EXTRAMEDULLARY INTRADURAL CAPILLARY HEMANGIOMA IN A PATIENT OF 03 MONTHS OF LIFE: A CASE REPORT

Hemangioma capilar intradural extramedular en paciente de 03 meses de vida: reporte de caso

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ABSTRACT

Introduction: Capillary hemangioma is an extremely rare endothelial cell neoplasm as an extramedullary intradural tumor. Its presentation in infants is anecdotal and only 2 cases have been previously reported in the literature.
Clinical case: a 03-month-old infant with severe abrupt-onset paraparesis. Spinal magnetic resonance imaging (MRI) showed an intraspinal tumor at the T7, T8, and T9 levels, which captures contrast. A T7-T9 laminectomy and microsurgical resection of the extramedullary spinal tumor were performed. The pathology was reported as lobulated angiomatous proliferation with focal vascular thrombus, with positive immunohistochemistry for CD 31 and 34, compatible with capillary hemangioma. The neurological evolution was favorable. Post-surgical MRI and spinal angiography showed little residual tumor and hydrosyringomyelia in remission.
Conclusion: Capillary hemangioma is a rare pathology in infants, this case being the 3rd case reported. Its diagnosis and surgical resection are important since most patients present an improvement in the neurological deficit after surgery. Surgical treatment avoids the risk of acute bleeding

Keywords: *Hemangioma, Capillary, Spinal Cord Neoplasms, Infant, Laminectomy. (Source: MeSH NLM)*

RESUMEN

Introducción: El Hemangioma capilar es una neoplasia de células endoteliales, extremadamente raro como tumor intradural extramedular. Su presentación en lactantes es anecdótica y solo 2 casos han sido reportados previamente en la literatura.
Caso clínico: Lactante de 03 meses con paraparesia severa de inicio brusco. La resonancia magnética (RM) de columna mostró una tumoración intrarraquídea a nivel T7, T8 y T9, que capta contraste. Se realizó una laminectomía T7-T9 y resección microquirúrgica de tumor espinal intradural extramedular. La patología fue informada como proliferación angiomatosa lobulada con trombo vascular y focal, con inmunohistoquímica positiva para CD 31 y 34, compatible con hemangioma capilar. La evolución neurológica fue favorable. La RM y la angiografía espinal postquirúrgica mostraron escaso tumor residual e hidrosiringomielia en remisión.
Conclusión: El hemangioma capilar es una patología rara en lactantes siendo este caso el 3er caso reportado. Su diagnóstico y resección quirúrgica son importantes puesto que la mayoría de los pacientes presentan una mejoría del déficit neurológico después de la cirugía. El tratamiento quirúrgico evita el riesgo de una hemorragia aguda.

Palabras Clave*: Hemangioma Capilar, Neoplasias de la Médula Espinal, Lactante, Laminectomía. (Fuente: DeCS Bireme)*

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A Capillary Hemangioma (CH) is a benign vascular neoplasm that appears in the cutaneous and mucous tissue of the head and neck, mainly in children. [1] In the nervous system it has been found in the spinal cord, cauda equina, and nerve roots, but more frequently in the vertebral body, being extremely rare as an extramedullary intradural tumor.[2,3]

Capillary hemangioma is found within the vascular lesions that constitute 2-7% of intradural spinal tumors. [4] It can cause chronic progressive myelopathy or radiculopathy or cause acute bleeding with consequent sudden neurological deterioration. [5]

Neuroimaging studies, particularly magnetic resonance imaging (MRI), are useful tools for evaluating tumor size

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and location. [6] The treatment of choice is total surgical resection to eliminate the mass effect and the risk of tumor bleeding that can cause acute compression of the spinal cord.

To date, 65 cases (last case reported in April 2020) of extramedullary spinal CH have been described. We present the third case of capillary hemangioma in young infants, reported in the literature.

CLINICAL CASE

History and examination: A 3-month-old female infant, from Tarapoto - Peru, with no prenatal history, suddenly presented severe paraparesis at 15 days of age, for which she was transferred to our hospital. On examination: Patient awake, Glasgow d scale: 15 points, normotensive fontanelle, severe flaccid paraparesis (2/5).

