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# Epidural lipomatosis with foci of hemorrhage and acute compression of the spinal cord in a child with CLOVES syndrome: illustrative case

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**BACKGROUND** Congenital lipomatous overgrowth, vascular malformations, epidermal nevi, spinal/skeletal anomalies, and/or scoliosis (CLOVES) syndrome is the most recently described combined vascular anomaly characterized by congenital excessive growth of adipose tissue, vascular malformations, epidermal nevi, and skeletal deformities. This condition exhibits a significant variability in clinical manifestations and a tendency for rapid progression and affects extensive anatomical regions. Information regarding the association of epidural lipomatosis with low-flow venous lymphatic malformations is rare, with few reports in the literature.

**OBSERVATIONS** The authors present a case of a 6-year-old girl who was admitted to the emergency department complaining of rapidly progressing weakness in her lower extremities and partial loss of sensation in the inguinal area. Radiologically, an extradural mass was identified at the T2–6 level, causing acute spinal cord compression. Urgent decompression and partial resection of the mass were performed. Despite satisfactory intraoperative hemo- and lymphostasis, postoperative lymphorrhea/seroma leakage was encountered as a delayed complication and was managed conservatively.

**LESSONS** CLOVES syndrome is characterized by the combination of various clinical symptoms, not all of which are included in the abbreviation, as well as a progressively deteriorating course, the emergence of new symptoms, and complications throughout the patient's life. This necessitates ongoing monitoring of such patients.

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**KEYWORDS** CLOVES syndrome; malformation; epidural lipomatosis; tumor mimicking; vascular-lymphatic anomaly; spinal cord compression

Congenital lipomatous overgrowth, vascular malformations, epidermal nevi, spinal/skeletal anomalies, and/or scoliosis (CLOVES) syndrome belongs to combined vascular anomalies and is one of the recently described combined forms of vascular anomalies in 2007.[1] Previously, this syndrome was not distinguished as a separate nosological unit, and diagnoses such as Proteus syndrome or Klippel-Trenaunay syndrome were established.[2] The name of the syndrome is an abbreviation for congenital lipomatous overgrowth, vascular malformations, and/or epidermal nevi, and, over time, skeletal deformities/scoliosis have also been added.[3] All combined vascular anomalies exhibit significant variability in clinical manifestations and a tendency for rapid progression and affect extensive anatomical areas. It is essential to consider the presence of associated pathologies that greatly complicate diagnosis and treatment: pathologically dilated veins, lymphatic vessels, low-flow venous lymphatic malformations, and significantly increasing intra- and postoperative risks. It is important to understand that patients with CLOVES syndrome can develop various pathologies and developmental defects, and the literature includes small cohorts or isolated presentations of clinical cases.

In this article, we present a clinical case of a 6-year-old patient with CLOVES syndrome and lipomatosis of the spinal canal characterized by acute compression of the spinal cord and severe neurological symptoms.

## Illustrative Case

### History and Examination

A 6-year-old girl with a history of CLOVES syndrome and under the care of a surgeon for lymphovenous malformations presented to

**ABBREVIATIONS** CLOVES = congenital lipomatous overgrowth, vascular malformations, epidermal nevi, spinal/skeletal anomalies, and/or scoliosis; CT = computed tomography; MRI = magnetic resonance imaging.

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our admissions department. Signs of CLOVES syndrome were as follows: congenital combined vascular malformations of a large size (lymphatic, venous, capillary) involving the chest, retroperitoneal space, pelvis, and left lower limb; adipose hyperplasia of the chest; congenital macrodactyly of the 2nd–3rd toes of the left foot; gigantism of the toes of the right foot (postoperative condition); and scoliotic deformation of the spine. In the patient's medical history, there had been multiple surgical interventions. At the age of 6 months, resection of the lymphatic malformation of the chest had been performed. Subsequent sclerosing of cystic lymphatic malformations had been performed using OK-432 and bleomycin.

At the time of admission, the patient's complaints included a disturbance in the function of the lower limbs. The mother had noted a gradual increase in weakness in both legs over the previous 5 days, which had occurred suddenly and shown a tendency to progress over the week. The child also complained of numbness in both groin folds. During objective examination of the child, vascular spots were present on the body, and there was an increase in the volume of the chest and anterior abdominal walls due to soft, elastic, tumor-like formations (Fig. 1). The child moved in a sitting position, using a wheelchair because of weakness in the legs.

