

Aggressive Adenoid Cystic Carcinoma With Asymptomatic Spinal Cord Compression Revealed By A “Curtain Sign”

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ABSTRACT

The authors present a case of unusually aggressive adenoid cystic carcinoma of the head and neck. The patient presented with sciatica one year after initial diagnosis. She was otherwise asymptomatic. Complete work-up for bone involvement, included bone scan and MRI. The patient had painful osteolytic sacral metastasis and asymptomatic thoracic (T5) vertebral metastasis revealed by a typical curtain sign on MRI. The curtain sign, originally described by Trolard, is due to the displacement of the Trolard's membrane with the median ligament attaching the dura to the bone while there is a dorsal displacement and loosening of the posterior longitudinal ligament. The patient benefited from radiotherapy and did not develop respiratory distress, paraplegia or pain but died of liver metastases.

CASE REPORT

A 54-year-old woman presented with an incomplete unilateral cauda equina syndrome. Her recent history was remarkable for adenoid cystic carcinoma of the submandibular gland pT2pN0M0. On pathology specimen, submandibular gland resection and homolateral selective neck dissection showed clear margins, grade II adenoid cystic carcinoma with solid component without perineural invasion (Fig. 1 and Fig. 2). Three months later, she presented with isolated nodal recurrence of her adenoid cystic carcinoma in upper cervical and submandibular areas. She underwent resection followed by postoperative irradiation. Six months later, she presented with atypical sciatica and incomplete cauda equina syndrome on physical examination. Lumbo-sacral and bone scan CT showed a bulky osteolytic metastasis of the second and third sacral vertebrae (Fig. 3). Bone scan also showed ambiguous abnormalities of the fifth dorsal vertebra. MRI showed metastasis of the fifth dorsal vertebra (T5) (Fig. 4 and Fig. 5), which was totally asymptomatic. There was a “curtain sign” (Fig. 6) pathognomonic for epidural involvement (1). She underwent oral steroid therapy and decompressive palliative

irradiation. With radiotherapy, she remained asymptomatic on her spine. However, she progressed in spite of two cycles of fluorouracil and carboplatin and further received imatinib for two weeks. She then died of liver metastases (Fig. 7).

DISCUSSION

Adenoid cystic carcinoma accounts for about 1% of all head and neck malignancies. It has a tendency for a prolonged clinical course, with local recurrences and distant metastases sometimes occurring many years after presentation. Standard treatment for salivary gland adenoid cystic carcinoma is surgery and post-operative radiotherapy (2). While prolonged survival (several years) has been observed with lung metastases, metastasis to bony sites is associated with a median survival of less than 12 months (3). Liver metastases are exceptional. Another primary cancer was ruled out. In the patient described the solid histological subtype and major

salivary gland involvement (4), the short intervals between the initial diagnosis and the two recurrences suggested aggressive tumor behavior. Such cases mandate more thorough work-up than only head and neck CT and chest X-ray. These should include a chest CT in lieu of an X-ray and may benefit from a bone scan or 18FDG Positron Emission Tomography. Epidural spinal cord compression usually occurs in patients with disseminated disease. Epidural spinal cord compression represents a medical emergency because delayed treatment can be responsible for irreversible neurological deficits, such as paralysis. In our case, vertebral body metastasis with epidural involvement was asymptomatic. A curtain sign was clearly identified on axial MRI views (Fig. 4 and Fig. 5). The anterior dural attachments are largely ignored, although they have been described since the last decades of the 19th century by Trolard (5). The curtain sign may also be identified on parasagittal views as ill-defined longitudinal margins of the posterior longitudinal ligament. The vertebral body fatty marrow contains 80% fat in adults and therefore exhibits a high signal. T1 weighted-sequences are useful to detect focal tumors as they usually are in relatively lower signal. T2-weighted sequences are useful for spinal cord compressions and to image the anterior epidural space, when invaded by metastases. Fat suppression sequences (so-called STIR sequences) can be performed to cancel the fat signal but also cancel signals identical to fat such as hematoma or contrast-enhanced tissue. Contrast medium is useful to enhance bony tumors and leptomeningeal metastases (6).