Spinal tomography (CT) showed a mass at the T7, T8, and T9 level, heterogeneous, intraspinal, with poorly defined hyperdense borders. (*Figure 1*). The MRI showed a hyperintense lesion in T2 and STIR, contrast-enhancing with vacuum signals, measuring 1.9 x 0.8 x 0.8 cm in the cephalocaudal, anteroposterior, and lateral directions respectively; It also showed an increased volume of the spinal cord due to tension hydrosyringomyelia from C5 to the medullary cone, with an extension of the edema to the medulla oblongata, in addition to a lipoma of the Filum terminale. (*Figure 2*)

Treatment: A T7-T9 laminectomy and subtotal microsurgical resection of an extramedullary intradural spinal tumor (mass attached to the dural sac and pia mater of the dorsal spinal cord, with a soft purplish nodule and another fibrous nodule of a yellowish color, hypervascularized) were performed. There was no alteration in the neurophysiological monitoring of motor and somatosensory evoked potentials (*Figure 3*).

The pathological anatomy was reported as lobulated angiomatous proliferation with vascular and focal thrombus, with dystrophic calcification, whose immunohistochemistry was positive for CD31 and CD34, compatible with capillary hemangioma (*Figure 4*).

Clinical evolution: The clinical evolution in the postoperative period was good with significant improvement in the neurological deficit (*Figure 6*). At the 3rd postoperative month, an MRI of the spine was performed, which showed malacic tissue in the surgical bed and a significant decrease in hydrosyringomyelia (T2-T3). At 7 months after surgery, a new MRI showed areas of nodular contrast uptake inside the vertebral canal from T8 to T12, extending through the right foramen at the level of T10. An MRI of the thoracic spine demonstrated the described lesion supplied by branches of the right intercostal arteries, from T8 to T12, but without a major mass effect (*Figure 5*).

At 10 postoperative months an aortography and selective spinal angiography of intercostal arteries were performed, from T3 to L2, observing a small zone of hypervascularization at the right T10-T11 level, of the same dimensions as in the previous study, without associated vascular injury. (*Figure 6*)

DISCUSSION

Spinal neoplasms constitute 15% of central nervous system tumors [7] and vascular lesions comprise 6-7% of intradural spinal tumors, among them are hemangioma, capillary telangiectasia, cavernous angioma, and arteriovenous malformation and venous. [8]

Hemangioma is a benign hamartomatous tumor that is subdivided into 4 subtypes: capillary, cavernous, mixed, and cellular, depending on the capillary or cavernous predominance, and that are differentiated histologically by the size of the vessels. [9]

