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CASE REPORT

Extramedullary plasmacytoma of the nasal cavity with kappa light chains in a young person: A case report

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ABSTRACT

BACKGROUND. Extramedullary plasmacytoma is a rare tumor, representing less than 1% of head and neck tumors. This specific type of tumor occurs in 90% of cases in the upper aero-digestive tract, with more frequent location in 80% of cases in the submucosa.
CASE REPORT. We report the case of a 39-year-old young man who presented for bilateral nasal obstruction with swelling of the left nasal region and intermittent epistaxis of moderate abundance. The clinical examination revealed a tumor mass in the nasal cavity bleeding on contact, with the absence of nasal airflow. The cranio-facial CT and MRI imaging with contrast agent showed a large, locally advanced tumor in the left nasal cavity, with destruction of the surrounding osteo-cartilaginous structures. The anatomopathological study of the biopsy specimen returned in favour of a plasmacytoma of the nasal cavity with kappa light chains. The biological assessment of the multiple myeloma confirmed that it was an extramedullary plasmacytoma. Radiotherapy treatment was proposed.
CONCLUSION. In the light of this case of extramedullary plasmacytoma, we will discuss the clinical, therapeutic and evolutionary characteristics of this tumour.
KYEWORDS: plasmacytoma, myeloma, extramedullary, kappa, nasal cavities.

INTRODUCTION

Plasma cell tumors form a group of neoplasms characterized by the monoclonal proliferation of B lymphocytes producing an immunoglobulin in a homogeneous manner. This immunoglobulin is of normal structure, either complete (2 heavy chains and 2 light chains) or incomplete, with a single kappa or lambda light chain. It is the indirect reflection of the tumor mass.

Plasma cell proliferations include multiple myeloma, solitary bone plasmacytoma, extramedullary plasmacytoma and plasma cell granuloma[1].

Extramedullary plasmacytoma is a rare tumor, representing less than 1% of head and neck tumors[2]. It occurs mainly in the elderly male subjects after 60 years. Its reference treatment, although it is radiotherapy, still remains a subject of controversy as to the dose and the target volumes[3].

CASE REPORT

A 39-year-old young man, without a notable pathological history, was admitted in our Department of Otorhinolaryngology for a tumor mass in the left nasal cavity evolving for 6 months. The evolution was marked by the progressive installation of a unilateral left nasal obstruction, which later became bilateral, with episodes of epistaxis of moderate abundance. The patient presented no other associated signs, in particular no decrease in visual acuity or tearing, no signs of intracranial hypertension.

The clinical examination showed (Figure 1): swelling of the left nasal region with filling of the left nasolabial sulcus; nasal flow abolished bilaterally; the presence of a budding mass from the left nasal fossa, bleeding on contact, reaching the nasal vestibule, pushing the nasal septum towards the contralateral side; unremarkable ophthalmic and neurological examination.

