

Case Report

Extra-Medullary Haematopoiesis in the Nose – A Rare Case

Ricardo Caiado*, Ana Melo, Joao Eloi, Jose Bastos, and Pedro

Tome

Department of Ear, Nose and Throat, Centro Hospitalar Universitário de Coimbra, Portugal

*Corresponding author

Ricardo Caiado, Department of Ear, Nose and Throat, Centro Hospitalar Universitário de Coimbra, Serviço ORL 10º piso, Praceta Mota Pinto, 3000-075 Coimbra, Portugal, Tel: 351-239-400-450; E-mail: ricardocaiado86@gmail.com

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Abstract

Introduction: In several hematological pathologies (eg myelofibrosis, chronic hemolytic anemia, etc.), when hematopoiesis function of the bone marrow is insufficient, the phenomenon of extramedullary hematopoiesis may occur. Although hematopoietic tissue has a tendency to form masses that can be found in almost any part of the body, it rarely appears in nose and paranasal sinuses.

Case presentation: The authors present a male patient, 75 years old, observed in Otorhinolaryngology consultation with epistaxis of the right nasal fossa and progressive nasal obstruction with a 6 months evolution. Besides intermittent cacosmiano other complaints were related by the patient.

Relevant previous medical history: myelofibrosis JAK 2 positive and chronic autoimmune hemolytic anemia.

The patient examination presented a friable injury in the right nasal cavity. A paranasal sinuses CT scan was requested to characterize the injury and a biopsy was scheduled. CT scan revealed an expansive lesion fully filling the right maxillary sinus with extension to the right nasal fossa. Two biopsies were performed but their results were inconclusive. A biopsy/excision of the lesion in the operating room lead to a extramedullary nasal hematopoiesis diagnostic.

Conclusion: Hematological diseases often have a clinical presentation with signs and symptoms of the otorhinolaryngological area. Whenever pathology with atypical evolution arises these should be considered as a possible diagnostic. Since extramedullary nasal and paranasal sinus hematopoiesis is a rare entity, each case should be individually addressed, according to the signs and symptoms, and the patient's comorbidities.

Keywords

- Extra-medullary haematopoiesis
- Nose
- Paranasal sinuses

ABBREVIATIONS

CT: Computer Tomography; MRI: Magnetic Resonance Imaging

INTRODUCTION

The authors present a case of a patient with complaints of progressive nasal obstruction and recurrent epistaxis with diagnosis of extramedullary hematopoiesis of the right nasal cavity. This type of phenomenon is extremely rare and few cases have been reported in the literature [1].

In several hematological pathologies (eg: lymphomas, leukemias, thalassemia, myelofibrosis, chronic hemolytic anemia, etc.), when hematopoiesis function of the bone marrow is insufficient, the phenomenon of extramedullary hematopoiesis may occur. Hematopoietic tissue has a tendency to form masses that can be found in almost any part of the body [2]. The most commonly affected areas are the paravertebral thoracic zones,

the liver, spleen and intra-abdominal lymph nodes. More rarely liver and spleen can be affected by masses that require differential diagnosis with neoplasias. Other less common areas are: intra-spinal canal, pre-sacral area, nasopharynx, paranasal sinuses, paratracheal area, pleural and precardiac space, kidneys and para-pelvic space, genito-urinary tract, gastrointestinal tract, central nervous system, prostate, perinium, skin, middle ear, lacrimal glands [1-3].

Systemic hematological diseases may have local manifestations in the otorhinolaryngological area that are not always easy to detect and diagnose. The diagnosis may be suggested by imaging tests such as CT scan and MRI, which characterizes better this type of lesions. The final diagnosis is obtained through histological studies. There is no treatment recommended for this pathology in the literature. Conservative medical treatment or more invasive surgical treatment are both valid options [1,4,5].

CASE PRESENTATION

The authors present a male patient, 75 years old, observed in Otorhinolaryngology consultation with epistaxis of the right nasal fossa and progressive nasal obstruction with a 6 months evolution. Besides intermittent cacosmia no other complaints were related by the patient.

Relevant previous medical history: myelofibrosis JAK 2 positive, chronic autoimmune hemolytic anemia (both diagnosed 5 years ago), myocardial revascularization surgery for coronary artery disease (2 years ago). The patient denied smoking and alcoholic habits. He also denied exposure to dust or animals in a professional or personal environment. The patient was medicated with folic acid, prednisolone, bisoprolol, aspirin and ruxolitinib.

The patient examination presented a friable lesion in the right nasal cavity. It was not possible to perform an endoscopic examination. In the left nasal cavity was observed a convex septum deviation with no other changes. A paranasal sinuses CT scan was requested to characterize the lesion and a biopsy was scheduled. CT scan showed an expansive formation that completely filled and distended the right maxillary sinus, with a discontinuity of the postero-lateral and medial wall and also of the papyraceous lamina inferior wall, with extension to the nasal fossa. The CT revealed a lesion with heterogeneous density, without significant enhancement after administration of contrast. It also exhibited ethmoidal involvement, with molding and destruction of some osseous trabeculae. Despite the mentioned discontinuity, the remaining sinus walls were thickened. The contralateral maxillary was partially filled, probably due to obstruction of ostiomeatal complex by the deviated septum (Figures 1,2).

A biopsy of the lesion was performed and its results were inconclusive. Nevertheless, it has raised a lymphoproliferative disease suspicion. The biopsy was repeated, and again had an inconclusive result. It was decided to perform a biopsy/excision of the lesion in the operating room. An endoscopic approach was performed. The lesion was completely removed with simple traction, similar to a removal of a single polyp with a remanent cavity. The patient subsequently performed a MRI (Figures 3, 4) that describes an absence of medial wall of right maxillary sinus and thickening of the remaining walls, and partial turbinectomy without any evidence of nasal foci of hematopoiesis.

