



Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Epidural angioliopoma: A rare cause of spinal cord compression

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ARTICLE INFO

Article history:

Received 30 January 2018

Received in revised form 23 February 2018

Accepted 11 March 2018

Available online 15 March 2018

Keywords:

Angioliopoma

Spinal neoplasms

Spinal cord compression, diagnosis

Treatment

ABSTRACT

INTRODUCTION: Angioliopomas are benign, slow growing lesions, almost always located subcutaneously in the trunk or limbs. They are composed of mature lipocytes admixed with abnormal blood vessels. Spinal epidural angioliopoma are rare accounting for approximately 0.14–1.2% of all spinal axis tumors and 2–3% of epidural spinal tumors.

PRESENTATION OF CASE: We report the case of a 65 years-old-woman, presenting with complete paraplegia installed since 7 months. Magnetic resonance imaging (MRI) showed an epidural dorsal fatty mass. The patient recovered immediately after surgery. The pathological examination concluded to an angioliopoma.

DISCUSSION: Angioliopoma patients most commonly have long-lasting pain and then develop progressive neurological symptoms secondary to spinal cord compression. The mean duration of symptom progression at diagnosis is 1 year. MRI is the most reliable examination for the diagnosis of spinal angioliopoma. Total resection is the treatment of choice. No adjuvant treatment is indicated.

Since SAL are very haemorrhagic lesions, preoperative embolization is recommended.

CONCLUSION: We think that spinal cord compression caused by angioliopoma have very good functional prognosis, even if tardily diagnosed

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1. Introduction

Spinal angioliopomas (SAL) are uncommon benign tumors composed of mature adipocytes and abnormal blood vessels. They comprise approximately 1.2% of all spinal axis tumors, and 3% of all spinal epidural neoplasms [1]. Given their rarity, these lesions are often overlooked in the differential diagnosis of space-occupying lesions within the spinal canal [2]. The symptoms of SAL usually appear gradually because of the progressive compression of the cord. However, sometimes, symptoms can rise rapidly because of intratumor hemorrhage and venous thrombosis. Most SAL have a good prognosis after surgical resection [3]. We report a case of spinal angioliopoma discovered after one -year-period of complete paraplegia, in which treatment using surgical removal resulted in an unexpected good outcome.

This work has been reported in line with the SCARE criteria [4]

2. Case report

A 65-year-old woman presented with a progressive weakness of the lower extremities evolving since one year. She became totally bedridden for 6 months with onset of sphincter disorders like urinary and faecal incontinence. In her history, we found primary sterility, surgery for ovarian cyst, and gonarthrosis. She never was under corticosteroids or hormones. The physical examination found a gynoid obesity with body mass index of 29.38 kg/m², complete paraplegia at 0/5 in muscular testing, with spasticity, tendon reflexes were hyperactive on both legs with bilateral Babinski sign and epileptoid trepidation. There was a hypoesthesia below the level of T8. The modified score of Japanese Orthopedic Association JOA applied to thoracic spinal lesions [5] was calculated preoperatively and was equal to 2/11 (0-1-1-0). No bed sores were noted.

The myelo-scan (Fig. 1) revealed an intra-canalicular space-occupying mass, located in the posterior epidural space, extending from T7 to T10, compressing the spinal cord. This mass measures 55 mm in height, 11 mm in width and 13 mm in thickness. It is of heterogeneous density, with a central zone fixing the contrast product after injection, and a peripheral hypodense zone. There were no bone abnormalities.

