Primary malignant myopericytoma with cancer cachexia
Report of the first case and review of literature

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Abstract

Rationale: Malignant myopericytoma is extremely rare, with a few cases described in the English literature.

Patient concerns: This novel study aimed to report a case of malignant myopericytoma with cancer cachexia arising in the left armpit. Also, it presented a review of the English literature regarding primary malignant myopericytoma, aiming to clarify the clinical features and potentially curative treatment. A 56-year-old male presented with an ulcerated and smelly mass involving her left armpit. The patient had obvious symptoms of cancer cachexia, including emaciation, anemia, and lower extremity edema.

Diagnoses: Computer tomography (CT) scan demonstrated a mass in the left armpit, with no evidence of metastasis according to the chest CT, abdominal ultrasound, and emission CT. The patient underwent a core biopsy of the mass, and a diagnosis of malignant myopericytoma was rendered.

Interventions: He received 2 standard courses of theprubicin combined with ifosfamide chemotherapy regimen with no tumor response. Then, he subsequently underwent complete excision of the tumor.

Outcomes: The symptoms of cancer cachexia disappeared gradually after operation. Recurrence and metastasis were not shown during follow-up for 5 years.

Lessons: Myopericytoma is generally considered benign with an indolent clinical course, and a few reports have described malignant myopericytoma in the literature. No standard treatment is available, and complete surgical excision of the lesion may be the only potentially curative treatment. The efficacy of chemotherapy and radiation is uncertain.

Abbreviations: CT = computer tomography, HMB-45 = human melanoma black-45, IFO = ifosfamide, SMA = muscle-specific actin, THP = theprubicin, WHO = World Health Organization.

Keywords: armpit, cancer cachexia, chemotherapy, malignant myopericytoma, surgical treatment

1. Introduction

The concept, myopericytoma, was described for the first time by Granter et al in 1998.[1] The World Health Organization (WHO) officially recognized the term “myopericytoma” in 2002 and referred to it as a member of the pericytic group in the Classification of Tumors of Soft Tissue and Bone.[2] Myopericytoma is a rare tumor that is predominantly located in the skin and superficial soft tissues of extremities, such as arm, thigh, leg, foot, and neck.[3] Most myopericytomas are benign in nature. Malignant myopericytoma is exceptionally rare, with only 8 cases described in the literature.[4–7] This study reported the ninth case, which is the first in the left armpit with a long history of 31 years.

2. Case report

A 56-year-old man presented to The Shaanxi Province Oncology Hospital with “an enlarging painful mass in his left armpit” in 2012. He had initially presented 31 years earlier with a painless nodule in the subcutaneous tissue of the left armpit. No other clinical symptoms were present at that time. The patient did not receive any treatment for 31 years. The nodule grew slowly. The mass expanded rapidly and ulcerated in the last year. Two months ago, the patient suffered from pain in the left shoulder during sports. At the same time, obvious symptoms of cancer cachexia, including emaciation, anemia, and lower extremity edema, were appeared. He had no medical history of surgery and trauma in the left armpit.

Physical examination showed an ulcerated and smelly mass in the left armpit (Fig. 1). The computer tomography (CT) scan demonstrated a mass of 6.1 × 5.3 cm^2 in the left armpit (Fig. 2), with no evidence of metastasis according to the chest CT, abdominal ultrasound, and emission CT. Other tests showed anemia and hypoproteinemia. He underwent biopsy of an
enlarging mass, and a diagnosis of malignant myopericytoma was rendered (Fig. 3). Immunohistochemical analysis revealed that the tumor had strong cytoplasmic reactivity for muscle-specific actin (SMA) and was negative for desmin and human melanoma black-45 (HMB-45).

The patient received 2 standard courses of thetrubicin (THP) combined with ifosfamide (IFO) chemotherapy regimen (THP: 60mg on day 1; IFO: 2g on days 1–3) with no tumor response according to the CT findings. He subsequently underwent complete resection by a surgical technologist. At the time of surgery, the mass was found to be arising from the axillary vein (Fig. 4). Postoperative pathology indicated that the tumor size was 6.5 × 5.5 cm², the margin was negative, and the lymph node was negative. After the surgery, clinical symptoms gradually disappeared. The follow-up was performed at 3 months in the first year, 6 months in the second year, and 1 year after 3 years. The last follow-up was March 2017. According to the clinical symptoms and imaging and laboratory tests, the patient did not show recurrence and metastasis during the follow-up for 5 years (Fig. 5).
Myopericytomas are generally considered benign with an indolent clinical course, and few reports have described malignant myopericytomas in the literature. The clinical and pathologic features of malignant myopericytomas are distinct from those of their benign counterparts. Myopericytomas are usually diagnosed after the age of 40, with a male-to-female ratio of approximately 2:1. They are seen in middle-aged adults. Although they are generally considered benign with an indolent clinical course, they have the potential to recur locally with distant metastases.

A study by Mentzel et al. [15] reported 1 patient in 2006, and Mainville et al. [16] reported 1 patient in 2012. Holling et al. [17] reported 1 patient in 2015 out of the 8 reported patients, 4 were males and 4 were females. The tumors were located in the extremities, neck, mediastinum, heart, and spine. All the patients underwent surgical excision, including 3 wide excisions and 1 marginal excision. The tumors were located in the extremities, neck, mediastinum, heart, and spine. All the patients underwent surgical excision, including 3 wide excisions and 1 marginal excision.

The largest series was reported by McMenamin and Fletcher [14] in 2002 and included 5 patients. The clinical and pathologic features of the 5 previously reported cases of malignant myopericytoma are summarized in Table 1. The clinical features and anatomic site were different. This case had the longest clinical history reported in the literature till now, with a follow-up of 15 years. The patient received 2 cycles of chemotherapy and radiotherapy. Four patients did not show recurrence, including 3 wide excisions and 1 marginal excision. The tumors were located in the extremities, neck, mediastinum, heart, and spine. All the patients underwent surgical excision, including 3 wide excisions and 1 marginal excision.

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4. Conclusion

Complete surgical excision of the lesion might be the only potentially curative treatment. The efficacy of chemotherapy and radiotherapy is currently uncertain. However, the currently reported cases of malignant myopericytomas, including tumors with phenotypic features of cutaneous adult myopericytoma, are rare. Therefore, further studies are needed to clarify the optimal treatment for this rare tumor.

3. Discussion

In 1996, Requena et al. [8] suggested using the term "myopericytoma" as an alternative name for some cutaneous adult myopericytomas. "Myopericytoma" was officially recognized by the WHO Classification of Tumors of Soft Tissue and Bone in 2012 [13]. Malignant myopericytoma is one of the members of the perivascular tumor family, including myofibromas, angioleiomyomas, glomus tumors, and angiosarcomas. Myopericytomas are generally considered benign with an indolent clinical course, and a few reports have described malignant myopericytomas in the literature. The clinical and pathologic features of malignant myopericytomas are distinct from those of their benign counterparts. Myopericytomas are usually diagnosed after the age of 40, with a male-to-female ratio of approximately 2:1. They are seen in middle-aged adults. Although they are generally considered benign with an indolent clinical course, they have the potential to recur locally with distant metastases.

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