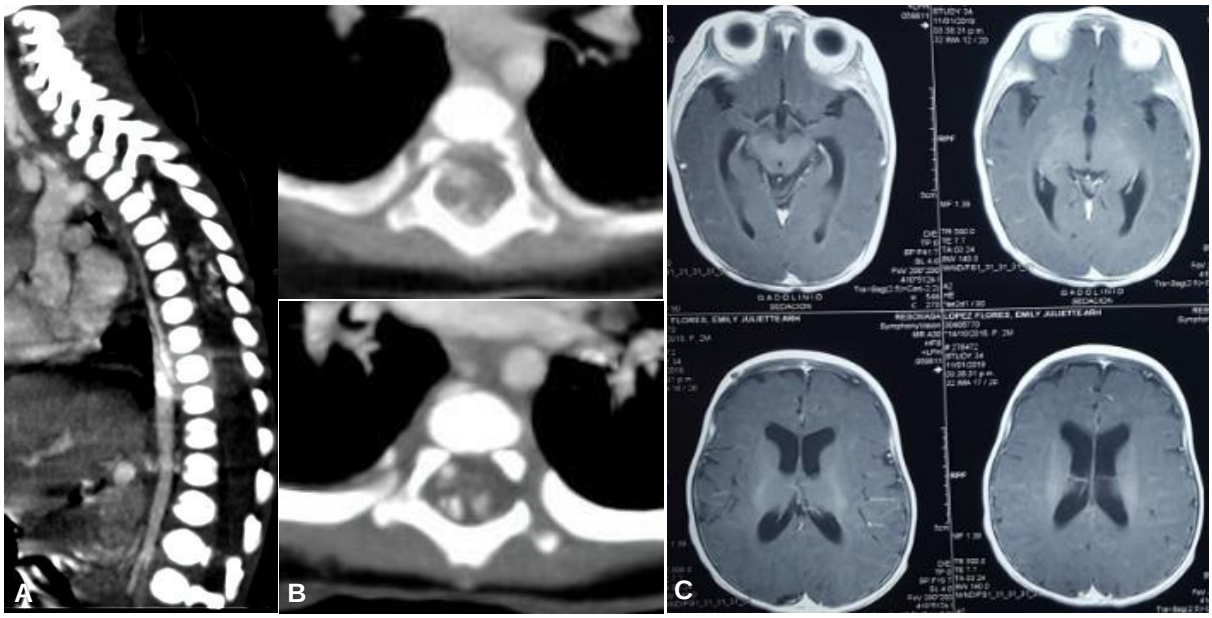

Fig 1. Tomography of the dorsal spine with contrast in sagittal and axial view **(A, B)** where a heterogeneous circumscribed mass with hyperdensity in a speckled pattern is observed, without bone erosion, at the intra-spinal level in segments T7 to T9. **(C)** Brain magnetic resonance imaging (MRI) without significant alterations.