Most of the responses to single-agent or combination chemotherapy or targeted therapies such as imatinib have yielded response rates in the 20% range in the metastatic setting. Responses have been partial and were of brief duration and these treatments have failed to demonstrate a survival benefit. Bone work-up and spinal MRI can be useful to screen and detect bony and leptomeningeal lesions. Palliative irradiation may be useful in an attempt to preserve the quality of life in aggressive cases with bony metastases.

TEACHING POINT

Adenoid cystic carcinoma is usually slow growing and distant metastases preferentially go to lungs. Some clinical and pathological factors, such as involvement of accessory salivary glands, size, histological grade, solid or cribriform adenoid cystic subtypes and perineural invasion, may indicate aggressive subtypes that require more thorough work-up. The curtain sign is a pathognomonic sign of epidural involvement and should be recognized early on MRI.

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FIGURES

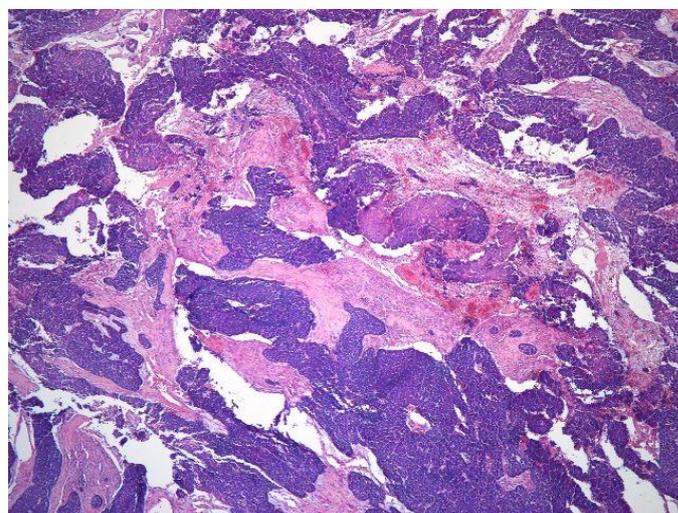


Figure 1: Histological image of adenoid cystic carcinoma. Massive infiltration by a solid adenoid cystic carcinoma. (HE X 40)

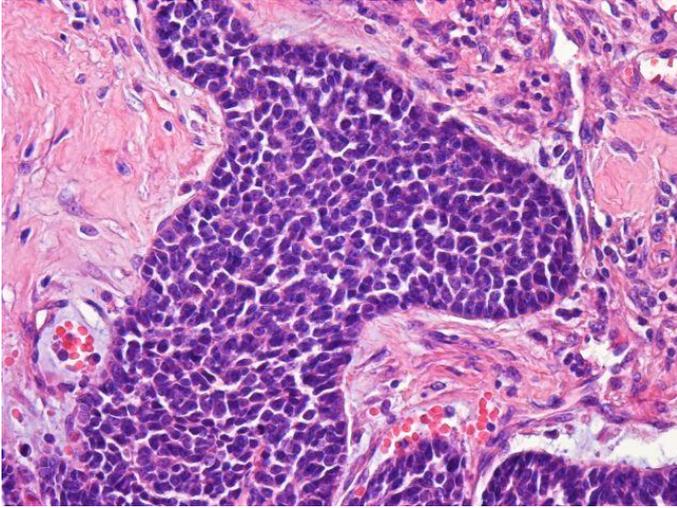


Figure 2: Histological image of adenoid cystic carcinoma. Tumor cells are small, hyperchromatic with numerous mitoses. (HE X 400)

