affects the diaphysis and metaphysis of the long bones of the upper and lower limbs as well as the flat bones of the pelvis and the trunk [5, 6, 7, 9, 10, 15, 17]. The location in the head and neck (Ewing's sarcoma of the head and neck - ES-HN) accounts for little percentage of all ES (1-9%) cases. ES-HN is most commonly found in the cervical spine, skull, mandible and paranasal sinuses, the scalp location is very rare [2, 5, 6, 8, 15, 16, 17, 18, 19, 20]. The first case of a 15-year-old boy with primary ES in the scalp was described by Suster et al. in 1988. Up to now, only a few cases of ES located in the scalp have been presented in literature as

the primary and secondary manifestation of this carcinoma. Haas et al. presented a case report of the innate ES of the scalp [2, 5, 15, 18, 19, 20].

ES is an aggressive carcinoma that quickly metastasizes. In the case of ES-HN, this is rarer compared to ES in other locations and the prognosis is better [1, 2, 5, 6, 8, 11, 12, 17, 21]. ES metastases are most often located in the lungs, pleura, other bones, liver and brain, and secondary affecting of the scalp is extremely rare [20]. Ewing's sarcoma tends to recur and recurs most often within 2 years [1, 6, 12]. For a localized disease, when adequate treatment is implemented, the 5-year overall survival is estimated at 65-87%. In a patient with distant metastases present at the time of diagnosis, the overall survival is about 30% [1, 2, 4, 6, 8, 14, 16, 17].

I.IV Diagnostics

The basis of ES diagnostics is the subjective and physical examination. The nonspecific symptoms that accompany this carcinoma include increasing pain, local edema and lymphadenopathy. Occasionally, inflamed and tender on palpation tissue thickening can be noticed. Fever, a significant weight loss in a short time, night sweats or fatigue can be alarming signals [2, 4, 6, 7, 8, 9, 10, 17, 19, 21]. Blood laboratory tests such as complete blood count with smear, erythrocyte sedimentation rate (ESR) or lactate dehydrogenase levels are simple tools that support diagnostics [1, 2, 4, 6, 10]. Imaging tests ordered in case of suspected ES and metastasis assessment include x-rays, computed tomography (CT) with or without intravenous contrast administration, magnetic resonance imaging (MRI) and positron emission tomography (PET scan). Bone biopsy and marrow punch biopsy are also performed to determine the disease progression [6, 8]. In imaging studies, extracostal ES has a slightly different form from a skeletal one because it does not directly affect the bone. It usually presents as a heterogeneous soft tissue mass. Hemorrhagic areas, cystic structures and necrosis areas are often present. ES can infiltrate surrounding tissues, leading to destruction [7, 8, 9, 10]. Literature emphasizes its heterogeneous enhancement in CT and MRI imaging after contrast administration [9].

There is little data on the ES-like features of cavernous hemangioma in imaging studies. Scientists present the case of ES of the sciatic nerve based on MRI as an oval, well limited lesion damaging the nerve, fed by a deep thigh vessel, which is enhanced after contrast administration [4]. In the case of pseudohemangioma of the spine, literature emphasizes benign, increased vascularization, with staining of the nerve opening at the height of the tumor [9]. There is a known case of Ewing's sarcoma of the back skin resembling an angiomatic tumor, which in dermoscopy appeared as multiple lobular red structures separated by fissures with a whitish veil [22].

I.V Treatment

Treatment of Ewing's sarcoma is multimodal. Intensive, multi-drug chemotherapy, surgery and radiotherapy are used [1, 2, 4, 6, 10, 11, 14, 16, 17, 21, 22]. Most of ESs respond well to chemotherapy. It is introduced before a surgical intervention to reduce tumor mass, but

also after a resection as adjuvant treatment [4, 6, 8, 14, 16]. In most cases, the patient undergoes 14-17 cycles of chemotherapy. Vincristine, Ifosfamide, Cyclophosphamide, Doxorubicin, Etoposide and Actinomycin B are most commonly used in various combinations. Some studies emphasize the importance of Dacarbazine and Dactinomycin. A surgery (a complete or partial resection of the tumor) and / or radiation therapy are to provide local control. When talking about the head and neck ES, local control may not always be aggressive due to a high risk of damage to adjacent nerve structures. This carries considerable morbidity [2, 6, 8, 11, 14, 16, 21]. Radiotherapy combined with chemotherapy are often reserved for inoperable cases or patients with residual disease [1, 2, 4, 6, 8, 14].

