Clinical Case Report

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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 are 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 following 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 $6.1 \times 5.3\,cm^2$ 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

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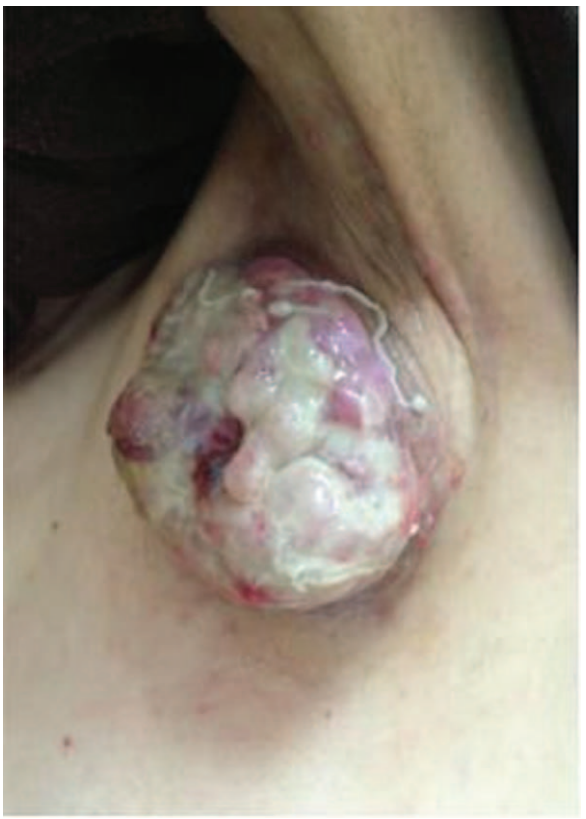

Figure 1. Physical examination showed a mass in the left armpit.

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 theprubicin (THP) combined with ifosfamide (IFO) chemotherapy regimen (THP: 60 mg on day 1; IFO: 2 g 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 $6.5 \times 5.5\,cm^2$, the margin was negative, and the lymph node was negative. After the surgery, clinical symptoms gradually

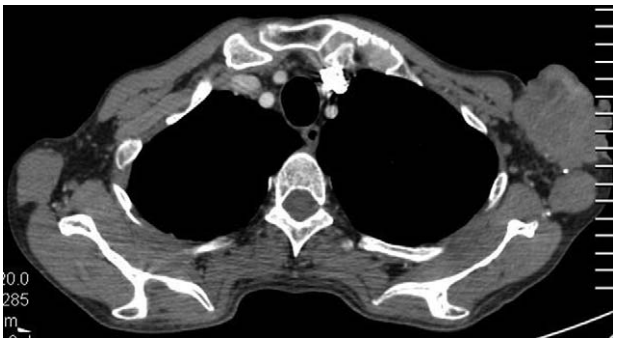

Figure 2. Computer tomography scan demonstrated a $6.1 \times 5.3\,cm^2$ mass in the left armpit.

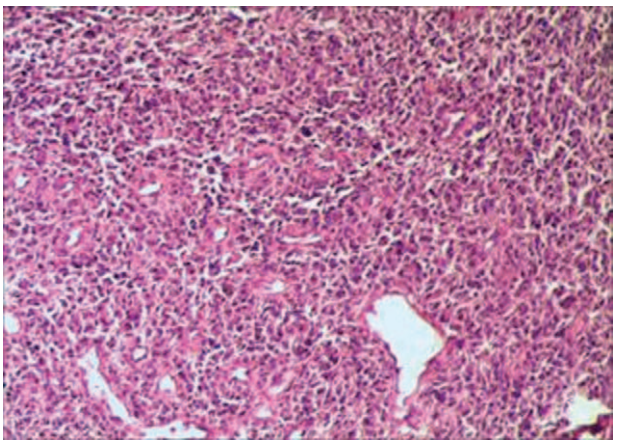

Figure 3. Proliferation of spindle-shaped myopericytic cell with characteristic concentric distribution (H&E, 10 x 10).

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).

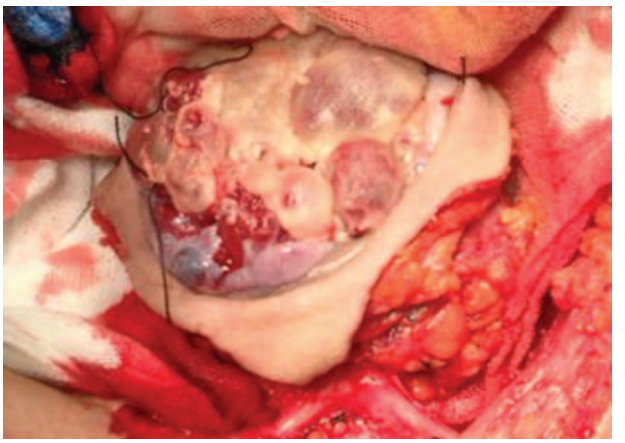

Figure 4. Appearance of the resected tumor.

