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Case report

Myeloid sarcoma (chloroma) a rare tumour of the chest wall

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Abstract

Introduction: **Myeloid sarcoma (previously chloroma or granulocytic sarcoma)** is a very rare malignant neoplasm associated with myeloproliferative neoplasms. This type of tumour can be located anywhere outside the bone marrow.

Aim: To describe the successful treatment and diagnostic of myeloid sarcoma.

Case study: Retrospective case review.

Results and discussion: This paper presents a case of myeloid sarcoma in a 50-year-old male. Patient presented due to pain lasting for 2 months in the left anterior axillary line region and a rapidly growing nodular lesion on the anterior chest wall. The lesion involved an extensive area of the ribs, causing rib destruction. Surgery was performed causing patients relief and chance of collecting histopathological tissue.

Conclusions: In myeloid sarcoma patients, surgical treatment does not only bring relief in symptomatic patients but also provides material for histopathological examination, which is necessary to confirm the diagnosis. An important part of the diagnosis is the implementation of a wide panel of immunohistochemical tests.

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1. INTRODUCTION

Myeloid sarcoma (MS, previously chloroma or granulocytic sarcoma) is a very rare malignant neoplasm developing from myelocytic lineage cells. This solid tumour localizes outside the myeloid cavity and co-occurs most commonly with acute myeloid leukaemia (AML; approximately 2.5%–9.0% of cases).[1] First described by Burns in 1811, the neoplasm was named chloroma due to its greenish colour resulting from myeloperoxidase reaction when exposed to air.[1,2] Since the neoplasm may also display other colours, it was renamed to granulocytic sarcoma in 1966.[2] The neoplasm is most commonly diagnosed in adults between 46 and 59 years of age, and almost 60% of cases are male. Apart from acute myeloid leukaemia, it can also accompany other myelodysplastic and myeloproliferative syndromes.[3] The lesion can occupy any area, most commonly the skin, soft tissues, lymph nodes, bones; a large proportion of cases remains asymptomatic.[4]

Diagnosis is a significant challenge and must be based on the entire clinical picture, with consideration imaging studies, histopathology, immunohistochemistry, cytogenetics.[5] Therapy includes chemotherapy, but in case of resistance to treatment or for tumours causing organ dysfunction – radiotherapy and surgery are used. Allogeneic hematopoietic cell transplantation (allo-HCT) and novel targeted therapies have recently been reported to have good therapeutic effect[6].

2. AIM

Hence the aim of this retrospective case report is to describe a case of successful treatment and diagnostic of myeloid sarcoma.

3. CASE STUDY

In June 2020, a 50-year-old male patient presented to the Department of Thoracic Surgery and Transplantation of the Pomeranian Medical University in Szczecin, Poland due to pain lasting for 2 months in the left anterior axillary line region and a rapidly growing nodular lesion on the anterior chest wall, present for 3 weeks. Physical examination revealed a lesion of about 6 cm in diameter on the left side, lateral to the pectoralis major in the anterior axillary line, compressive soreness and inflammatory reaction around the lesion. No other abnormalities were found. The family history of neoplastic and hematological diseases was negative. The patient did not report any concomitant diseases. The diagnostics was enhanced to include chest wall ultrasound, which showed a 7.5 × 5.0 cm lesion probably originating from the rib and extensive infiltrative changes. Fine-needle aspiration biopsy of the lesion was performed. The cytology was suggestive of lymphoma. There were no suspicious oncological lesions in the abdominal cavity or involvement of the lymph nodes in the supraclavicular fossae. Blood count showed elevated WBC (white blood cells) count with moderate neutrophilia, basophilia and thrombocytosis. The patient's plasma carcino-embryonic antigen (CEA) concentration was within normal range. A computed tomography (CT) of the chest was performed, which showed an extensive pathological mass 10.0 × 7.0 × 9.5 cm on the left side of the chest (Figure 1). A resection of the chest wall tumour with a fragment of rib IV and V was performed (Figures 2 and 3). On intraoperative evaluation the tumour was soft and friable, clustered in structure, contained a fluid space, causing destruction of rib IV and infiltrating the intercostal space and rib V. Intraoperative histopathological evaluation was suggestive of a lymphoid malignancy. The postoperative period was uncomplicated and the patient was discharged home on day 4 in good condition with further pain relief.