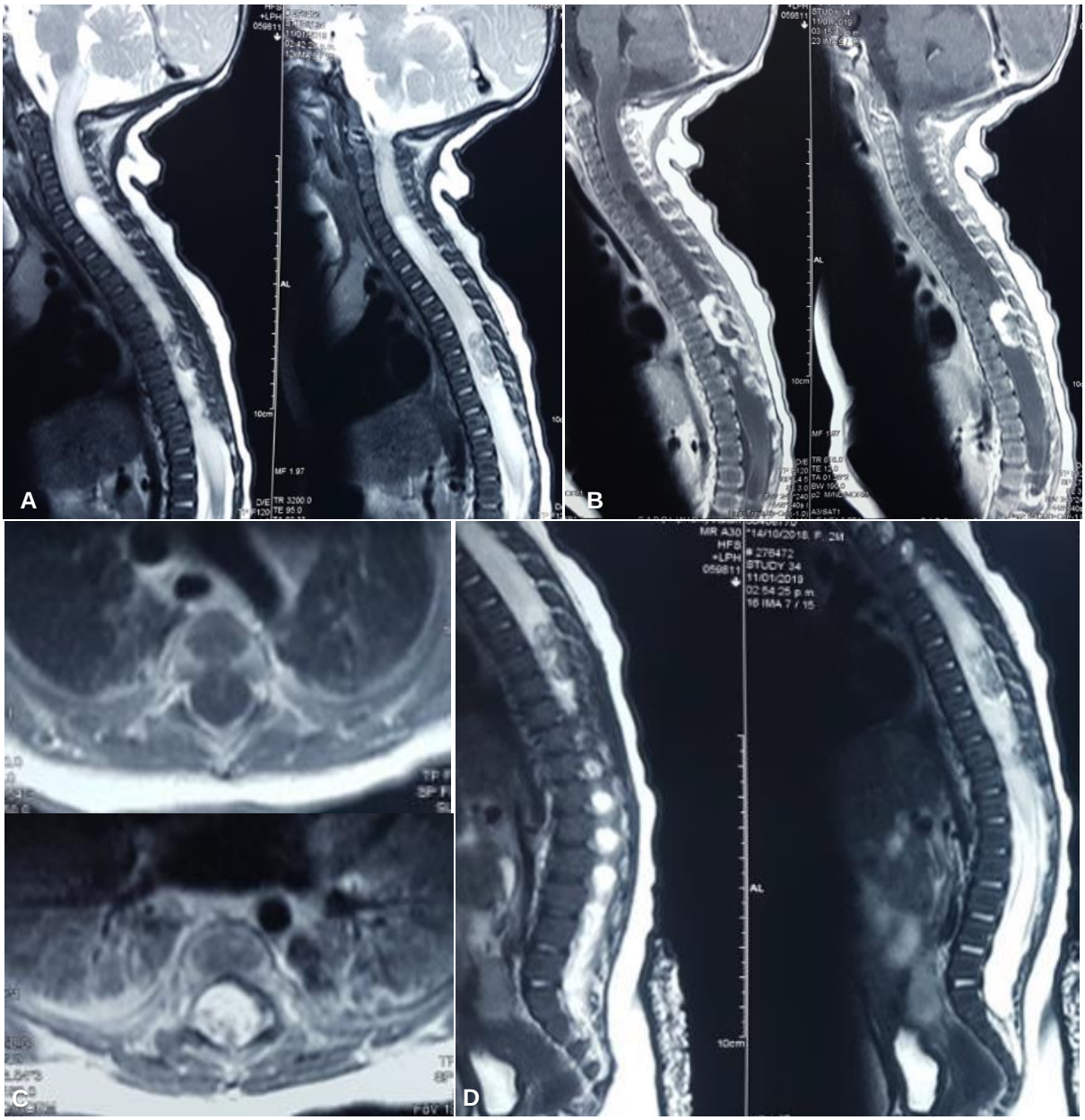

Fig 2. MRI of the total spinal cord in **(A)** sagittal T2 sequences, showing an extramedullary intradural tumor of heterogeneous intensity, **(B)** sagittal T1 with contrast, it shows intradural tumor that captures contrast; **(C)** Axial T1 with contrast and **(D)** Sagittal T2, showing dilated serpentine vascular structures on the dorsal surface and caudal to the tumor.

A cutaneous capillary hemangioma (CH) is more common in the pediatric population, unlike spinal CH which is more frequent in the adult population, with a mean age of 48 years (range 0.3-74 years) and a male / female ratio from 4 to 1. The most frequent location is at the thoracic level (75% of cases) and affects 1 or 2 vertebral segments. Due to the rarity of the tumor, incidence and mortality rates remain unclear.

Its natural history and pathogenesis are still unknown; some postulates are the following: alteration in the migration and differentiation of the primitive mesoderm at the level of the embryonic mesodermal plate (at the time when angioblastic differentiation begins on day 21-24 of embryogenesis), as the origin of hamartomatous cell proliferations endothelial, as a congenital malformation with an autosomal dominant pattern, and as vascular structures within the epineurium of the nerve roots affected during their development. [5, 10]

Capillary hemangioma originates from the blood vessels of the nerve roots, the inner layer of the dura mater (like the sign of the dural tail), or the pial surface of the spinal cord.[2] It is a highly vascularized lesion, the risk of bleeding still It is yet to be determined, but there are factors related to bleeding such as tumor size, tumor growth rate, hypertensive episodes, coagulopathy, or increased venous pressure.[11, 12]

In our case, the origin of probable bleeding is unknown intratumoral, but it could be due to a rapid growth rate that caused the rupture of the labile venous capillaries.

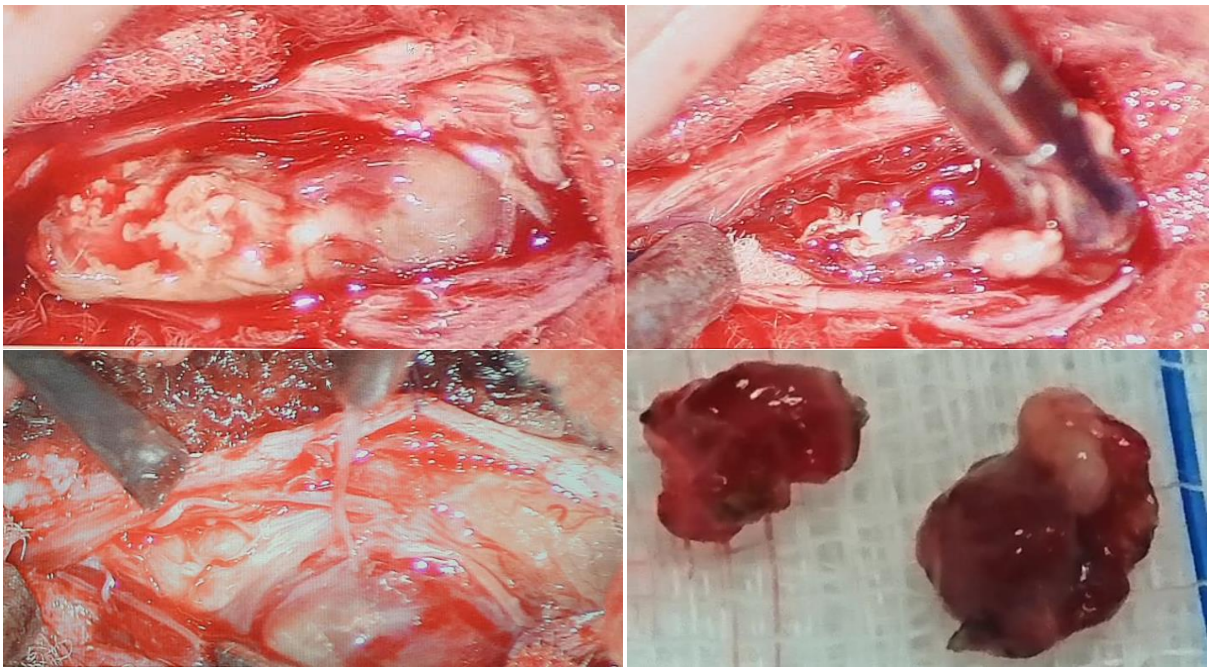

Fig 3. Intraoperative images of the microsurgical resection of the hypervascularized and lobulated extramedullary intradural tumor firmly adhered to the pia and dura mater.

