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Atypical presentation of a cervical breast-cancer metastasis mimicking a dumbbell-shaped neurinoma

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ABSTRACT

INTRODUCTION: Spinal metastases are frequently encountered in patients with breast cancer. Because of recent improvements in oncologic therapies a growing incidence of symptomatic leptomeningeal metastases (LM) should be expected. The differential diagnosis of LM comprises a wide range of conditions, including neurinoma. The radiologic discrimination between metastases and neurinomas is primarily based on distinct neuroimaging features, particularly number, size and growth pattern.
PRESENTATION OF CASE: We report the first case of a solitary leptomeningeal metastasis of a cervical nerve-root, which mimicked a benign dumbbell-shaped neurinoma, using neuroimaging and visualized intraoperatively. The tumor was successfully treated with surgery followed by adjuvant radiochemotherapy (RCT).
DISCUSSION: While the patient history directs towards a metastasis, the localization, growth pattern and MRI signal were concordant with a cervical neurinoma. The current literature is not conclusive concerning the optimal choice of treatment; the therapy is strictly palliative and indications for surgery remain individual decisions. However, due to recent improvements in survival of patients with LM require reconsideration of established strategies.
CONCLUSION: The present case report and the reviewed literature point towards a growing clinical relevance of symptomatic LM in cancer patients and their possible atypical presentations and locations.

1. Introduction

In advanced stages, solid tumors can metastasize to various organs, including the central nervous system. In particular, breast, lung and prostate cancer metastases are frequently encountered in the spine.[1] Current operative concepts are focused on lesions to the vertebra, while metastases to non-osseous spinal structures are rare and usually associated with advanced disease.[2–5] The ongoing advancements in oncologic therapies have led to improvements in the life expectancy of cancer patients, even in palliative situations. Therefore, an increasing incidence of the formerly rare symptomatic leptomeningeal metastases (LM) should be expected.[1,2] LM cause obstruction of cerebrospinal fluid (CSF) circulation and compression of neurologic tissues.[6] The differential diagnosis of LM includes ruling out a wide range of malignant and benign conditions, such as congenital and degenerative lesions, infectious and autoimmune diseases and neurinoma.[7] The radiologic discrimination between metastases and neurinomas is primarily based on distinct neuroimaging features, particularly number, size and growth pattern.[7] Whereas LM are often encountered as multiple small nodules at ower spinal structures (e.g. the cauda equina), presumably due to gravity,[7] neurinomas appear as single lesions in the neuroforamen and might present at any height.[8] The clinical presentation of LM depends on the location and growth-pattern, often resulting in general symptoms such as nausea and head-aches due to interruption of the CSF flow and, later signs of myelopathy due to compression of the spinal cord.[7,9] On the other hand, neurinomas usually affect single nerve roots with typical clinical symptoms in the associated dermatomes and peripheral nerves.[10]

The present study reports the first case of a solitary leptomeningeal metastasis, mimicking a benign dumbbell-shaped neurinoma of a cervical nerve-root that was successfully treated with surgery and adjuvant radiochemotherapy (RCT).

Abbreviations: BC, breast cancer; CSF, cerebrospinal fluid; CK, cytokeratin; CT, computed tomography; ER, estrogen receptor; HER2, human epidermal growth factor receptor 2; IRS, immunoreactive score; LM, leptomeningeal metastases; MRI, magnetic resonance imaging; PR, progesterone receptor; RCT, radiochemotherapy.

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2. Presentation of case

2.1. History

A 47-year-old woman was referred to our institution because of progressive unilateral arm elevation weakness with an onset 8 weeks prior. The patient did not complain of pain or numbness. She presented with a clearly visible atrophy of the biceps and triceps muscles and substantial reduction of both muscular strength in arm elevation and elbow flexion and extension. Neurological examination revealed hyperactive biceps and triceps tendon reflexes and hypesthesia confined to C5–7 dermatomes, indicating cervical myelopathy.

The patient was diagnosed with breast cancer (BC) 9 years earlier and underwent unilateral mastectomy (pT2m, pN3a, M0, G2, R0, estrogen receptor (ER) immunoreactive score (IRS) 12, progesterone receptor (PR) IRS 4, and human epidermal growth factor receptor 2 (HER2/neu) negative). She rejected adjuvant therapy while undergoing alternative therapeutic measures. Five years later, supraclavicular lymph node-metastases on her right side were found; again, the patient declined chemotherapy. Six months later, a brain metastasis was diagnosed and RCT was initiated (corticosteroids, bevacizumab and capecitabine). The patient rejected being treated with zoledronate for suspected bone metastasis, and she aborted RCT 14 months later. Until the occurrence of the current neurologic symptoms, regular follow-up examinations showed that the patient had stable disease.