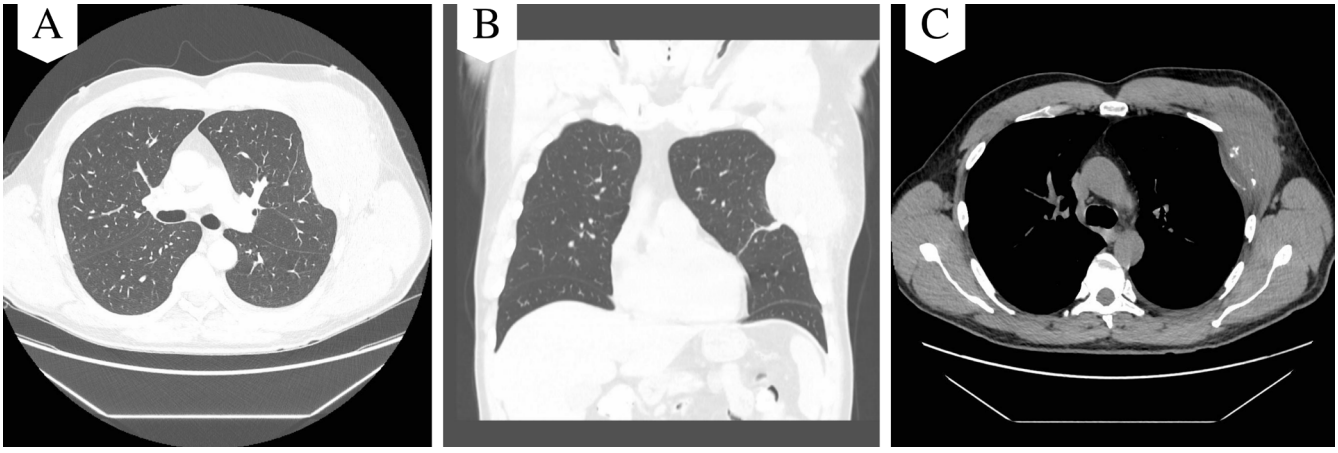

Figure 1. (A) CT scan of the chest: an extensive pathological mass is present on the left side of the chest, centred on the destruction and widening of the outlines of the anterior segment of the 4th rib suggesting the starting point of the lesion. The tumour also involves the 5th rib where periosteal reactions are present. The tumour infiltrates the pectoralis major muscle, with obliteration of its outlines. (B and C) The tumor infiltrates segments III and IV of the left lung to a depth of up to 23 mm, with the presence of few small nodules in the adjacent parenchyma and band-like airless lesions in segments IV and V of the left lung.

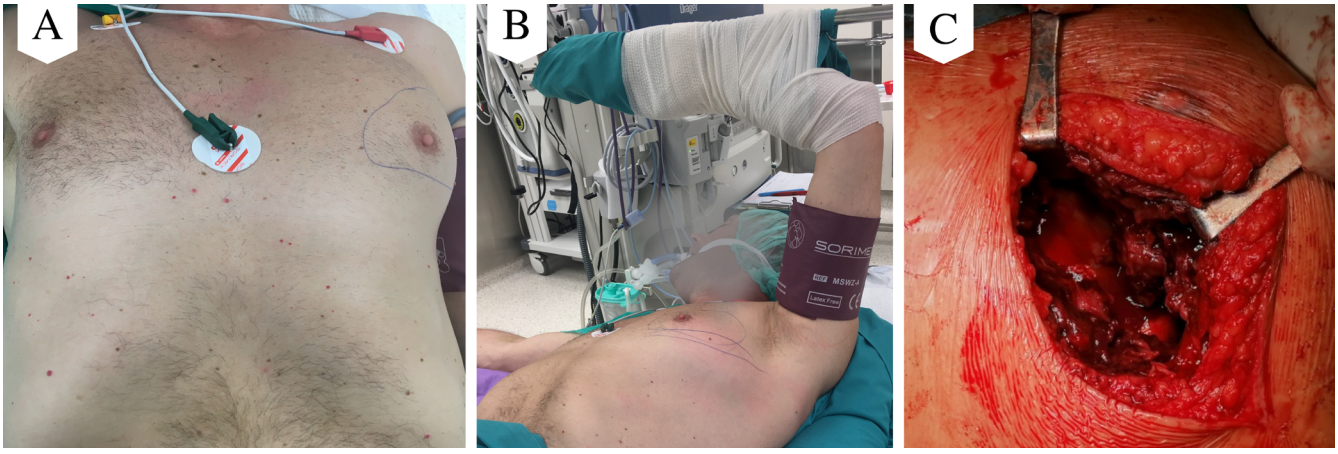

Figure 2. (A) Preoperative view of the chest; (B) Position of the patient on the operating table; (C) View of the operating field.

Histopathological examination of the obtained tissue material showed a solid infiltrate of large blastic cells and locally numerous myelocytes and granulocytes with acidophilic granules (Figure 4).

Immunohistochemical examination of the material showed a positive reaction with CD45/LCA antibody. Reaction with CD20, CD79a, CD3, CD34, CD138 antibodies was negative and CD56 reaction was found in some tumour cells. The histopathology and immunohistochemical findings were consistent with MS.