Spinal arachnoiditis is a chronic inflammation of the pia-arachnoid due to subarachnoid hemorrhage, meningitis, spinal trauma, or intrathecal injection. When there is peritumoral arachnoiditis, as detailed by Holtzman et al., It is due to small bleeding or erythrocyte diapédesis.[15] Similarly, Lee J. et al. associated this inflammatory process of thickened arachnoid (strongly adherent pia mater of the HC) to old tumor subarachnoid hemorrhage, for which arachnoiditis could be considered as a diagnostic factor of suspicion for vascular tumor.[1] This arachnoiditis can cause alteration of the fluid pressure gradient cerebrospinal at the level of the central canal of the spinal cord, producing interstitial edema. [13]

The clinical picture begins with back pain (60% of cases) that progresses to radicular pain, especially when the tumor is in the lumbar spine, and/or neurological deficit.[14] Like any occupying lesion, it will be clinically accompanied by Myelopathy and/or radiculopathy that progresses gradually, over a period of months to years, that very rarely produce severe neurological deficit due to its discrete and non-infiltrative nature. It may also present intra- or peritumoral hemorrhage with acute symptoms, although bleeding is rare, being more common in cavernous hemangioma. [15]

MRI is the main test for the diagnosis of CH. On MRI, it is described as a well-defined regular lesion, isointense in T1 sequence, isointense or hyperintense in T2, avid and homogeneous contrast enhancement; Unlike the cavernous hemangioma that presents heterogeneous enhancement with areas of high and low signal that would constitute chronic subacute bleeding and hemosiderin deposits. [16]

Tekin et al. described the identification of void signals in T2 MRI sequence as dilated serpentine venous structures around the lesion that could lead to suspicion of CH, [17] as demonstrated in our patient.

The definitive diagnosis is still histopathology, but some authors propose performing a spinal angiography where a hypervascular lesion can be seen with early tumor blush and peripheral enhancement, becoming more intense and homogeneous as it fills from the periphery towards the center, with rapid opacification of the perimedullary venous plexus; [14] Then, endovascular treatment is proposed before surgery, since embolization reduces the abundant bleeding that occurs during surgical resection.[18] The risk of intraoperative hemorrhage could be associated with tumor size, hence when the lesion spreads on two vertebral bodies preoperative embolization is recommended. [19]

The pathology is like capillary hemangioma of the skin and mucosa. On macroscopic examination, it is a reddish or brownish lesion, elastic, and lobular in appearance. It has no parenchyma between the vessels and is well-differentiated from the surrounding tissue, unlike intradural arteriovenous malformation and capillary telangiectasia.[18]

At microscopy, a well-encapsulated highly vascularized lesion is appreciated, characterized by nodules of capillary-sized vessels lined by flattened and hardened endothelium, by a single layer of oval cells, in some cases lobed by fibrous septa; It may also have a partial cavernous component, being considered a transitional form9,[20] unlike cavernous hemangioma which is composed of dilated sinusoidal vascular channels, large and close together, irregular, lined by a monolayer of benign endothelium, without intervening neural tissue.[21] Occasional mitoses can also be seen. Neoplastic cells are immunoreactive to GLUT-T1, CD31, and CD34, confirming their endothelial nature, being negative for inhibin, epithelial membrane antigen (EMA), S100, WT1, and GFAP.

