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clinical practice

A rare case of myeloid sarcoma presenting as anal fissure

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SUMMARY: **A rare case of myeloid sarcoma presenting as anal fissure.**

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Myeloid sarcoma is a tumor composed of myeloblasts occurring at an extramedullary site. It may develop in patients with acute myeloid leukemia, myeloproliferative or myelodysplastic syndrome, sometimes preceding onset of the systemic disease. Frequent sites of myeloid sarcoma are bones or various soft tissues. Gastrointestinal involvement is very rare. We report a unique case of myeloid sarcoma presenting as a painful anal fissure, in a patient with a history of acute myeloid leukemia. The diagnosis was achieved by a surgical excisional biopsy and immunoistochemical staining.

KEY WORDS: Anal fissure - Myeloid sarcoma - Excisional biopsy.

Introduction

Myeloid sarcoma is a tumor composed by immature myeloid cells or myeloblasts at an extramedullary site. It has also been termed chloroma, granulocytic sarcoma, myeloblastoma neoplasm or extramidollary myeloid tumor (1). Myeloid sarcoma may develop in patients with acute myeloid leukemia, myeloproliferative neoplasms or myelodisplastic syndrome, sometimes-occurring months or years before the initial manifestation of the systemic disease. It is usually diagnosed in bones, soft tissue and reproductive organs (2). Gastrointestinal tract is very rarely involved (1) and isolated anal location is unique. We report a case of myeloid sarcoma located in the anal canal and clinically presenting as a painful anal fissure.

Case report

A 65-year-old man complaining of severe anal pain was referred to our Department of Surgery. He complained of anal pain, especially during defecation, starting few weeks earlier. His family history was insignificant. He denied any abdominal symptoms.

A previous history of acute myeloid leukemia was referred. Six months earlier, he was submitted to a peripheral blood smear for marked asthenia, which showed dysplastic changes in all hematopoietic lineages including hypogranulation of the neutrophils. Bone marrow aspirate yielded a diagnosis of mielodysplastic syndrome (refractory anemia with 2% blasts) with karyotype 46, XY. Recombinant human erythropoietin (Epoetin alfa at dose of 40.000 U.I. weekly) was used without clinical or hematologic benefit. After few months of follow-up, because of worsening in general conditions with appearance of fever, weight loss and weakness, the patient underwent a new bone marrow aspirate that allowed diagnosis of acute myeloid leukemia 46 XY with inv (9), FLT3 w.t., NPM1 mut.

At the time of our observation in the Department of Surgery, upon physical examination a typical anal fissure with a skin tag was observed. Colonoscopy was negative. Surgery was scheduled and a sphincterotomy was performed. During the operation, the skin tag was removed.

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The pathological examination of the removed skin tag revealed a fibro-epithelial polyp.

The specimen was fixed in formalin 10% buffered and embedded in paraffin. The sections were stained with hematoxylin-eosin and Giemsa. Immunostaining was performed using paraffin sections by Envision methods (DAKO) with the following antibodies: myeloperoxidase (MPO), CD34, CD43, CD45, CD20, CD3, terminal deoxynucleotidyl transferase (TdT) (all by DAKO), according to the manufacturer's instructions. Hystologically, the connectival tissue was diffusely infiltrated by atypical cells of medium and large size, which showed immature myeloid elements (myeloblasts and promyelocytes) (Figure 1). On immunohistochemistry sections, CD43 and CD45 were diffusely positive, MPO was positive in about 40-50% of the neoplastic cells (Figure 2),

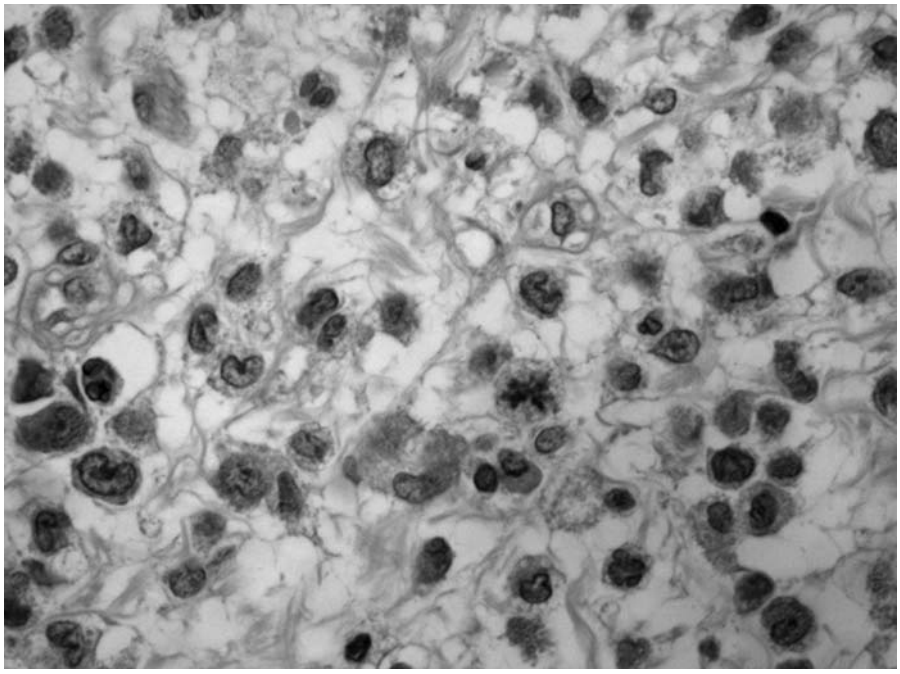

Fig. 1 - Pathological findings (hematoxylin-eosin 63x): infiltration by large cells and multinucleated giant elements with abundant eosinophilic cytoplasm (myeloblasts and promyelocytes).

