CASE REPORT

Primary abdominal muscle lymphoma

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SUMMARY

Primary skeletal muscle lymphoma accounts for <1% of all lymphomas; a fraction of these arise within the abdominal muscles. Here, we describe the case of an 84-year-old woman who presented with multiple painful abdominal masses that proved to be diffuse large B-cell lymphoma and we discuss the main features of the disease. Clinical differential diagnosis from soft tissue sarcoma can be extremely difficult. Therefore, accurate pathological diagnosis including optimal tissue processing and immunohistochemical staining is vital to ensuring proper treatment.

BACKGROUND

Primary lymphoma is not usually included in the differential diagnosis of a muscle mass, leading to an unnecessary, non-curable surgical excision. This case report reminds both the physician and the pathologist that keeping an open mind is the cornerstone of any diagnosis.

CASE PRESENTATION

An 84-year-old woman presented with a 2-month history of a painful swelling in her lower abdomen. On examination, the abdomen was soft with diffused tenderness. Palpation revealed three masses in the umbilical and left lateral regions. No B symptoms (fever, night sweats, weight loss) were reported. Her medical history was notable for ischaemic heart disease with an acute coronary syndrome approximately 30 years before her presentation. She was an ex-smoker with a total of 20 pack-years and had abstained from smoking since the age of 55. She did not drink alcohol or use illicit drugs. Her medication included aspirin, ramipril, metoprolol and omeprazole.

INVESTIGATIONS

MRI performed with the administration of intravenous contrast material revealed three enhancing masses measuring 12×9.3×3 cm, 3×4×1.8 cm and 4.3×3.1×2.2 cm, respectively, arising from the vertical anterior and left anterolateral abdominal muscles (figure 1). Core biopsy was performed and the pathological examination confirmed the diagnosis of diffuse large B-cell lymphoma (DLBCL). The immunohistochemistry staining was positive for CD19, CD20, sIgM and bcl-6 and negative for CD5, CD10 and MUM1, further classifying it as germinal center B-cell like according to the Hans algorithm.¹ Subsequent staging did not reveal other sites of disease. Her Eastern Cooperative Oncology Group (ECOG) performance status was 3 and the international prognostic index was 3, indicating high intermediate risk.

DIFFERENTIAL DIAGNOSIS

Skeletal muscle lymphoma is a rare and clinically diverse disease, making the diagnosis challenging.^{2,3} The main differential diagnoses are tumours of mesenchymal origin such as sarcomas and metastatic carcinomas to soft tissue. Clinical features such as pain and tenderness, elevated levels of lactate dehydrogenase, lymphadenopathy and ipsilateral extremity swelling can help in the diagnosis of primary extremity lymphoma.³ The MRI features of skeletal muscle lymphoma have been described in a retrospective analysis of 20 pathologically proven cases. Muscle lymphomas are isointense to normal muscle on T1-weighted images and have increased or intermediate signal intensity compared with fat on T2-weighted images, with homogeneous diffuse enhancement in most cases. Multiple muscular masses were reported in 14 of 20 patients.⁴ However, clinical history, examination and MRI are insufficient to differentiate between sarcoma and primary muscle lymphoma.⁵ Accurate diagnosis requires pathological examination; core biopsy established the diagnosis with a sensitivity of 93% in one case series.² Most soft tissue lymphomas are of B-cell origin, most commonly DLBCL. Uncommon histological subtypes include anaplastic large-cell lymphoma, low-grade histology, T-cell lymphomas and Burkitt's lymphoma.⁶

TREATMENT

Treatment usually includes chemotherapy with or without radiotherapy. Surgical excision followed by chemotherapy has also been successfully applied in a patient with abdominal wall lymphoma.⁷ The patient was treated with cyclophosphamide 750 mg/m² (day 1), adriamycin 25 mg/m² (day 1),

Figure 1 Axial T1-weighted image of an isointense to normal muscle mass (asterisk).

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oncovin 1.4 mg/m² (day 1), prednisone 40 mg/m² (days 1–5) and rituximab 375 mg/m² (day 1) every 21 days (R-mini-CHOP-21). Immunochemotherapy has become the standard for the treatment of aggressive lymphomas in elderly patients⁸ and was used in our case, albeit with a lower dose of adriamycin due to age-related concerns.

OUTCOME AND FOLLOW-UP

After the first cycle of immunochemotherapy the patient reported markedly improved symptoms. No masses could be palpated and the ECOG performance status improved to 1. She completed six cycles uneventfully and attained a complete remission as evidenced by repeat MRI. Currently, the patient is in remission 10 months after the initial diagnosis. Long-term disease-free survival is achieved by a majority of patients in most retrospective case series. Specific prognostic factors for skeletal muscle lymphoma have not been described.

DISCUSSION

Although extra-nodal disease is fairly common among non-Hodgkin's lymphomas, primary soft tissue lymphoma is a rare entity with an incidence rate of <1% of all lymphomas.⁹ In a literature review, Chim *et al*¹⁰ identified 32 cases of primary skeletal muscle lymphoma. The median age was 64.5 years (range: 15–87 years), with a predominance of male patients. The most common sites of occurrence were the extremities; no cases of abdominal wall lymphoma were reported.¹⁰ In a similar fashion, in a recent literature review of soft tissue lymphomas reported in the English literature only one case of abdominal wall lymphoma was identified during the past 20 years.⁶ On the contrary, Lanham *et al*¹¹ reported in their series that the thoraco-abdominal wall was the second most common (n=22) site of occurrence among 75 patients with primary soft tissue lymphoma. Skeletal muscle lymphomas are associated with previous injury or surgery; however, such an association did not apply to our patient.¹²

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- ▶ Primary skeletal muscle lymphoma accounts for <1% of all lymphomas; a fraction of them arise from the abdominal muscles.
- ▶ Clinical differential diagnosis between muscle lymphoma and sarcoma can be challenging.
- ▶ Accurate and early pathological diagnosis avoids inappropriate resection and permits institution of correct treatment.
- ▶ Successful remissions can be attained even in older patients using modern immunochemotherapeutic regimens albeit with the necessary dose modifications for cardiac and other toxicities.

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