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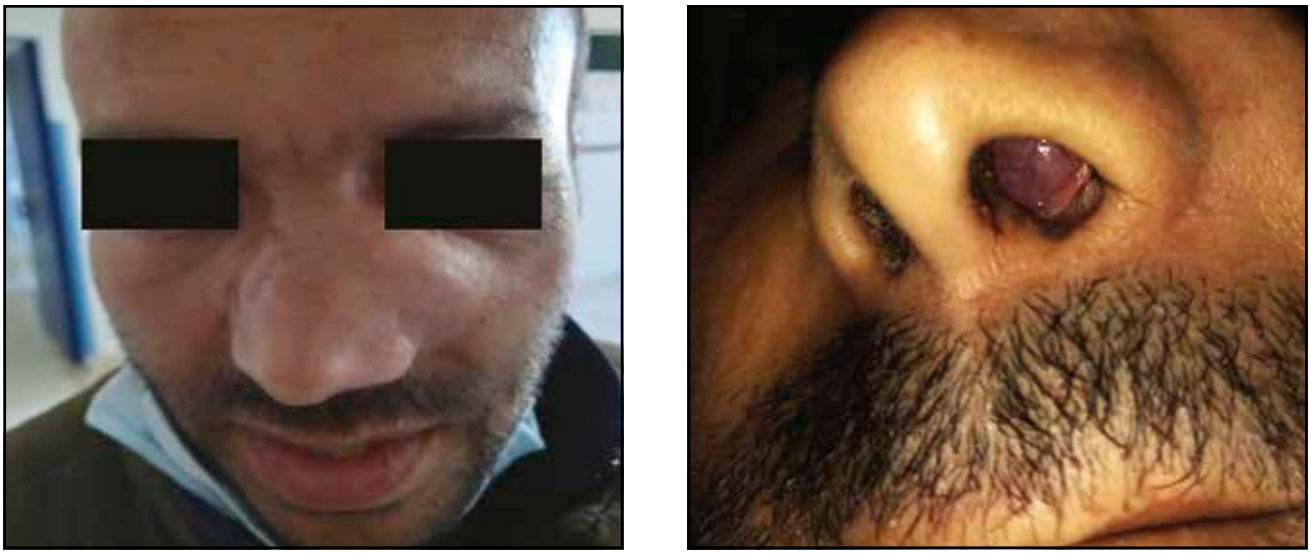

Figure 1. Swelling of the left nasal region with filling of the left nasolabial sulcus and a purplish mass in the left nasal fossa, reaching the nasal vestibule.

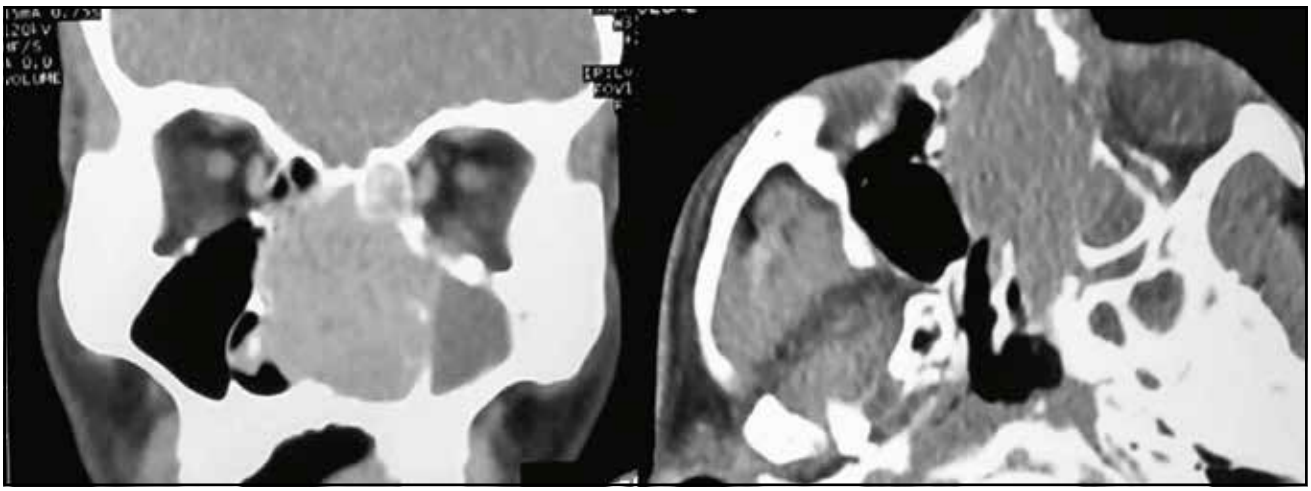

Figure 2. Contrast-enhanced cranio-facial CT scan, axial and coronal sections, showing a speculated reaction of the cortical bone of the left nasal bone.