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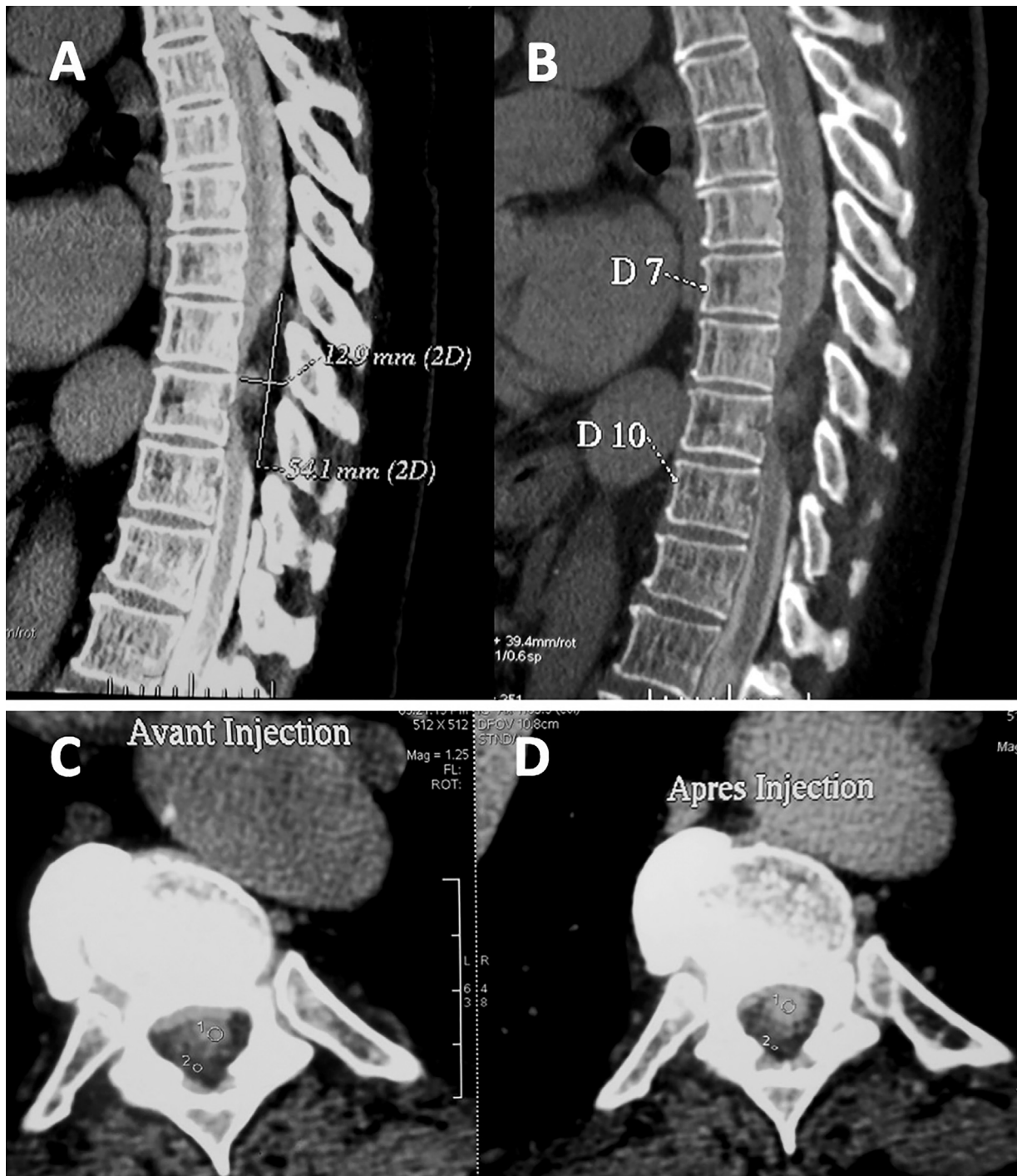


Fig 1. Myelo-scan: sagittal (A, B) and axial (C: without contrast, D: with contrast) images showing an epidural posterior mass extending from T7 to T10 with marked cord compression. The lesion has a heterogenous density with a central spontaneous hyperdense zone enhancing with contrast [1] and a peripheral hypodense zone [2].

On magnetic resonance imaging (MRI) (Fig. 2), the epidural spinal mass showed mostly a hyper-signal on T1-weighted sequences except a hypo-signal central zone. On T2 sequences, the lesion is homogeneously in hyper signal. On T1 contrast injected sequences, enhancement was heterogeneous: intense in the centre, inconspicuous or absent in the periphery. The neural foramina were widened and scalloped by the tumor.

The patient underwent an operation. Afterwards, a laminectomy of T7 to T10, an epidural reddish- yellow coloured fatty lesion was encountered. This lesion was friable, easily cleavable from the dura

mater, fusing on the lateral faces of the ME and through the neural foramina. However, the mass was extremely haemorrhagic, and the patient had to be transfused. Total resection was finally obtained by morcellation.

Pathological study revealed a benign mesenchymal proliferation, made of mature benign adipose cells, without atypia, associated with a network of small vessels having narrow lumen and edged with flattened endothelial cells. No histological signs of malignancy were found. These results were consistent with an angioliipoma (Fig. 3).