The anatomopathological study of the lesion revealed a focus of extramedullary nasal hematopoiesis. The patient also underwent abdominal and lumbar CT scanning to detect other sites of extramedullary hematopoiesis but the results were negative.

DISCUSSION

Extramedullary hematopoiesis with a focus on the paranasal sinuses is extremely rare with few cases described in the literature and there are no guidelines concerning it.

In this clinical case with the patient's hematological antecedents the hypothesis of haematological disorders gained more strength. The presence of a space occupant lesion always



Figure 1 CT scan coronal view.

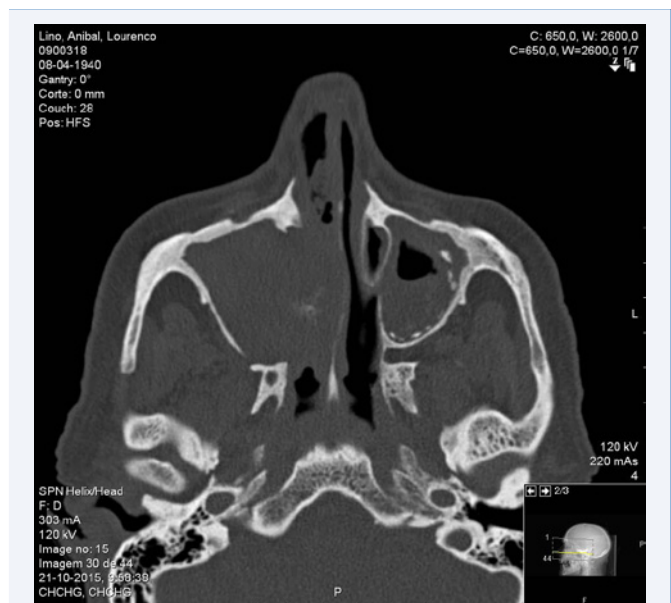


Figure 2 CT scan axial view.

requires its study. As any lesion occupying space requires an imaging staging with CT scan and eventually MRI must be performed. Histological diagnosis using biopsy is fundamental to exclude other neoplasms and to define an effective treatment.

As stated previously there are no guidelines for this pathology and each case must be individually oriented. Therapy can be adjusted for the underlying pathology to try to increase bone marrow function by decreasing the stimulation of extramedullary hematopoiesis. Another solution involves removal of the lesion. The decision between a more or less invasive treatment should be based on the patient's complaints, the accessibility of the

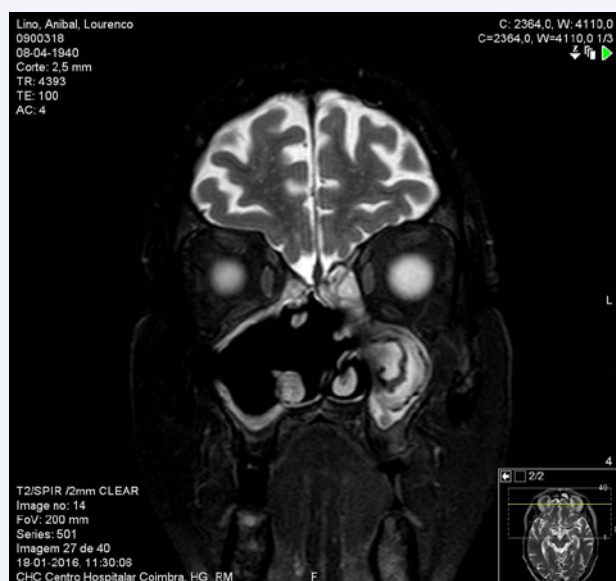


Figure 3 MRI coronal view.

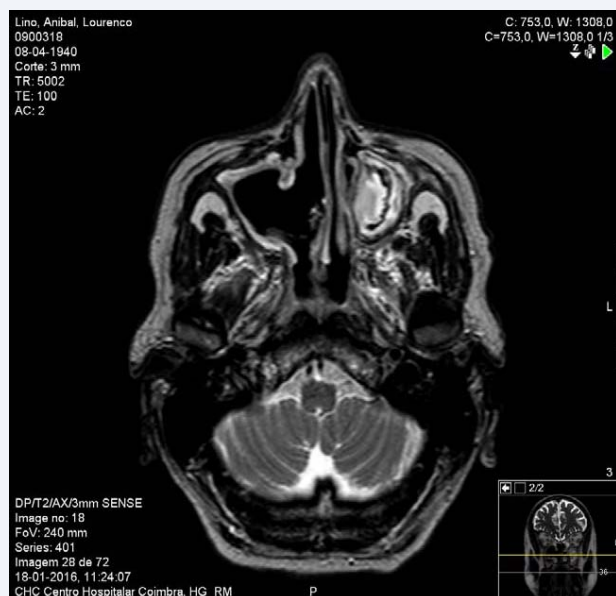


Figure 4 MRI axial view.

lesion and the general condition and comorbidities of the patient that may constitute a contraindication for a more invasive procedure. In the future, with the cumulative evidence and the aware of this unusual location of hematopoiesis in patients with bone marrow disorders, with better knowledge of lesions features in imagiologic examination, probably in some cases invasive procedures will be avoided. In this case, in spite of the cardiac comorbidities of the patient, the lesion had easy surgical access by endoscopy, and the clinical symptoms were significant. The underlying pathology, regardless of surgical excision, must be adjusted to prevent relapse and new outbreaks of disease.

CONCLUSION

The hematological pathologies are frequently manifested with signs and symptoms of the otorhinolaryngological area and should therefore always be present in the list of differential diagnoses. This principle becomes more relevant if the evolution is atypical and the histological diagnosis proves difficult to perform.

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