Within the most frequent differential diagnosis, we have schwannoma and meningioma, there are also paraganglioma, hemangioblastoma, hemangiopericytoma, hemangioendothelioma, cavernoma, angiomyolipoma, solitary fibrous tumor, terminal filum ependymoma, sarcoidosis, lymphoma, and metastasis. [7, 22]

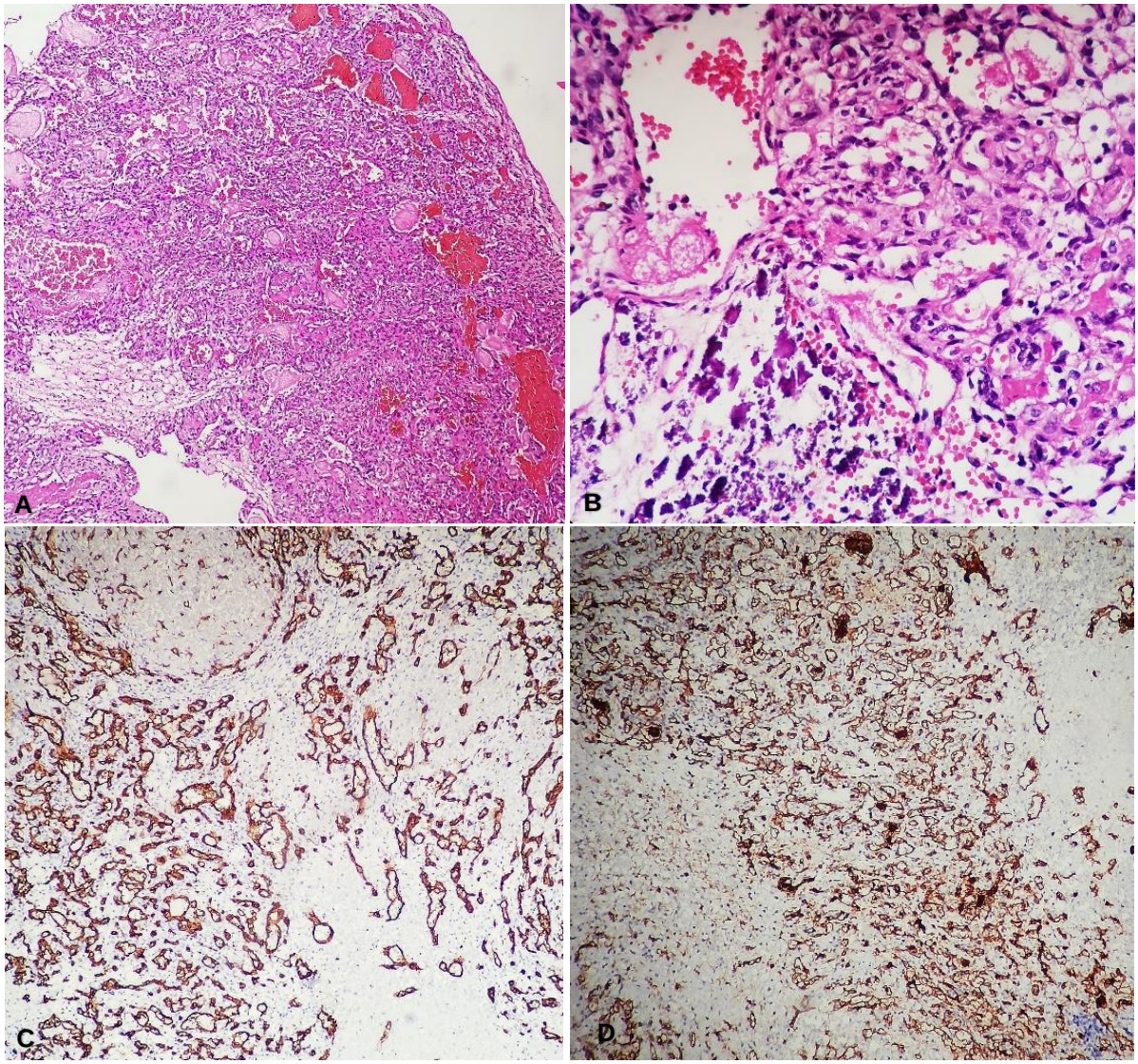

Fig 4. Microphotography of the pathological anatomy of the tumor showing: **(A)** in the central zone, proliferation of small-caliber vessels, large lacunar-type vascular dilations on both sides (HE 10x); **(B)** In the lower part of the picture, proliferation of capillaries with endothelial hyperplasia and dystrophic calcification (HE 40x). **(C)** Positivity for CD31 (IHC 40x), and **(D)** for CD34 (IHC 40x). Characteristics were compatible with capillary hemangioma.

