Abstract

Dumbbell shaped epidural cavernomas are uncommon lesions. Of these, the extraforaminal extension is usually limited, and the lesions are removed surgically by an extended laminectomy. We present two cases of dumbbell shaped epidural cavernomas with a considerable intrathoracic growth. They were resected through a combined thoracotomy and laminectomy. Only in the case in which a wide approach to the posterior mediastinum was performed it was possible to completely remove the lesion. Since it is possible to anticipate this histological diagnosis by the magnetic resonance imaging characteristics, we propose that a wide transthoracic approach should be performed associated to a laminectomy if a complete resection of the lesion is the goal.

Keywords: Cavernous angioma; Epidural; Laminectomy; Thoracotomy; Thoracic spine

1. Introduction

Cavernomas are benign vascular malformations very uncommonly found within the thoracic cavity, where they may appear as extensions of epidural lesions [1]. Although posterolateral surgical exposures have been advocated to remove these lesions [1,2], their unique surgical features would recommend a wide exposure if the mediastinal extension is large. We present two cases of epidural intraspinal cavernomas with thoracic invasion approached by two different combinations of thoracotomy and laminectomy.

2. Case reports

2.1. Case 1

A 74-year-old woman presented with a 6-month history of progressive paraparesis and numbness in both legs. Neurological examination revealed a right 3/5 and left 4/5 paraparesis and a Th5 symmetric distal hypoesthesia. Magnetic resonance (MR) imaging showed a left posterolateral epidural mass compressing the spinal cord at the Th4–Th5 vertebral level, and a left paravertebral intrathoracic mass displacing the superior pulmonary lobe, measuring 2×2.5 cm axially and 4.5 cm craniocaudally, contacting the descendent aorta. The mass was isointense with the T1 and hyperintense with the T2 sequence (Fig. 1A). Similar lesions were found at the vertebral bodies of Th3, Th4, Th5, Th7 and Th12. The operation was performed with the patient in right lateral decubitus position. A thoracic surgical team performed a left posterolateral thoracotomy, and a smooth red-brown tumor adherent to the 4th intercostal nerve extending from the 4th–5th intervertebral foramen to the descendent aorta was dissected and removed. Simultaneously, a neurosurgical team performed a 4th, 5th and 6th laminectomy and the epidural portion was removed. The histological diagnosis was a cavernoma. There were no postoperative complications. The control MR showed a complete removal (Fig. 1B). The rest of the vertebral lesions have remained unchanged after 72 months of follow-up. The patient presents only a 4th left intercostal dysesthesia.
2.2. Case 2

A 57-year-old woman presented a 2-year history of right cervical, shoulder and chest pain. Neurological exam showed a right 4/5 paraparesia and left hemihyposthesia. MR showed an epidural mass exiting the 5th intervertebral foramen continuous with a right paravertebral mass measuring $6 \times 3.5 \times 3$ cm in the transverse, antero-posterior and craniocaudal diameters, isointense in the T1 and hyperintense in the T2 sequence (Fig. 2A). Other similar lesions appeared within the vertebral bodies of Th10 and Th12. The patient was positioned in left lateral decubitus. A right posterolateral incision continued by a paravertebral one was made. After a muscular flap was developed, a 4th–5th laminectomy and foraminotomy was performed and the intraspinal component of the lesion removed. Then, the head and posterior part of the 5th rib removed and the intrathoracic cavity entered. A reddish mass exiting the 5th intervertebral foramen was piecemeal removed. The histopathological diagnosis was a cavernoma. The postoperative MR revealed a $2.5 \times 1.5 \times 1.2$ paravertebral mass (Fig. 2B). The patient presents only local wound pain.

3. Discussion

Cavernous hemangiomas (also known as cavernomas or cavernous angiomas) are benign hamartomatous vascular malformations without mitotic activity, although they may grow due to repeated hemorrhage, or thrombosis and recanalization [1]. They consist of packed sinusoidal vascular spaces lined by a single endothelial layer without elastic or muscular tissue [1]. Although they may be surrounded by a pseudocapsule and lack a gross feeding vessel [3], these thin walled soft friable lesions are prone to bleed and to break during surgery, and both factors are known causes of incomplete resection [2].