II. Aim of study

The aim of our study is to focus on the rare case of a pediatric patient with Ewing's sarcoma located in the scalp, which adopted a cavernous angioma mask, and to assess the available knowledge on the diagnosis and treatment of the extracostal ES located in the scalp.

III. Methods and materials

The research method was a case study. The research material was the patient's medical record, and the technique used was an analysis. An unsystematic review of Polish and English-language scientific literature was conducted. Electronic databases: PubMed, SCOPUS and Google Scholar were searched using the keywords: sarcoma, Ewing, hemangioma, pseudohemangioma, the head, the scalp.

IV. Findings – a case report

We present a case report of a 17-month-old Caucasian girl with suspected Ewing sarcoma of the scalp admitted to the Clinic of Hematology, Oncology and Pediatric Transplantation to make the final diagnosis, determine the stage of the disease and implement appropriate treatment.

In February 2018, the parents of the six-month-old girl observed a lump in the parietal region on their daughter's head. Since August 2018, the lesion was growing rapidly, prompting her parents to consult doctors - first a pediatrician, and then a surgeon.

A Doppler ultrasound (USG) examination confirmed subcutaneous location of the nodule in the median line of the head, its smooth contours and dimensions of 20x10 mm. The presence of single calcifications, vascular flow traits and heterogeneous echogenicity were described. The lesion did not infiltrate the surrounding tissues.

Based on the clinical picture, a Doppler ultrasound (USG) and magnetic resonance imaging (MRI) examinations of the central nervous system (CNS) performed in October 2018, cavernous hemangioma was diagnosed. Following this diagnosis, it was decided to treat the vascular lesion conservatively, and Propranolol was recommended to accelerate the reduction of tumor mass. Due to a congenital heart defect, the child was under strict medical supervision of

the Cardiology Unit while receiving the medication, where no contraindications for it were found. Unfortunately, parents administered the medication irregularly and the change increased.

In January 2019, an angio-CT scanning with contrast was performed to assess arterial and venous vessels, and a neurosurgical consultation was commissioned. The extracranial location of the tumor at the frontoparietal border in the midline and on the left side was confirmed. This change underwent strong, heterogeneous contrast enhancement and clinically corresponded to hemangioma. The connection to the central nervous system (CNS) vasculature was excluded. The tumor had strong vascularization from the common carotid artery.

At the end of January 2019, the girl was qualified for an elective total resection of the extracranial tumor under general anesthesia. The collected material was sent for histopathological examination. The macroscopically removed tissue was cohesive, livid grey, 4.2x4.2x3.5 cm, covered with macroscopically unchanged skin flap. In the cross-sections there were numerous cystic spaces filled with blood. The surgical margin was a pseudocapsule. The microscopic image of the removed lesion indicated a monomorphic, microcelllular texture of the tumor located subcutaneously, with poorly expressed formation of rosette structures full of hemorrhages.

Immunohistochemistry revealed CD99 (diffuse +), Vimentin (diffuse +), S-100 (focal +), CK AE 1/3 (-), LCA (-/+), FLI-1 (+), Synaphtophisin (focal +) , CD56 (- / trace +), CD3 (-), CD79a (-), Myogenin (-), Desmin (-) reaction. PAS and PAS-D (+) staining confirmed the presence of glycogen in the tested material. Summing up, the micro- and macroscopic image and the results of the immunohistochemical examination indicated hyperplasia characteristic of Ewing’s Sarcoma Family of Tumors.

The FISH genetic test was commissioned, where after counting 100 cells, more than 30% of the obtained ones bore the present rearrangements of the EWSR1 gene (22, q12.2) with t(11.22) (q24, q12) or t(21.22) (q22, q12) translocation. After verification of the material by an independent pathomorphologist, the final diagnosis of Ewing’s sarcoma with a present rearrangement of the EWSR1 gene was made.