On neurological examination, tendon reflexes from the legs were significantly reduced; there were markedly diminished knee reflexes and absent Achilles reflexes in both limbs. Abdominal reflexes were reduced. There was a lower deep paraparesis. Muscle strength was rated as 1/5 in both legs (Medical Research Council scale for muscle strength). A pathological Babinski reflex was present in both legs. Sensory examination revealed a hypoesthetic area in the groin folds on both sides, with sensation more affected distally in the extremities. Sensation in the anogenital region was unaffected. The function of the pelvic organs was preserved; there was independent urination without straining, and defecation was independent and unobstructed.

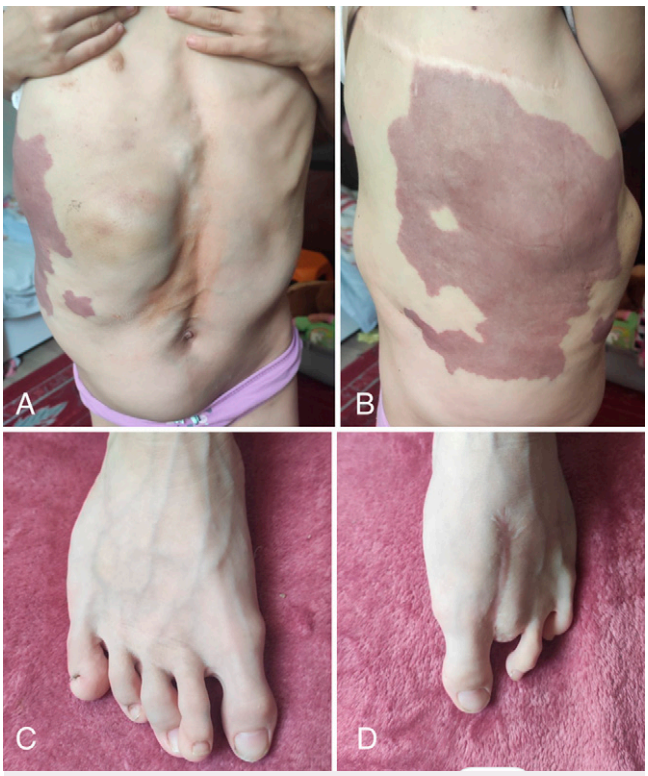


**FIG. 1. A:** Tumor-like formation on the chest and anterior abdominal wall. **B:** "Vine spot" on the chest, right side. **C:** Right foot. **D:** Left foot, postoperative condition after the gigantism of the toes.

Magnetic resonance imaging (MRI) had been performed at another hospital 1 day before admission to our clinic. According to the radiologist, there was an intradural extramedullary formation at the level of T2–3–4 segments, predominantly on the left, with a critical subtotally stenosed dural canal and compression of the spinal cord. This finding was more consistent with venous malformation with hemorrhage (Fig. 2). Computed tomography (CT) scanning of the thoracic spine at the time of admission revealed a formation at the level of T2–5 paravertebrally, extending through the intervertebral foramina. Differentiation of the spinal cord at this level was challenging. There were no definite signs of hemorrhage into the spinal cord space. Taking into consideration the child's underlying pathology and the tendency for the development of lipomas in various locations, CT and MRI examinations have been conducted repeatedly throughout her life, including studies of the spinal canal. None of these investigations had revealed deviations from normal. Considering the significant neurological deficit and spinal cord compression, urgent surgery was performed.

## Operation

Blood clotting factors (activated partial thromboplastin time, prothrombin time, fibrinogen, D-dimer) were monitored, and plasma, packed red blood cells, and thrombocyte concentrates were ordered for potential blood transfusion. Given signs of local intravascular coagulopathy (a more than 5-fold increase in D-dimer levels in peripheral blood), the child received perioperative therapy with low-molecular-weight heparin at a dose of 100 IU/kg.

The surgical procedure was conducted in a specialized neurosurgical operating room. The child was positioned on her abdomen with rolls placed under the pelvis and chest to avoid pressure on the lymphoma of the anterior abdominal wall. Intraoperative level marking was performed using fluoroscopy. A midline incision was made at the level of T1–6. With the aid of monopolar coagulation, the bony processes and arches of the vertebrae were skeletonized on both sides. Karl Storz power equipment was used to perform laminectomy at the T3–5 level. The posterior structures of the T2 body were removed with Kerrison nippers due to the high risk of massive bleeding when dealing with malformations using power equipment.