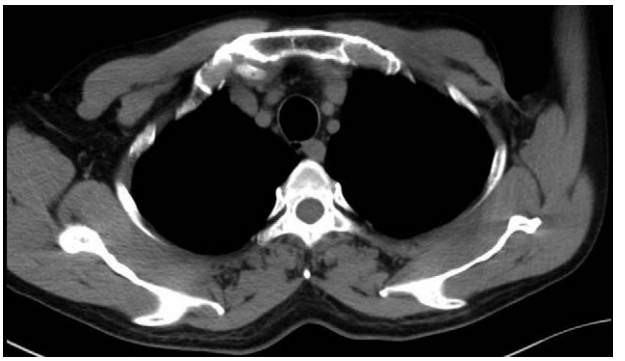

Figure 5. Computer tomography scan demonstrated no mass in the left armpit.

This case report was approved by the ethics committee, and informed consent form was obtained from the patient.

3. Discussion

In 1996, Requena et al[8] suggested "myopericytoma" as an alternative name for some cutaneous adult myofibromas. Later, in 1998, Granter et al[9] described a group of vascular tumors, including tumors with phenotypic features of cutaneous adult myofibroma, glomus tumors, glomangiopericytoma, and myopericytoma. The term "myopericytoma" was officially recognized by the WHO Classification of Tumors of Soft Tissue and Bone in 2012.[10]

Myopericytoma is one of the members of the perivascular tumor family, including myofibroma, angioleiomyoma, glomus tumor, and myopericytoma.[11] So far, the cause of myopericytomas is unclear. Some authors suggested that the cause seemed to be related to the Epstein–Barr virus.[12] The immunohistochemical analysis showed that SMA was positive, supporting a pericytic/myofibroblastic differentiation. Desmin positivity was seen in rare cases. Clinically, most myopericytomas 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.[13]

Malignant myopericytoma are exquisitely rare. They show aggressive clinical behavior and poor prognosis. Only 8 cases have been reported in the literature to date. The largest series was reported by McMenamin and Fletcher[14] in 2002 and included 5 patients. 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 extremities, neck, mediastinum, heart, and spine. All the patients underwent surgical excision. Four patients did not show recurrence, including 3 wide excisions and 1 marginal excision and radiotherapy. Four patients developed metastases, and 3 died of disease within 1 year. The clinical features of the 8 previously reported cases in addition to this one are summarized in Table 1.

This study reported a case of malignant myopericytoma arising in the left armpit, the histopathologic and immunophenotypic features of which were similar to those previously reported, but clinical features and anatomic site were different. This case had the longest clinical history reported in the literature till now, with obvious cancer cachexia symptoms. No standard treatment exists for malignant myopericytoma. The patient received 2 cycles of THP combined with IFO chemotherapy regimen with no tumor response. Consequently, chemotherapy might not be necessary. Complete surgical excision of the lesion might be the only potentially curative treatment, and the timing and frequency of follow-up are essential.

4. Conclusion

Myopericytoma are generally considered benign with an indolent clinical course, and a few reports have described malignant myopericytoma in the literature. The clinical and pathologic features of these cases of malignant myopericytoma were summarized in this study. 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. However, the currently reported rare cases may not be sufficient to allow the clinical outcome to be fully evaluated, requiring the involvement of large samples.

Table 1

Clinical characteristics of all reported cases of malignant myopericytoma.

Case	First authors	Sex/Age	Site	Presenting symptoms	Past medical history	Mass size	Treatment	Outcome
1	McMenamin and Fletcher[14]	F/81	Left side of neck	Rapidly growing painless mass of 2 mo	Excised melanoma at the same location	20 mm	Marginal excision	Metastases to the liver at 14, alive with disease at 24 mo
2	McMenamin and Fletcher[14]	M/46	Left thigh	Painful mass of unstated duration	No	130 mm	Marginal excision, postoperative radiotherapy	Widespread metastasis at 6 mo, died of disease at 7 mo
3	McMenamin and Fletcher[14]	M/19	Heel of right foot	Growing painful mass of 2 mo	Poorly circumscribed glomus tumor of right foot 6 y ago, radiotherapy 5 y ago, excised "glomus" tumors 4 y ago	40 mm	A right below-knee amputation	Metastases and died within 1 y (precise time are not available)
4	McMenamin and Fletcher[14]	F/80	Left arm	Painful mass of unstated duration	No	15 mm	Wide excision	No recurrence and metastasis at 20 mo
5	McMenamin and Fletcher[14]	F/67	Superior mediastinum	Superior vena cava syndrome, skin metastases developed rapidly	No metastatic deposit	100 mm	Excised cutaneous metastatic deposit	Dead of respiratory failure within less than 1 mo
6	Mentzel et al[15]	F/61	Lower leg	Subcutaneous mass of unstated duration	No	Infiltrative mass (size is unavailable)	Wide excision	No recurrence and metastasis at 3 y
7	Gisele N	M/52	Left atrial wall	Progressive blackening in the left field of vision	Wide excised malignant melanoma of left arm and right leg 23 y ago by radiotherapy	53 mm	Marginal excision of cardiac mass, excised metastatic brain tumor followed by radiotherapy	Alive with disease at 8 mo
8	Markus Holling	M/38	Intraspinal dorsal calf	Progressing painful in the right during sport of 5 mo	No	Unavailable	Complete resection	No recurrence and metastasis at 18 months
9	This study	M/56	Left armpit	Pain of left shoulder mass and cachexia	No	610 mm	Complete resection	No recurrence and metastasis at 5 y

F = female; M = male.

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