Fig. 1. (A) Preoperative spine and thoracic MR corresponding to case 1 showing a posterolateral epidural mass isointense with the T1 sequence located at the Th4–Th5 vertebral level, entering into thoracic cavity through the left intervertebral foramen, in contact with the descendent aorta. (B) Postoperative MR of case 1, showing no residual tumor.

Fig. 2. (A) Preoperative spine and thoracic MR corresponding to case 2 showing a posterolateral epidural mass isointense with the T1 sequence located at the Th4–Th5 vertebral level, entering into thoracic cavity through the right intervertebral foramen (B) Postoperative MR of case 1, showing a residual mass.
Cavernomas are most commonly located in the skin, subcutaneous tissue, brain, several viscerae and vertebral bodies, sometimes as multiple lesions [6]. Pure epidural cavernomas, more frequent at the thoracic spine [3], comprise 12% of all spinal cavernomas [4]. Clinically they may present with symptoms of spinal cord or nerve root compression. They are very rare within the thorax, either at the anterior mediastinum or the lung [4]. Another source of posterior mediastinal cavernomas may be a growth of an epidural cavernoma through the intervertebral foramen, adopting a ‘dumbbell’ shape. Including ours, only 13 cases of dumbbell shaped epidural cavernomas have been reported to date [1,2,5,6]. A huge intrathoracic component is seldom reported.

MR may provide the differential diagnosis from other dumbbell lesions [7]. Cavernomas are iso or hypointense in T1 and hyperintense in T2 weighted sequences, enhancing homogeneously or slightly heterogeneously with contrast, contrary to ependymomas or schwannomas, which show a hyperintensive rim. The existence of multiple vertebral lesions may support the diagnosis. Other lesions, such as herniated discs, meningiomas, lymphomas, Ewing’s sarcomas, chondromas or angiolipomas show different characteristics [1].

Although malignant degeneration of epidural cavernomas has not been reported [3], incomplete tumor removal may lead to recurrence of the symptoms [7], so a complete resection is advisable [2,7]. Up to now, the reported cases of dumbbell shaped cavernomas have been approached through a laminectomy, either standard or extended with a foraminotomy or costo-transversectomy [1,7]. However, these approaches provide a limited exposure of the posterior mediastinum and may lead to incomplete resection of an intrathoracic extension, which may require a further thoracotomy [2]. Since piecemeal resection or profuse bleeding are known factors preventing a complete removal [2], a sufficient exposure of the whole lesion is necessary for a radical excision. These, along with intercostal nerve involvement, may recommend a wider transthoracic approach combined with the laminectomy if there is an intrathoracic extension [3]. In the case of other dumbbell tumors, Grillo and co-workers [8] advocate a posterolateral thoracotomy extended posteriorly by an upward paravertebral branch that allows access to the selected intercostal space and vertebral laminae. On the contrary, Akwari and co-workers [9] propose a paravertebral incision for the laminectomy and another one for the thoracotomy. Ricci et al. consider that Grillo’s approach is sufficient for tumors involving only one foramen and with a limited thoracic extension, while for greater or more complex lesions they recommend Akwari’s approach [10]. In our first case we chose Akwari’s technique for the possible aortic involvement and the size of intrathoracic component. A simultaneous approach by two teams permitted to shorten the procedure duration and a safer control of possible residual lesions. In the second case, the lesion was approached by Grillo’s technique, which was not sufficient to control the whole lesion, permitting only a partial resection. The residual lesion may be reoperated through a thoracotomy, which should be indicated if symptoms recur. More controversial is the use of radiation therapy [1,2,7], which might be indicated in the case of asymptomatic growth or persistent pain.

We conclude that a huge intrathoracic extraforaminal extension of an epidural cavernoma should be approached through a simultaneous laminectomy and thoracotomy permitting a wide exposure of the lesion if the complete surgical removal is the goal.

References