After the laminectomy, subsequent stages of the surgical procedure were performed using an intraoperative microscope (Carl Zeiss OPMI Pentero 9000). During the flavectomy, there was visualization of epidural fat accumulation, which had a significantly increased density, intermittently adhered to the dura mater, and in some areas grew into it. The fatty tissue extended into the lateral pouches, exerting compressive effects on the spinal cord, and within the structure, numerous small vessels were visualized (Fig. 3). At the T2 level, there was visualization of a cluster of dilated veins of significant diameter, bluish black in color, with pathological tortuosity. Intraoperative Doppler ultrasound was performed to monitor blood flow in the vessels, revealing no significant blood flow (Fig. 4).

Stepwise dissection of the fatty tissue with coagulation of small feeding vessels at the periphery was performed. During the operation, pathological vessels were incised, resulting in the release of

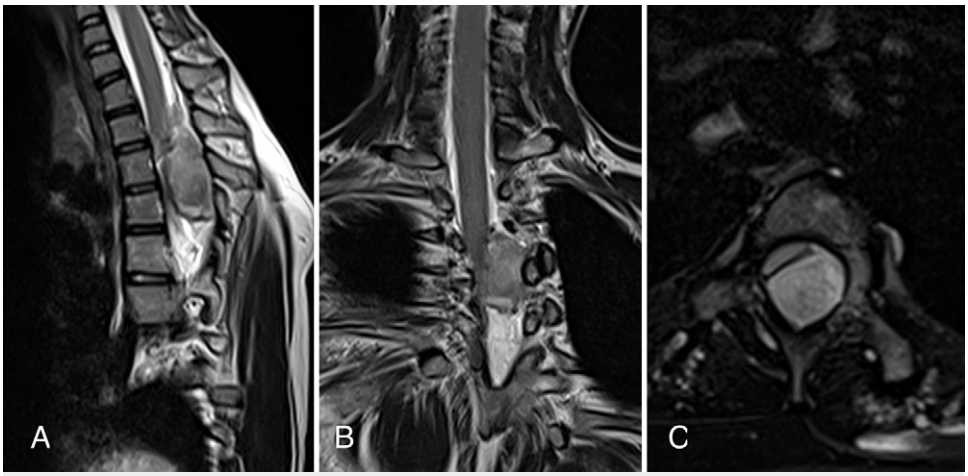


**FIG. 2.** Preoperative sagittal (**A**), coronal (**B**), and axial (**C**) MRI of the thoracic spine showing an intradural extramedullary formation at the levels of T2–3–4, predominantly on the left, with absolute stenosis of the spinal canal and compression of the spinal cord.

dark, liquefied blood. The neoplasm was gradually removed subtotally. Residual elements of the neoplasm were visualized on the anterior surface of the spinal canal epidurally, at the T3 level below the nerve root. Considering the complexity of accessing the remnants of the neoplasm (the need for pediculotomy, given the inability to retract the dural sac along with the spinal cord to avoid the risk of secondary damage), the decision was made not to perform a total removal. The bed of the voluminous formation and feeding vessels were coagulated and lined with SurgiCel Fibrillar. Hemostasis and laminoplasty were performed at the T3–5 levels, with decompressive laminectomy at T2. During the surgery, no opening of the dura mater, including iatrogenic damage, was conducted. At the stage of wound closure, there were no signs of active bleeding, lymphatic leakage, or cerebrospinal fluid leakage.

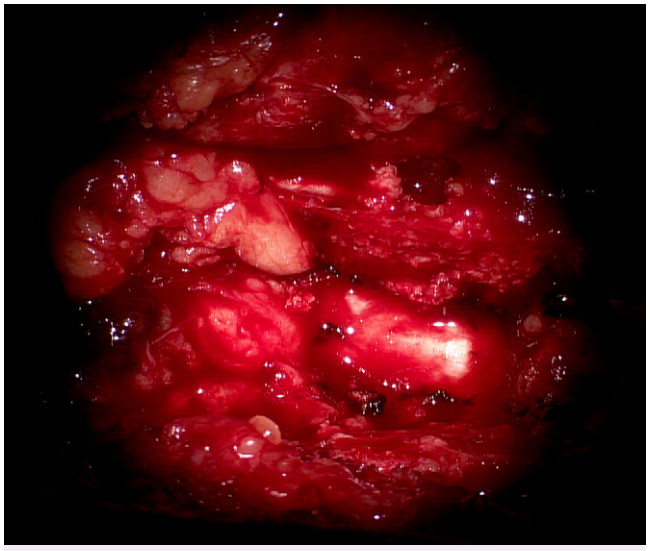

**FIG. 3.** View from the microscope showing an epidural fat mass with significantly increased density and occasional adhesions to the dura mater; some areas growing into it were also visualized. The fat extended into the lateral recesses, causing compression on the spinal cord. In the fat tissue, numerous small vessels were visualized.