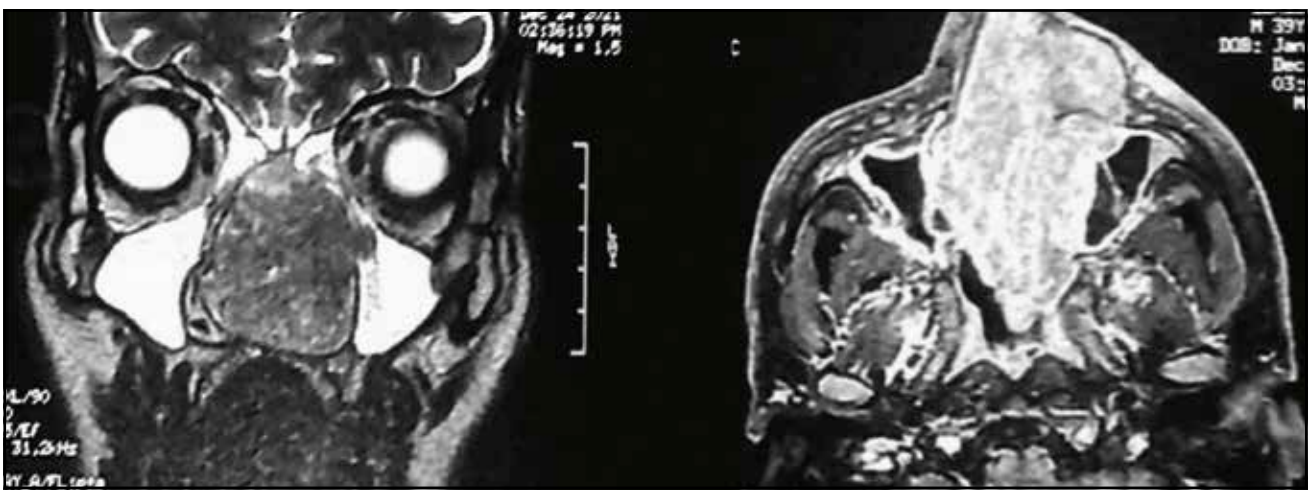

Figure 3. Cranio-facial MRI, coronal section T2 sequence and axial section T1 FAT SAT sequence, objectified an iso-signal tumor lesion of the left nasal fossa, locally advanced, but without endocranial or endo-orbital extension.

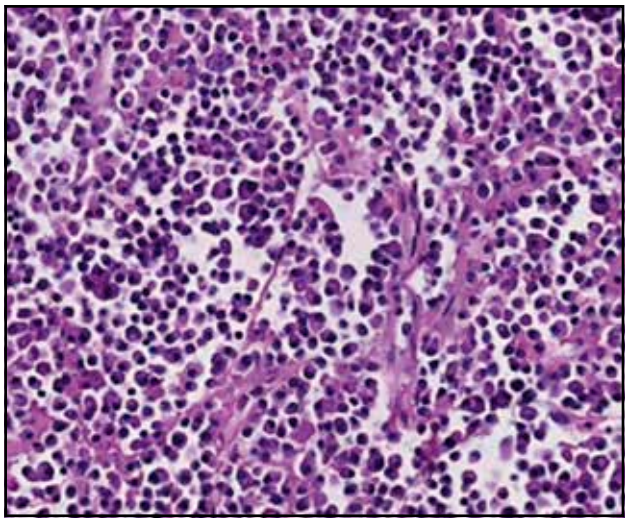

Figure 4. Immunohistochemistry study shows intense membrane marking in network by CD138; it expresses Kappa light chains (high-resolution electron microscopy with haematoxylin-eosin staining).

The contrast-enhanced cranio-facial CT scan showed a tissue mass filling the left nasal cavity, enhanced after contrast measuring 86*47*50 mm. The tumor presented an intimate contact with the nasal septum, which was deviated towards the contralateral side; it came in contact with the left nasal bone and maxillary bone bilaterally, responsible for a spiculated reaction of the cortical bone of the left nasal bone (Figure 2).