The girl went to the Clinic of Hematology, Oncology and Pediatric Transplantation almost thirteen months after the first symptoms appeared. She was qualified for a treatment in the EWING2008 protocol. The patient underwent all the tests recommended in the Euro EWING protocol. Based on laboratory, imaging (MRI, CT of the chest, abdominal ultrasound), biopsy and marrow punch biopsy, no tumor metastases were found.

In early March 2019, the patient being in a good general condition received 1 cycle of VIDE. She tolerated the treatment well. No side effects were observed. After the first cycle of chemotherapy, the administration of Filgrastim was started.

V. Discussion

Ewing's sarcoma is an aggressive, malignant tumor of bones and soft tissues in children and young adults [1, 2, 3, 4, 5, 6, 7, 8, 9, 10]. The disease is most often detected in patients aged

10-20 years (the age median is 15 years). The occurrence in children under 10 years of age is rarely reported [2, 6, 7, 9, 11]. The girl, whose patient's history we discussed, was 6 months old when the first symptoms appeared. In the case of the ES scalp described by Chao et al. the boy was 11 years old, and in the study of Suster et al. the age was 15 years [15, 19]. ES slightly more often affects young boys than girls, and of the Caucasian race rather than those of an African origin [1, 2, 5, 6, 7, 9, 10, 12, 13].

Ewing's sarcoma usually affects the diaphysis of the long bones the limbs as well as the flat bones of the pelvis and the trunk bones [5, 6, 7, 9, 10, 15, 17]. The scalp becomes hardly ever affected by ES. Haas et al. described the case of an innate ES of the scalp [18]. In literature we met only a few case reports of primary ES in this location [15, 18, 19]. We collected them **in table no. 1 (Table 1).**

Tabele 1: Comparison of case reports of Ewing's sarcoma of the scalp.

No.	Case report Characteristic	Our case report	Suster et al.	Chao et al.
1.	**The age at which the first symptoms appeared**	6 months	15 years	11 years
2.	**Gender**	Female	Male	Male
3.	**First symptoms**	Rapidly growing lesion on the head covered with unchanged skin	Fast-growing head tumor covered with smooth skin	Lymphadenopathy, head and neck pain
4.	**Scalp location of the tumor**	Parietal region, midline and left side	Parietal region, on the right side	No data
5.	**Tumor size**	4,2x4,2x3,5 cm	3.5 cm in diameter	
6.	**Features in imaging studies**	Angio-CT: strong, heterogeneous contrast enhancement	X-ray: small, localized irregularity on the outer plaque below the lying parietal bone without invading the bone	
7.	**Macroscopic features of the tumor**	Cohesive, gray-blue mass, covered macroscopically with unchanged skin flap, numerous cross-sections of cystic blood-filled cysts, surrounded by a pseudocapsule	Lobular arrangement of the tumor cells, which are occasionally separated by broad collagen bands and show focal areas of hemorrhage and necrosis	
8.	**Microscopic features of the tumor**	Monomorphic, small-cell tumor weaving, with poorly expressed formation of rosette structures, rich in hemorrhages. PAS and PAS-D (+).	Fairly uniform, hyperchromatic, round to oval, compactly arranged cells, PAS (+)	
9.	**Immunohistochemistry**	CD99 (diffuse +), Vimentin (diffuse +), S-100 (focal +), Cytokeratin AE 1/3 (-), LCA (-/+), FLI-1 (+), Synaphtophisin (focal +), CD56 (-/+), CD3 (-), CD79a (-), Myogenin (-), Desmin (-).	Vimentin (+), LCA (-), NSE (-), Cytokeratin (-), Desmin (-)	
10.	**Chemotherapy**	VIDE – Vincristine, Ifosfamide, Cyclophosphamide, Doxorubicin, Etoposide	No	Yes
11.	**Tumor resection**	Yes	Wide tumor resection	Yes
12.	**Radiotherapy**	No	No	2000 cGy