### Immediate Postoperative Course

The day after surgery, MRI of the thoracic spine was performed. Epidural hemorrhagic layers, fluid accumulations, and air bubbles were observed epidurally. The previously identified formation with a fatty component at the T5 level was not fully discernible. Against the background of postoperative changes, the presence of a residual component of the known formation, including at the T3 level on the left epidurally, could not be ruled out. Focal lesions, compressions, and infarctions of the spinal cord at the specified level were not identified (Fig. 5).

The day after the operation, the child started rehabilitation sessions with a physiotherapist, and 1 week later, she was discharged from the hospital without any neurological deficits. The total hospital

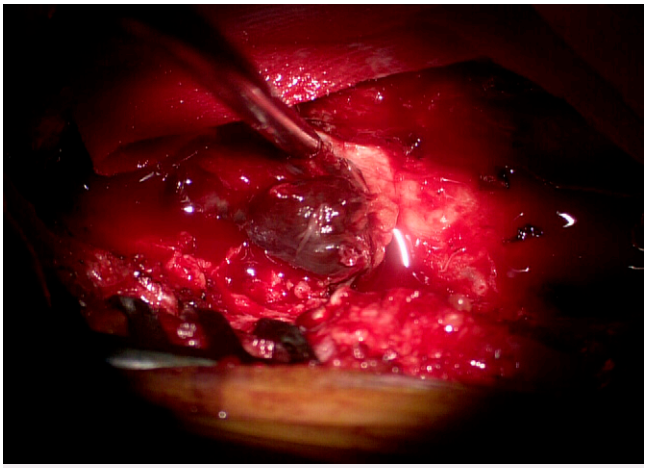

**FIG. 4.** View from the microscope showing dilated veins with a bluish black color and pathological tortuosity. Intraoperative Doppler ultrasound was used to control bloodstream in the vessels, which did not reveal significant blood flow.

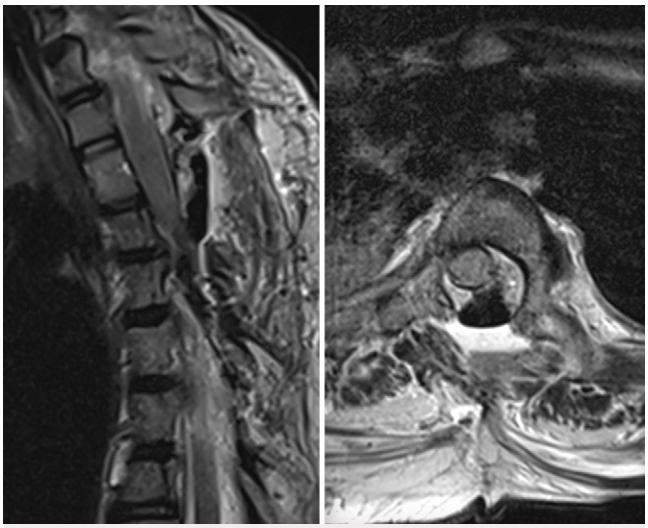

**FIG. 5.** MRI the day after surgery showing epidural hemorrhagic layers, fluid accumulations, and air bubbles. In comparison with the preoperative images, the previously identified formation with a fatty component at the T2–5 level was no longer visualized in its entirety. Residual components of the known formation at the T3 level on the left could not be excluded because of postoperative changes.

stay was 7 days. Throughout the stay, coagulogram indicators were monitored; however, the isolated increase in D-dimer (0.6 mg/L) did not affect postoperative treatment.

### Histopathological Findings

Lipoma with areas of hemorrhage and increased vascularity, including large-diameter veins, were noted. Within the thickness of the lipoma, we remarked lymphatic ducts forming small malformations with small-diameter venous vessels.