The patient underwent complex hematological procedures based on chemotherapy, immunotherapy and allo-HCT being still due to observation.

4. DISCUSSION

MS according to WHO is a tumour containing myeloblasts with or without features of maturation, localizing outside the bone marrow and disrupting the tissue architecture.[1]

More than a dozen cases of MS localization within the ribs have been reported in the literature.[7–12] The overall tumour incidence is 2 per 100 000 for adults and 0.7 per 100 000 for children,[13] and isolated tumours are 2 cases per million adults.[1] MS often co-occurs with AML, myeloproliferative neoplasms (MPN), myelodysplastic syndrome (MDS), MPN/MDS, or blast crisis in the course of chronic myeloid leukaemia.[1] It is estimated that in 80%–90% of patients with primary MS, AML is manifested upon diagnosis or at a later stage.[3] In almost every case of MS rib tumour known to the literature, myeloproliferative neoplasms of the bone marrow were found to coexist before or after the diagnosis of sarcoma. In the work of Takahashi and Pomeranz, as well as in the case presented herein, MS cooccurred with CML.[7–12]

This paper presents a case of a man aged 50, with a MS in the chest wall. Previous papers of MS in the ribs described varied age and sex of initial diagnosis of MS.[3,7–12] MS can occupy any area, most commonly the skin, soft tissues, lymph nodes, and bones.[2,3] Infiltration of the skin can also be referred to as leukemia cutis and is associated with poor prognosis. Manifestation of isolated skin involvement is related with pre-

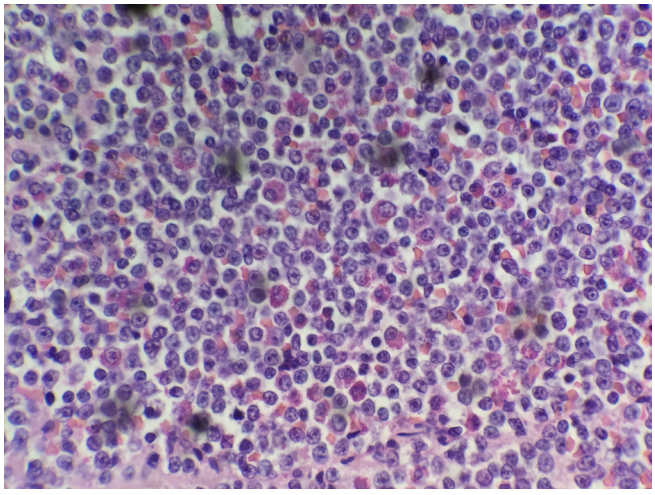

Figure 3. Macroscopic image of fragments of the removed tumour with a visible greenish hue.

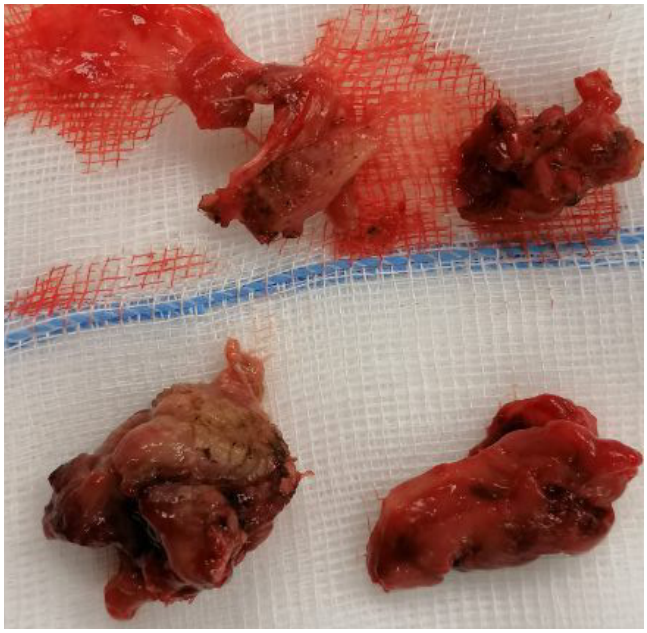

Figure 4. Histopathological examination of the obtained tissue material showed a solid infiltrate of large blastic cells and locally numerous myelocytes and granulocytes with acidophilic granules (H&E staining; magnification ×200).

ceding diagnosis of systemic leukemia by several months or years, depending on the type of leukaemia.[17] The tendency to occupy bone and periosteum is often due to direct infiltration of the tumour from the bone marrow, as suggested by the CT description in the case in question.[2,3] Isolated tumours cause symptoms depending on their location, mass effect, organ infiltration, and remain asymptomatic in about 50%.[2,6] In the presented case, as well as in the cases described by Guermazi, Raucci or Takahashi, the predominant symptom was left-sided chest pain. Hermann reports flu-like symptoms in an adult, and fever, underweight and tachypnoea in an infant.[7–9]