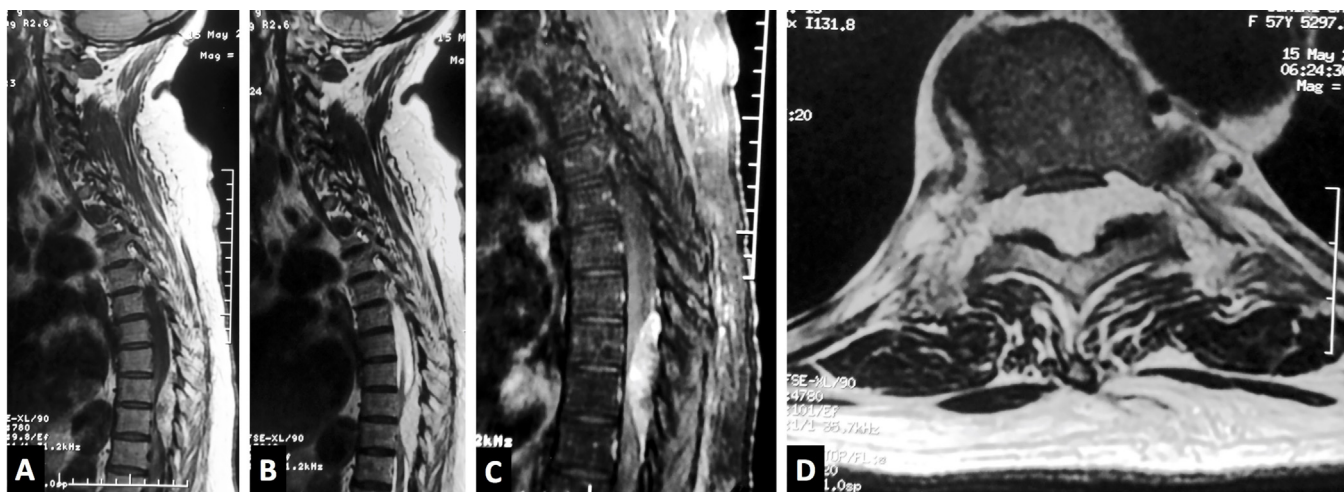


Fig. 2. MRI: (A) sagittal T1-weighted image showing an epidural mass mostly hyperintense except a hypo-signal central portion corresponding to vascular elements. (B) sagittal T2-weighted image: the lesion is homogeneously in hyper signal similar to adipose tissue. (C) Fat suppressed T1-weighted sagittal image with gadolinium: shows a highly enhanced mass. (D) axial T2-weighted image: showing the mass along the posterior epidural spinal canal compressing and displacing the spinal cord anteriorly. The right neural foramina is widened and scalloped by the tumor, with paraspinous soft tissues extension.

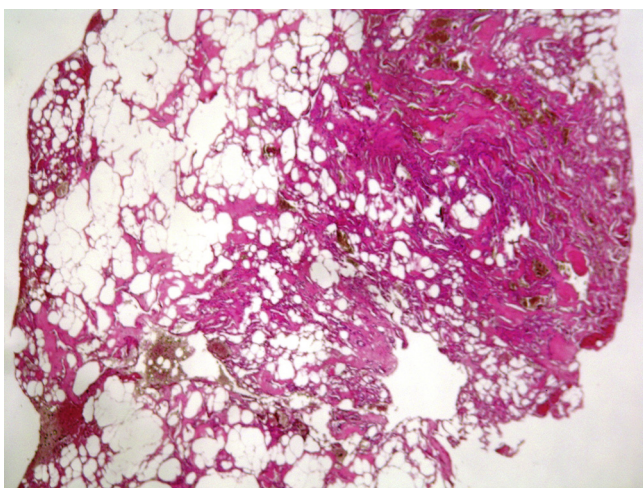


Fig. 3. Histology. The tumor is characterized by a biphasic proliferation of mature adipocyte lobules separated by thin-walled, cavernous vascular channels. Hematoxylin and eosin stain, magnification 40.

The recovery of motor skills has been very fast. Already at day 1, the patient was moving her toes and flexing her knees. At one week, she could stand up and walk with a zimmer frame. At one month she could walk with a knobstick, and at 6 months, she has completely recovered a normal walk. Normal function was obtained 6 months postoperatively though paraplegia was lasting for one year. The sphincter disorders have also improved but there still some urinary leaks. The score of the modified JOA postoperatively was 9/11 with a JOA recovery score [6] of 77.77%. Five years later, the patient remains stable and there is no recurrency on MRI control.

3. Discussion

Angiolipomas are benign tumors that look like nodules located in the subcutaneous spaces of the forearms, trunk, and neck. Very rarely, they develop in the spinal canal and constitute the spinal angiolipomas SAL which represent 0.14–1.2% of all spinal tumors, about 2–3% of epidural spinal tumors and 16–35% of spinal lipomas [1]. SAL are located in thoracic spine 78%, lumbar column 10%, and

less than 1% cervical or sacral column [7]. SAL occur in middle-aged women with a sex ratio of 3/2 [8].

The first case of AES was described in 1820 by Berenbruch in a 16-year-old boy with a spinal tumor. The diagnosis was made at autopsy, after the death of the patient following the surgery [9]. However, the angiolipoma entity was established in 1960 by Howard and Helwig, as a separate pathological entity, containing both mature adipose cell proliferation and vascular proliferation [10].