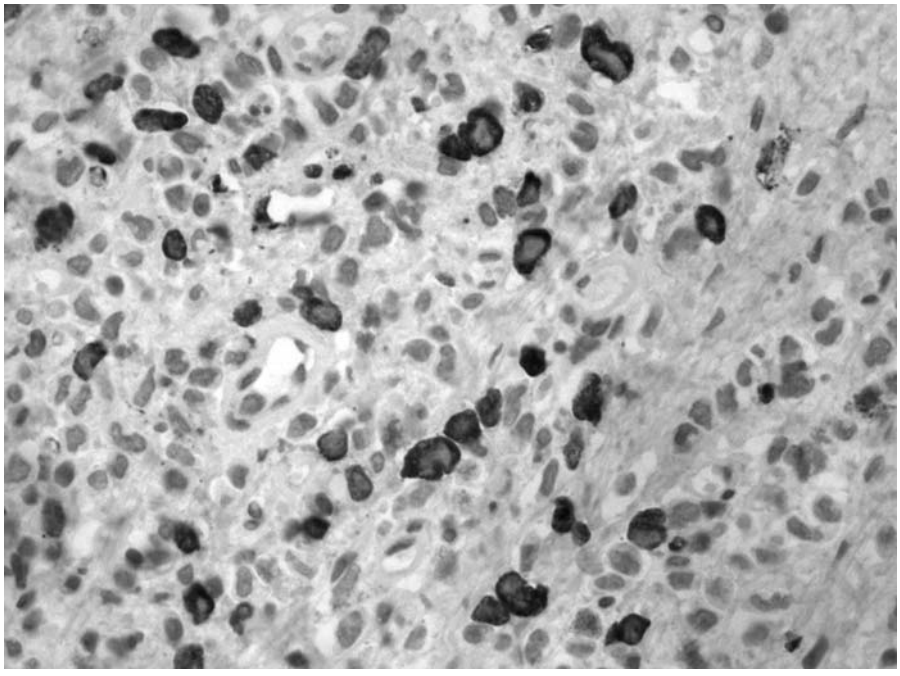

Fig. 2 - Immunochemistry pathological sections (MPO, 40x): positivity in about 40-50% of the neoplastic cells.

while lymphoid markers (CD20 and CD3) and TdT were negative, as well as the more undifferentiated (blastic) CD34 marker.

The final diagnosis was myeloid sarcoma with a unique location in the anal canal.

Afterwards, the patients was re-evaluated for his hematologic disease and chemotherapy was started. In the surgical follow-up, pain improved with healing of the anal fissure.

Discussion and conclusion

According to the WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues, a myeloid sarcoma is a tumor mass consisting of myeloid blasts with or without maturation occurring at anatomical sites other than the bone marrow. Infiltrates of any site of the body by myeloid blasts in leukemic patients are not classified as myeloid sarcoma, unless they present with tumor masses in which the tissue architecture is effaced (3).

Myeloid sarcoma may be observed in advanced stages of acute myeloblastic leukemia. Sometimes, it is a manifestation of relapse of myeloproliferative neoplasm previously treated or even after bone marrow and stem cell transplantation for acute or chronic leukemia (4). Only occasionally myeloid sarcoma may precede the onset of acute myeloid leukemia, which in these cases can develop months or years later.

It can occur as a solitary mass or multiple nodules and it is usually observed in skin, bone, and soft tissue, such as oral cavity, mucosal or genital tract, and sometimes in the central nervous system (2, 4-6). Very rarely it can involve the gastrointestinal tract (1, 2), and in these cases it may involve the mucosa from the oral cavity to the colon-rectum. Location in gallbladder, bile duct, pancreas and liver has also been described (1).

Only one case of involvement of the anus associated with lesions observed in the rectum and the sigmoid colon, has been described in the Literature (1). The isolated location of myeloid sarcoma in the anal canal, as observed in our case, have never been reported.

Diagnosis of myeloid sarcoma may be challenging, depending on the location of the disease. Previous clinical history of the patient might be of help. Biopsy is crucial for the diagnosis. Hystopathologically, myeloid sarcoma has to be differentiated by non-Hodgkin lymphoma or undifferentiated carcinoma. Staining with immunohystochemical markers allows the definitive diagnosis (1), and in these cases, differential diagnosis with other diseases can be challenging (7, 8).

In the patient of this report, the anal canal myeloid sarcoma presented with anal pain and clinical findings showing an anal fissure. The manifestation as an anal fissure could have induced in error. Signs and symptoms in isolated location of anal canal of myeloid sarcoma are not specific and the differential diagnosis with idiopathic anal fissure is extremely difficult. We achieved the final diagnosis through the hystopathological examination of a skin lesion close to the anal fissure resembling a skin tag.

In conclusion, although it is very rare to observe an involvement of the anal canal, in patients with acute myeloid leukemia, it is imperative to consider in the diagnosis of anal canal disease the occurrence of myeloid sarcoma. With a clinical history of acute myeloid leukemia, we should have thought to the final diagnosis.

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