Ewing's sarcoma tends to metastasize quickly. It is reported that for the location of the primary ES in the head and neck this frequency is less than for the primary ES in other locations. This is associated with a better prognosis for the patients affected by the former one [1, 2, 5, 6, 8, 11, 12,

17, 21]. Secondary ES location most often includes the lungs, pleura, other bones or liver. Turgut et al. point to rare scalp metastases [20]. There is little data to give basis for multi-specialist medical teams to deepen their knowledge, which would translate into faster diagnosis and the best adjustment of treatment to a specific patient. Up to the present, there have been no clear patterns developed for the management of a patient with suspected ES located in the scalp.

ES diagnostics is based on subjective and physical examination. Among the ES (primary and metastatic) symptoms of the scalp, which have been described in literature, the predominant ones are pain, local edema as well as the presence of tender and palpable tumor. Cases of lymphadenopathy, fever, weight loss and fatigue have been reported [15, 18, 19]. The study of Suster et al., who were the first to describe the primary ES of a scalp in a 15-year-old boy, indicated a rapid increase in the lesion of 3.5 cm in diameter, located in the right parietal area [15]. Chao et al. recalled the case of an 11-year-old boy with a scalp tumor, in whom the first manifestation of the disease was lymphadenopathy and the head and neck pain [19]. In our study, anxiety among doctors was aroused by the lesion rapidly expanding, located in the parietal region of the head in the midline and slightly to the left.

Clinicians often use supplementary test results such as blood count with smears, erythrocyte sedimentation rate, and lactic acid dehydrogenase test. Anemia, leukocytosis, elevated erythrocyte sedimentation rate (ESR) and elevated lactate dehydrogenase levels are characteristic but nonspecific [1, 4, 6, 10]. In imaging tests, extraskeletal ES looks different from the skeletal one because it does not affect bones directly. The study of Suster et al. points to the irregularity of a small lesion and the lack of infiltration in the skull bones [15]. In the case we described, based on vascular flow features found in the ultrasound examination, atypical cavernous hemangioma was suspected. Pseudohemangioma did not respond to Propranolol treatment. MRI and angio-CT of the head were performed to confirm the diagnosis. The tumor underwent a heterogeneous, strong enhancement after contrast administration. It had no connection to the CNS vasculature and was fed by the common carotid artery. Macroscopically, ES most often resembles a soft mass with necrotic and hemorrhagic areas and cystic structures [7, 8, 9, 10]. The resected tumor was livid grey, cohesive, with cystic spaces filled with blood and covered with a pseudocapsule. Suster et al. described ES of the scalp that was of a lobular texture. The tumor was collagen coated and contained hemorrhagic and necrotic areas [15].

The ES microscopic image of the scalp bears the features in common with the image of ES tumors in other locations. The literature mentions small, round blue cells with a poor glycogen-containing cytoplasm. When we talk about Ewing's sarcoma, the formation of rosette structures is less expressed in ES than in the case of other ESFT tumors [1, 4, 6, 7, 8, 9, 11, 12, 14, 15, 16]. In each of the cases mentioned, the histopathological examination with immunohistochemical assessment was decisive, which resulted in the diagnosis of extraskeletal Ewing’s sarcoma [15, 18, 19]. In the study of Suster et al. the tumor was microcellular, with hyperchromatic cells ranging from round to oval ones. In our study, the result of the examination also showed a monomorphic, microcellular texture of the tumor located subcutaneously with few rosette

structures. In both cases, the presence of glycogen with PAS staining was confirmed. Immunohistochemistry allows scientists to perform more accurate differential diagnosis. Both in the case presented by us and in the study of Suster et al. the tumor cells were positively reported for Vimentin and negatively for Desmin and Cytokeratin [15]. Due to technological progress and the availability of genetic tests in the case described by us, doctors were able to detect by means of FISH the rearrangement of the EWSR1 gene characteristic of tumors within this family [2, 3, 6, 7, 8, 9, 11, 12, 13, 14, 15].