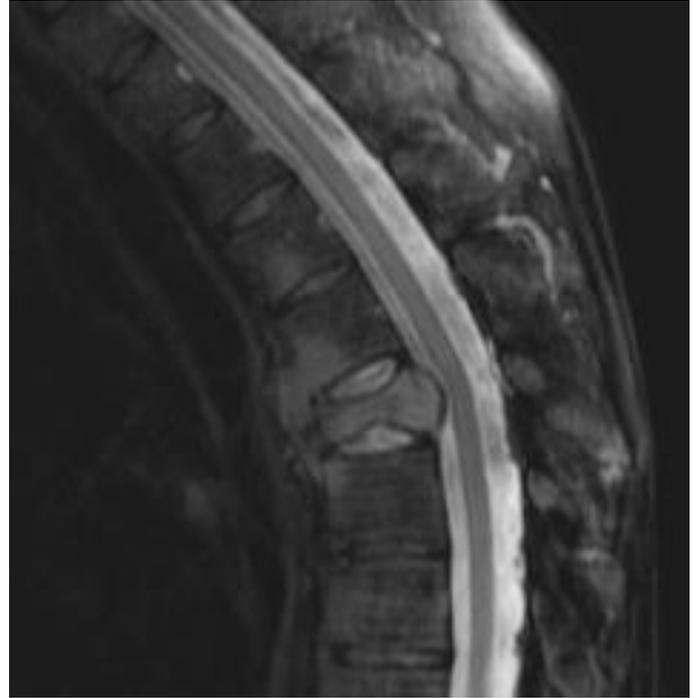


Figure 4: MRI (T2) of the thoracic spine without contrast: metastasis of an adenoid cystic carcinoma. Tumor extension into the anterior part of the epidural space, well seen on sagittal view on T2-weighted sequences. Para-sagittal views show loosening of the posterior longitudinal ligament.



Figure 3: CT of the pelvis with contrast: metastasis of an adenoid cystic carcinoma. CT scan showing a large osteolytic metastasis of the sacrum.



Figure 5: MRI (T1) of the thoracic spine with contrast: metastasis of an adenoid cystic carcinoma. Tumor extension into the anterior part of the epidural space, well seen on sagittal view on gadolinium T1-weighted sequences. Para-sagittal views show loosening of the posterior longitudinal ligament.

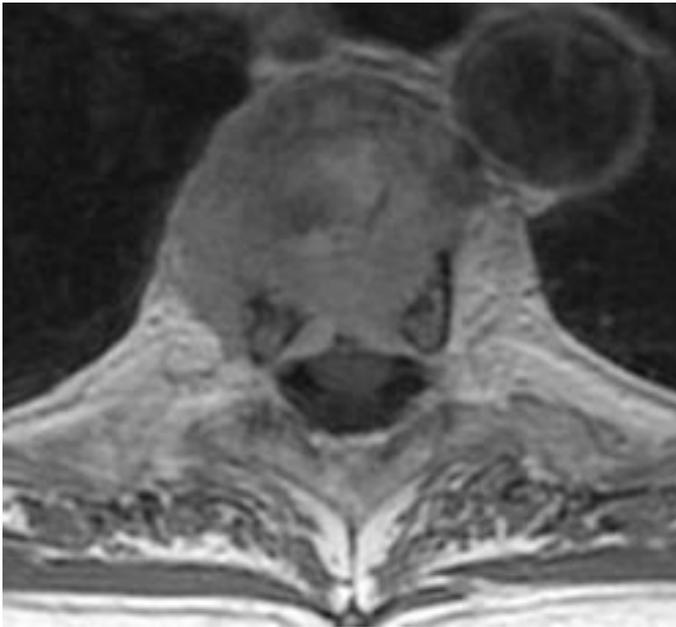


Figure 6: MRI (T1) of the thoracic spine with contrast: metastasis of an adenoma cystic carcinoma. Axial gadolinium-enhanced T1-weighted image at the level of the thoracic vertebral metastasis shows a typical curtain sign due to bony tumor extension into the anterior part of the epidural space. The curtain sign is due to the presence of the Trolard membrane with the median ligament attaching the dura to the bone while there is a dorsal displacement and loosening of the posterior longitudinal ligament.

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Figure 7: CT of the abdomen with contrast: metastasis of an adenoid cystic carcinoma. CT scan showing a large necrotic metastasis in the liver.