The objective of surgery is total resection (in bloc resection) of the lesion, as the only curative treatment. Partial resection can lead to recurrence and residual tumor hemorrhage with the reappearance of neurological symptoms. [3,24] This occurs when a surgical resection type "debulking" or intratumoral decompression is performed, which causes excessive intraoperative bleeding, aggravating the already established arachnoiditis; however, if the integrity of the spinal cord is at risk, this technique is acceptable, as in the case presented.

Partial resection of the tumor can cause relapses, for which some authors recommend post-surgical radiotherapy (with acceptable results), and steroids and interferon-alpha are used as adjuvant therapy. [21]

There are reported cases of focal arachnoiditis that could be treated with subarachnoid dissection and decompression plus duroplasty; In the case of longitudinally extensive arachnoiditis, arachnoid microdissection and Ventriculo-subarachnoid bypass is used as a novel and effective method, described by Mitsuyama et al. [25]

The prognosis depends on the degree of neurological deterioration and the time elapsed from the onset of symptoms to surgery. However, most patients present an improvement in neurological deficit after surgical resection.

CONCLUSION

A capillary hemangioma (CH) is a rare entity, but it must be considered within the differential diagnosis of extramedullary intradural tumors. Early diagnosis and surgical resection are curative, prevent complications, and improve prognosis.