Chemotherapy, surgery, and radiation are used to treat Ewing's sarcoma. Due to the high sensitivity of ES to chemotherapy, there are many schemes for drug combinations, also at different doses [1, 2, 4, 6, 8, 10, 11, 14, 16, 17, 21, 22]. Up to the present, no common position has been reached on ESHN chemotherapy. In literature we met with the use of Vincristine, Doxorubicin, Etoposide and Isophosphamide. Following the analyzed articles, it can be concluded that it does not differ from ES treatment in other locations. The location of ES in the head and neck often limits the possibilities of tumor resection due to the proximity of the nervous system, which may affect the patient's morbidity [2, 6, 8, 11, 14, 16, 21]. The patient discussed by Suster et al. underwent only an extensive tumor resection, perhaps due to limited knowledge of chemotherapy at that time. The boy mentioned by Chao et al. also received chemotherapy and radiotherapy at a dose of 2000cGy. We do not have more accurate data on the patient's treatment plan described by Chao et al. Our patient underwent tumor resection and only after obtaining the results of histopathological examination extended by immunohistochemistry and genetic examination her chemotherapy according to the VIDE scheme was implemented [15, 19].

In the case we described, apart from the atypical location, it is curious that the original ES of the scalp adopted a hemangioma mask. The results of ultrasound, MRI and angio-CT were nonspecific, and this highlights the significance of early histopathological diagnosis. This delayed the introduction of appropriate treatment. There is little data concerning the features of the ES clinical picture resembling cavernous hemangioma. In literature we found a case of ES of the spine, sciatic nerve and soft tissues of the back that resembled hemangiomas [4, 10, 22]. We collected the data in **table no. 2 (Table 2)**.

Tabele 2: Comparison of case reports of EES masking as hemangiomas.

No.	Case report Characteristic	Our case report	Nortes et al.	Dhua et al.	Bemporad et al.
1.	**Age**	6 months	29 years	6 years	8 years
2.	**Gender**	Female	Female	Male	Male
3.	**First symptoms**	A rapidly growing lesion on the head covered with unchanged skin	Asymptomatic back tumor, which appeared four months earlier and gradually increased	Painful, localized deep swelling of the back of the left thigh, difficulty walking for 4 months, fever	Neck and upper back pain, paresthesia of both arms, diffuse wrist pain, weakness, bending of the neck, point pain in the lower cervical spine with limited movement range, bilateral weakness of the upper limbs
4.	**Tumor location**	Parietal region, midline and left side	Back	Left sciatic nerve	Cervical spine
5.	**Mask**	Cavernous hemangioma	Angiomatous tumor	Infected hemangioma, hematoma, or absces	Aggressive hemangioma
6.	**Tumor size**	4,2x4,2x3,5 cm	2,5–3 cm	5 cm	No data
7.	**Treatment**	Propranolol, tumor resection and chemotherapy	Surgical excision	Painkillers, antibiotics, biopsy, surgical excision, peroneal nerve transplant, chemotherapy, rehabilitation	Tumor resection and stabilization of the operated spine, chemotherapy (Cytoxane, Doxorubicin, Vincristine and G-CSF)
8.	**Tumor features in imaging studies**	Angio-CT: strong, heterogeneous contrast enhancement, vascularization of the common carotid artery, no connection to the CNS vasculature	No data	USG: infected abscess Angio-CT and MRI: Infected hematoma or hemorrhage in non-involuting hemangioma with close proximity to the sciatic nerve. CT with contrast: well-defined oval lesion in close proximity to the sciatic nerve with a feeder vessel (arrow) from profunda femoris vessel and enhancement of the lesion with contra	Non-contact CT: coarse trabecular body pattern C7, anesthesia epidural soft tissue mass extending from the left nerve hole. Image of the central sagittal T1 scale: homogeneous extracellular mass compressing the spinal cord at the back, weak in signal and roughly isointensive for gray matter. The C7 circle is compressed and the signal decreases while maintaining the space between the intervertebral discs. Sagittal T2 image of the cervical spine with a slight shift to the left of the median line: preservation of intervertebral spaces. The epidural mass is quite homogeneous and has a medium to high signal. C7 compression with increasing anteroposterior diameter. Central sagittal image T1 with contrast: heterogeneous strengthening of the epidural mass and the C7 vertebral body. Arteriography: benign, pathological hypervascularization in the body of C7, with paravertebral staining of the nerve connection at tumor level
9.	**Macroscopic features of the tumor**	Cohesive, gray-blue mass, covered with macroscopically unchanged skin flap, numerous cross-sections of cysts filled	Nonulcerated well demarcated, reddish tumor, dermoscopy: multiple lobular red structures separated from each other by fissures with a radial distribution. A whitish veil was	A variegated consistency mass in the substance of sciatic nerve with no gross involvement of the surrounding tissue	No data