Physiopathology of angiolipomas is still uncertain. The most accepted hypothesis is that the mesenchymal stem cell, will differentiate, under the influence of unknown factors, either in lipoma or angioma, or in an intermediate entity: the angiolipoma. Hemangiomas and lipomas thus represent the ends of a spectrum, within which angiolipomas constitute an intermediate entity [11].

From a clinical point of view, SAL do not differ from other benign spinal tumors. Angiolipoma patients most commonly have long-lasting pain and then develop progressive neurological symptoms secondary to spinal cord compression [8]. The mean duration of symptom progression at diagnosis is 1 year [12].

Several factors will contribute to the growth of these lesions and thus to symptoms onset: traumatic, inflammatory and hormonal stimuli. Hormonal factors are the most incriminated, evidenced by the prevalence of peri-menopause (82.2% of patients [1]). Also, pregnancy was an aggravating factor in 15% of women with epidural angiolipomas. This phenomenon is also observed in patients under steroids or in cases of excessive weight gain [13]. The vascular and hormone-dependent characteristics of angiolipomas, make that the clinical history is made of an alternation of periods of aggravation and remission. The sudden onset or aggravation of neurological signs, due to sudden increase in tumor volume, is caused by intra-tumoral thrombosis, haemorrhage or steal syndrome [14].

Computed tomography (CT) reveals a lipomatous extradural mass. In most cases the lesion is hypo-dense, but it can be hyper-dense depending on the size of the vascular component. Bone erosion can be found due to tumor compression. The CT scan findings are not specific [15]. MRI is the most reliable examination for the diagnosis of SAL [16], showing a fatty tissue, typically hyperintense in T1 and T2 sequences. This tissue is trabeculated by vascular elements which enhance variably after contrast injection. Contrast enhancement is clearly observed on T1 sequences with gadolin-

ium injection and fat saturation (fat-saturation sequences) [17]. The presence of large zones in T1 void signal, is due to the richness in vessels, and can predict preoperatively a high vascularized tumor [18]. MRI helps making differential diagnosis with lipomas, spinal vascular tumors such as fistulas, metastases, meningiomas and hemopathies. Angiography, if done, can help with differential diagnosis, assess vascularization of the lesion, and may be used for embolization of the tumor which helps with removal [17].

Macroscopically, the tumor is an encapsulated or unencapsulated, reddish soft mass, easily cleavable from the dura mater. Histologically, it is composed of a proliferation of normal mature adipocyte lobules and blood vessels, normal in appearance or mimicking capillary angiomas, cavernous angiomas or arteriovenous malformations. The fatty tissue is of mature type without any particularity. The fat/vessel ratio is variable from 1:3 to 2:3. Tumors containing smooth muscle cell proliferation are subclassed into angiomyolipomas. A thin capsule, which is missing in places, surrounds the lesion. Neither atypia, pleomorphism or mitotic figures or karyotypical abnormalities were found [1].

Total resection is the treatment of choice. In case of impossibility of surgical treatment, or residue, a diet with weight loss improves the symptoms in 81.8% of cases [19]. The surgical approach varies according to the extension of the lesion. Most often a laminectomy alone is indicated. Anterolateral approach is indicated for vertebral extension with or without osteosynthesis [20,21]. No adjuvant treatment is indicated (radiotherapy, chemotherapy) as long as it is a benign lesion, even in the case of incomplete resection [1].

Since SAL are very haemorrhagic lesions, preoperative embolization is recommended [17,21]. Recurrence is exceptional. One lonely case has been reported in the literature 12 years after the first procedure [22]. No other case of recurrence has been described since, even in the case of SAL titled as “infiltrating” [23] or in case of partial resection [18]. No malignant transformation has been reported in the literature [1]. Surgery guarantees complete recovery without sequelae in more than 90% of cases even in cases of severe neurological involvement [24].

4. Conclusion

Spinal epidural angioliopoma is a rare cause of progressive spinal cord compression. MRI is necessary for diagnosis. Necessary precautions before and during surgery have to be taken (embolisation, transfusion). The treatment consists on complete surgical excision, adjuvant treatment is not indicated. The prognosis after surgical management of this lesion is excellent even at the stage of complete paraplegia.

Conflicts of interest

No conflict of interest

Funding source

No funding.

Ethical approval

The Ethic's Committee of the Faculty of Medicine of Tunis approved this study prospectively.

Consent

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

A copy of the written consent is available for review by the Editor of this journal.

Author contribution

All listed authors have made substantial contribution to the following aspects of the manuscript:

MR, SA, MAK and AZ participated in diagnosing and treating the patient, acquisition of data.

MR collected the findings and drafted the manuscript.

KB and IZ revised the manuscript.

The authors read and approved the final manuscript.

The integrity of this work is guaranteed by Dr. Mouna Rkhami and Dr Ihsen Zammel.

Guarantor

The integrity of this work is guaranteed by Dr Ihsen Zammel.

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