Extensive imaging, histopathological, molecular, cytogenetic diagnostics should be performed to make the diagnosis. Imaging is one of the essential elements of differentiation. The use of ultrasound is justified in lesions located superficially e.g. on the skin, testes or – as it is in the case in question – in the chest wall.[2] MS appears on CT as an isodense mass in relation to skeletal muscle with homogeneous contrast enhancement.[3] In the presented case as well as in the works of Guermazi, Hermann, Raucci, Takahashi, a mass causing rib destruction was found. Furthermore, the last two authors described the rib as the starting point of the lesion. In the imaging studies of the presented patient, a radiograph of a 10-month-old girl and a CT scan of the chest of a 30-year-old patient described a broadening of the rib outlines.[7–10] CT and magnetic resonance imaging (MRI) may exclude the presence of hematomas and abscesses occurring in the course of AML.[6]

The presence of pathological changes in imaging studies in the presence of coexisting bone marrow malignancy may suggest the presence of MS. In patients without symptoms suggestive of marrow pathology, the presence of a soft tissue tumour will require a radiologically guided core needle biopsy and extended immunohistochemical studies. In the patient presented here, a fine-needle biopsy was used; however, core needle biopsy performed under imaging guidance is more effective.[1,8]

Myeloblasts are predominant in the histological picture of the MS. More mature cells with differentiation into radiolucent or neutrophils can also be seen, as in the case presented.

If MS arises from a myeloproliferative neoplasm, erythroid cells and megakaryocyte precursors may also be observed.[4] MS can be divided into five subtypes: immature granulocytic sarcoma, differentiated granulocytic sarcoma, monoblastic sarcoma, monocytic sarcoma, myelomonocytic sarcoma.[16]

As many as 47% of MS cases are incorrectly diagnosed, most commonly as diffuse large B-cell lymphoma.[3] Therefore, i.a. immunohistochemical examination is essential to correctly establish the diagnosis.[5] Positive reaction to markers MPO, CD43, CD68, CD15, CD117, CD 99, CD34, TdT, CD56, CD61, CD30, CD4, glycophorin A and lysozyme present on MS cells are important for diagnosis. In the case described here, a positive reaction to CD56 and a negative reaction to CD34 were reported, with prevalence according to Pileri at 13.0% and 43.4%, respectively.[4,13,14] To exclude lesions of B or T cell origin, the absence of a reaction should be demonstrated using CD20, CD45RO, CD79a, CD3 antibodies.[6] Immunohistochemical studies are essential in the differentiation of small cell tumours into lymphoid malignancies, small cell sarcomas such as Ewing sarcoma, rhabdomyosarcoma, synovial sarcoma, MS and also medulloblastoma, neuroblastoma.[4,15]

The prognosis of MS is poor and untreated patients die within a short time. An untreated isolated MS tumour transforms to AML after about a year.[1] The average survival rate for MS is 7.5 months after diagnosis.[2] There are no clear conclusions on prognostic factors like gender, age, location, MPN and history of MDS. The probability of higher survival rates in patients treated with systemic chemotherapy or after allo-HCT is observed.[1,2,4]

There are no established guidelines for the treatment of MS, however, the best results are observed with systemic treatment regardless of AML co-morbidity or after complete resection of an isolated tumour. Chemotherapy with anthracycline with cytarabine is preferred. Systemic treatment prolongs disease-free survival time but there is no evidence of a protective effect against MS transformation to AML.[1,3,6] Radiotherapy and surgical treatment are important in the local treatment of MS. Surgery is indicated in symptomatic patients and before starting chemotherapy, particularly to confirm the diagnosis based on surgical specimens. In the MS cases described by Raucci and Takahashi, as it is the case in the patient presented here, the main purpose of the surgery was symptomatic treatment and the need to collect material for histopathological examination to establish a definitive diagnosis of the disease. Recent reports recommend allo-HCTas first-line treatment in MS patients after induction of a remission.[3,7,8,13] Novel targeted therapy using tyrosine kinase inhibitors (TKI) is also a promising treatment option.[1]

5. CONCLUSIONS

(1) Surgical treatment provides symptomatic patients with relief and enables the collection of material for histopathological examination, which is decisive in the challenging diagnosis of MS.
(2) Haematological status of patients with isolated MS should be closely monitored due a risk of onset of myeloproliferative neoplasm.
(3) Further treatment with therapy using chemotherapy, immunotherapy, allo-HCT and radiotherapy provide promising results.

Conflict of interest

The authors declare that they have no conflict of interest.

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