		with blood, surrounded by a pseudocapsule	also observed		
10.	**Microscopic features of the tumor**	Monomorphic, small-cell tumor weaving, with poorly expressed formation of rosette structures, rich in hemorrhages. PAS and PAS-D (+). Involvement of the surrounding tissue	Well-defined blue and small cell with vesicular nuclei and eosinophilic cytoplasm tumor located in the papillary dermis with numerous hemorrhagic spaces	Uniform small round cells with mitotic figures, having a vesicular to dark chromatin with prominent nucleoli surrounded by a vacuolated to clear cytoplasm, homer-Wright and perivascular pseudo rosette formation	Densely packed, small, round to oval, hyperchromatic cells with a pycnotic nucleus and scanty cytoplasm, areas of necrosis with diffuse mitotic forms. The tumor invaded the spinal cord space. There are weak thin-walled vascular channels
11.	**Immunohistochemistry**	CD99 (diffuse +), Vimentin (diffuse y+), S-100 (focal +), Cytokeratin AE 1/3 (-), LCA (-/+), FLI-1 (+), Synaphtophisin (focal +), CD56 (-/+), CD3 (-), CD79a (-), Myogenin (-), Desmin (-)	CD99 (+), Vimentin (+), Cytokeratin AE1–AE3 (focal +), EMA (focal +), 35% of the cells were positive for Ki67 antigen, S-100 (-), Melan A (-), HMB45 (-), Synaptophysin (-), Chromogranin (-), CD31 (-),CD56 (-), CD34 (-)	No data	CD99 (+), LCA (-), PGP 9.5 (-)

Clinical manifestations of these tumors depended on their location. Nortes et al. described an asymptomatic angiomatic tumor of the back in a 29-year-old woman, which was gradually increasing [22]. Dhua et al. presented the case of a 6-year-old boy with a painful swelling located deep in the muscles of the posterior compartment of the left thigh, which imitated an infected hemangioma, hematoma or abscess. The boy had had a difficulty with walking for 4 months and he was feverish [4]. Bemporad et al. recalled the case of an 8-year-old boy with pain and bent neck, upper back pain, paresthesia of both arms and widespread wrist pain. There was also point pain in the lower cervical spine with limited range of motion. The patient felt bilateral weakness in the upper limbs. Doctors suspected an aggressive hemangioma [10]. In the girl we discussed in our study, there appeared a lump in the parietal area of the head, which was quickly expanding and was not responsive to Propranolol treatment.

VI. Conclusions

It is vital to remember about unusual locations of Ewing's sarcoma, such as soft tissues of the top of the head and masks which it adopts in the clinical picture - in the case described, a cavernous angioma mask. Despite little data on this location present in literature, oncological vigilance must be maintained. Due to the rare occurrence and low recognition of ES in soft tissues under the mask of pseudohemangioma, it represents a diagnostic and therapeutic challenge. Making the right diagnosis constituted a real challenge for a multidisciplinary team of

doctors, who had been being suggested the diagnosis of cavernous hemangioma by the clinical picture for a long time. Histopathological examination supplemented with immunohistochemical assessment and genetic examination of the removed lesion allowed to make the final diagnosis and introduce the treatment targeted at Ewing's sarcoma. The long time that has passed between the appearance of the nodule and the surgical resection of the tumor and the implementation of chemotherapy do not guarantee the girl the promising future. In oncology, it is important to quickly recognize and immediately start a therapy, which affects the patient's future prognosis.

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