2.2. Examination

After admission, total spine and cranial magnetic resonance imaging (MRI) scans were performed and revealed the previously diagnosed right parietal intracerebral metastasis and a solitary lesion in the cervical spinal canal. The spinal tumor was located in the right neural foramen between C3/4 with an extraforaminal extension, thereby compressing the cervical plexus at levels C3–5. The osseous structures showed no erosions. Thus, the neuroimaging findings were consistent with a dumbbell-shaped neurinoma (Fig. 1a–c).

An abdominal staging computed tomography (CT) scan and a three-phase bone scintigraphy showed no further lesions. The case was reviewed at an interdisciplinary tumor board, and according to the histological findings, resection followed by adjuvant therapy was recommended.

2.3. Operation

Surgery was performed under general anesthesia using a ventral approach to the cervical spine, with the patient in the supine position. After microsurgical exposure, the tumor presented as completely intradural with the characteristic appearance of a neurinoma (Fig. 2). Following resection of the intraspinal tumor and corpectomy, the vertebral bodies C3 and C4 were replaced (Pyramesh-Cage, Medtronic GmbH, Meerbusch, Germany) and ventral plate-stabilization (CSLP, Synthes GmbH, Umkirch, Germany) from C2–5 was performed (Fig. 3).

2.4. Pathological findings

Macroscopic pathological examination revealed a soft, yellow-gray tumor. The intraoperative rapid frozen section showed disorganized nerve root tissue with cell nests of unknown entity. The definitive histological examination of the tissue revealed epithelioid cells with irregular and hyperchromatic nuclei and scattered mitosis arranged in nests embedded in stroma rich in collagen fibers (Fig. 4a). Immunohistochemical staining revealed that the tumor cells were positive for cytokeratin (CK) 8 (Fig. 4b), 85% were positive for ER (Fig. 4c), 10% for HER2/neu (Fig. 4d) and 2% for PR. There were no CD3, CD20, S-100 or vimentin-positive cells. The Ki67 proliferation index was 20%. Fluorescence in situ hybridization showed no positive signals for HER2/neu. The final histopathology

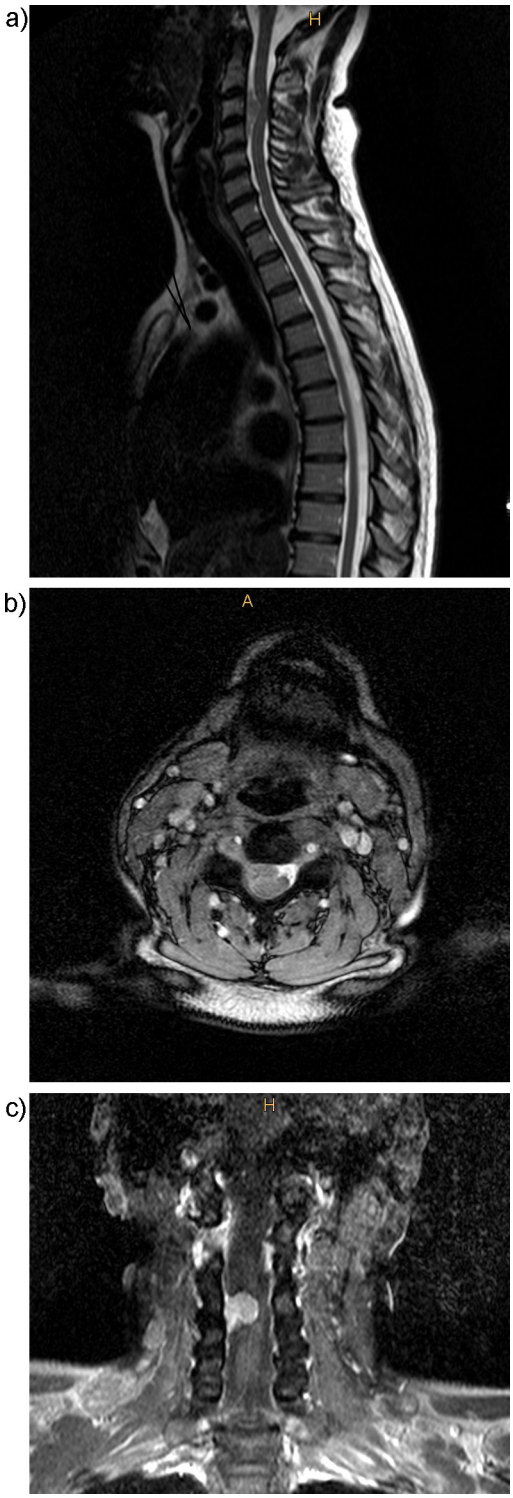

Fig. 1. (a)–(c) MRI of the cervical spine with the tumor (a) mid-sagittal plane T2/TSE; the tumor is compressing the spinal cord in a triangular shape, expanding from the second to the fourth cervical vertebral body (C2–C4), (b) axial plane B-TFE/TRA; the dumbbell-shaped intra-neuroforaminal growth of the tumor on the right side is depicted. The spinal cord is shifted and compressed and (c) coronal plane T1/TSE. The tumor is visible at the right side, located at the height of the third and fourth cervical vertebral body (C3–C4).