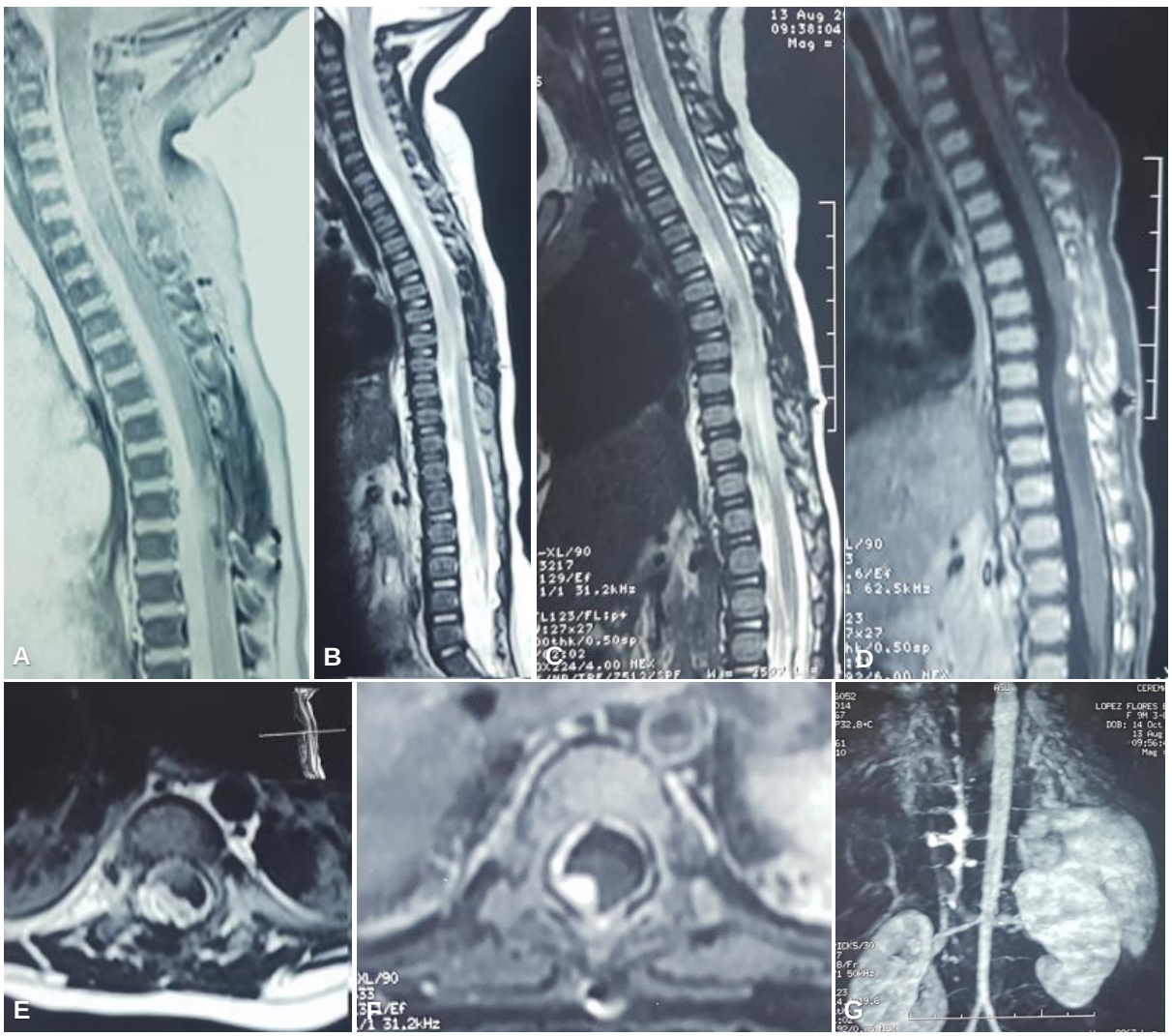

Fig 5. Postoperative holomedullary MRI. **(A, B, E)** MRI at 3 months after surgery, where myelomalacia is observed in the operative bed and a significant decrease in hydrosyringomyelia. **(C, D, F)** MRI at 07 months postoperative where a small recurrent contrast-enhancing nodule is observed at the T8 level, which extends to the right foramina and at the extraforaminal level at T10. **(G)** Angio MRI of the dorsal spine that show the dorsal lesion supplied by right intercostal branches.

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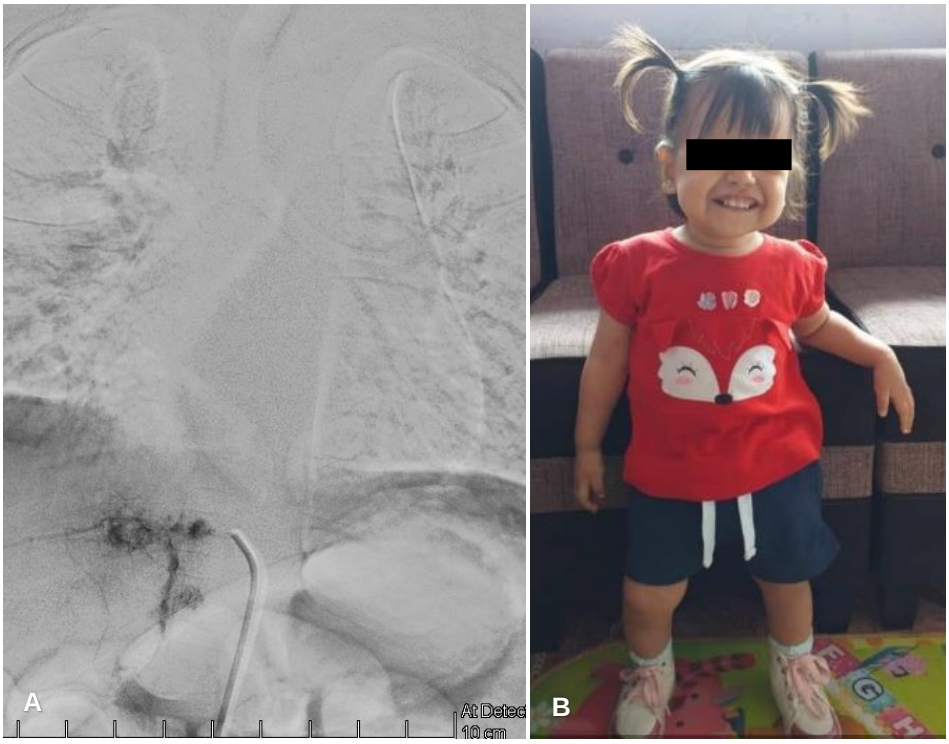

Fig 6. (A) Spinal angiography 10 months after surgery showing a small area of hypervascularization at the right T10-T11 level, without associated vascular injury. **(B)** Patient at 1 year and 07 months of age with good clinical evolution of neurological deficit (paraparesis).

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Declaración de conflicto de intereses

Los autores reportan que no existe conflicto de interés en lo concerniente a los materiales y métodos usados en este estudio o a los hallazgos específicos en este artículo.

Contribución de los autores

Concepción y diseño: Todos los autores. Redacción del artículo: Urquizo. *Revisión crítica del artículo*: Basurco, Urquizo. *Revisó la versión reenviada del artículo*: Urquizo. *Aprobó la versión final del artículo en nombre de todos los autores*: Urquizo.

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