### Two Weeks After Surgery

Eight days after discharge from the hospital, 2 weeks after the surgical intervention, the child's mother contacted us with complaints of continuous leakage of "clear, transparent fluid from the upper edge of the wound, which permeates all dressings and does not stop." We recommended that the patient come to the clinic for a follow-up examination. Upon arrival at the hospital (the patient coming from another city), no discharge from the wound was observed. The child's mother explained that during the train journey, a "large amount of clear fluid suddenly leaked from the wound, and then it stopped." The wound was examined, and during the dressing change, the postoperative wound appeared healed. There was a diastasis up to 0.5 cm in the upper part with slight swelling around it, moisture around the diastasis, and areas of redness. Palpation was painless, without fluctuation. At the time of examination, there were no secretions even with active palpation of the paravertebral area. The stitches were properly placed. To visualize possible cavities and fluid accumulations, an ultrasound of the postoperative scar area and the paravertebral region was performed. No cavities, cysts, or significant fluid accumulations were detected. Considering the elevated risk of lymphorrhea in this patient, given the presence of multiple large vascular malformations (lymphatic, venous, capillary) in the chest, retroperitoneal space, pelvic area, and lower left limb, as well as the absence of any discharge from the wound that would enable differentiation between seroma and lymph (given the lack of dura mater exposure, the possibility of cerebrospinal fluid leakage was not considered), it was decided not to remove the stitches and additionally to seal the wound with Dermabond skin glue.

The patient was observed in the clinic for an additional 5 days with regular ultrasound examinations of the paravertebral area. She was discharged from the hospital in a satisfactory condition. Communication with the patient 2 months after the surgical intervention revealed a satisfactory condition. The patient's mother had not noticed any discharge from the wound or other problems. The stitches were removed at the end of the 3rd month postsurgery.

### Patient Informed Consent

The necessary patient informed consent was obtained in this study.

## Discussion

### Observations

Combined vascular malformations are rare pathologies with an occurrence of less than 1 case per 100,0000 persons (for CLOVES, Proteus, and Klippel-Trenaunay syndromes). Such diseases are classified as orphan diseases and are treated at specialized centers with the involvement of a multidisciplinary team of experts, including surgeons, orthopedists, vascular and thoracic surgeons, geneticists, dermatologists, and others.

Given the limited accumulated experience in treating this condition, many manifestations may still be unknown or are encountered so rarely and sporadically that they are not documented in the literature. One such manifestation is epidural lipomatosis with the formation of venous lymphatic low-flow malformations. The combination of venous lymphatic malformations with epidural lipomatosis is extremely rare, and, in a search for literature sources, only 3 case reports[4–6] including approximately 15 patients with vascular pathologies epidurally were found. All patients were treated with a combined approach using endovascular embolization and microsurgical removal of the malformation or only endovascular treatment and observation.[5]

Some authors described the acute onset of neurological deficit in a 12-year-old patient with CLOVES syndrome.[4] However, in that case, it was not known whether the vascular malformation had existed from birth or developed de novo.

In a 54-year-old patient with CLOVES syndrome, an arteriovenous fistula at the C5–6 level manifested with severe headaches and myelopathy.[6] The treatment was exclusively endovascular. In the patient's extended treatment history within a single clinic, where routine CT and MRI, including spinal imaging, were performed at admission and during follow-up, no pathological changes were identified on any of the scans.

### Lessons

It is not definitively known what precisely triggered the acute neurological symptoms in our patient—whether it was the significantly thickened adipose tissue or the progressing enlargement of the low-flow venous lymphatic malformation. The malformation, believed to have a valvular mechanism, seemed to grow gradually but progressively, compressing the spinal cord until it led to a compromise in its compensatory capacities, resulting in the development of acute neurological deficits. Despite the primary diagnosis, this

pathological condition necessitated urgent intervention and decompression of the spinal canal to ensure adequate spinal cord perfusion and restore its functions.

The question of removing the tumor, considering the uncertainty of the structures based on neurovisualization results (tumor with hemorrhage, arteriovenous malformation, hemorrhage), was left for the intraoperative stage. This surgical intervention was performed in a high-tech, specialized, and well-equipped neurosurgical operating room. However, even in the absence of specific equipment, such conditions must be addressed by a neurosurgical team under any circumstances.

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### Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

### Author Contributions

Conception and design: all authors. Acquisition of data: Ishchenko. Analysis and interpretation of data: all authors. Drafting the article: Ishchenko, Holoborodko. Critically revising the article: Ishchenko, Holoborodko. Reviewed submitted version of manuscript: Ishchenko. Approved the final version of the manuscript on behalf of all authors: Ishchenko. Statistical analysis: Holoborodko. Administrative/technical/material support: Ishchenko. Study supervision: Ishchenko, Holoborodko.

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