The cranio-facial MRI with contrast agent objectified a tumor mass centered at the level of the left nasal fossa, isosignaled in T1 and T2, of tissue density on the scanner, measuring 86*47*50 mm in diameter. The tumor presented the following ratios and extensions (Figure 3):

- Anteriorly – extension to the nostril orifices bilaterally, and towards the subcutaneous space opposite;
- Laterally – invasion of the nasal septum, the right nasal fossa; with lysis of the internal walls of the maxillary sinus bilaterally with endosinusal extension; it enlarges the left nasal tear duct and infiltrates the extra conical fat of the left orbit and it comes into contact with the internal rectus muscle;
- Inferiorly – lysis of the hard palate in places, without endobuccal extension;
- Superiorly – lysis of the ethmoidal floor with endosinusal extension, without endocranial extension;
- Posteriorly, it protrudes into the nasopharyngeal lumen.

The patient underwent a biopsy under local anaesthesia. The anatomopathological study returned in favour of a plasma cell population. An immunohistochemistry study was carried out using high-resolution electron microscopy with haematoxylin-eosin staining, tumor proliferation showing partial labelling with CD45 without obvious labelling with pan-cytokeratin or with AML. This population presents an intense membrane network labelling by CD138; it expresses the Kappa light chains with a proportion of 100% of the cells, as opposed to the Lambda light chains, which represent less than 1% of the cells infiltrating the chorion. So, the result came back in favour of an infiltration of the nasal mucosa by a monotypic plasma cell population (a Kappa monotype), evoking a plasmacytoma (Figure 4).

The multiple myeloma assessment was negative, without anemia, with proper renal function, calcemia, and normal levels of beta2 micro globulin. The electrophoresis of serum proteins objectified detection of a low intensity band in Gamma-globulins, but there was a peak of a serum Kappa light chain (at 226 mg/l) and positive Bence-Jones proteinuria. The bone marrow biopsy objectified a normal and balanced representation of the granular and erythroblast lines with the presence of 4% plasma cells.

In total, it was a solitary extramedullary plasmacytoma with the following 4 criteria:

1- Isolated soft tissue lesion with evidence of clonal plasma cells proven by biopsy.
2- Normal bone marrow with absence of clonal plasmacytosis.
3- Absence of skeletal anomaly of the spine and pelvis.
4- Absence of CRAB criteria (C: normal calcemia, R: correct renal function, A: anemia "no anemia", B: normal bone marrow biopsy).

The final diagnosis was extramedullary plasmacytoma of the left nasal cavity with major local extensions (stage II) and kappa light chains. It should be noted that the presence of a monoclonal light chain (Kappa or lambda) in the serum and/or in the urine is not a reason for excluding the diagnosis of extramedullary plasmacytoma, because the 4 criteria are confirmed.

Exclusive radiotherapy was instituted (47Gy in 3 fields, 2 lateral and one anterior with eye protection), 34 sessions over 6 weeks. The evolution was favourable with complete remission.

Quarterly alternating ENT/Radiotherapist monitoring was initiated with complete Blood Count and Protein Electrophoresis. Post-radiation endoscopic and radiological control (cervico-facial MRI) did not detect any tumor recurrence after an 18-month follow-up.

DISCUSSIONS

Extramedullary plasmacytoma is a rare tumor, representing less than 1% of head and neck tumors[4]. In 80% of cases, it is a submucosal tumor of the upper aero-digestive tract, mainly in the naso-sinus cavities (75%) (like our case), the larynx (15%) and the oropharynx (10%)[5-7].

The average age is 65 years old and 90% of patients are over 40 years old[8,9]. In our case, the patient is 39 years old and, as seen reviewing the literature, this

type of tumor is very rare at this age. Extramedullary plasmacytoma is more frequent in men than in women, with a sex ratio of 3/1 to 4/1[9].

The etiology of extramedullary plasmacytomas remains unknown. Genetic factors, exposure to radiation, chronic antigenic stimulation are put forward in the literature as risk factors for developing plasma cell tumours, but no association has ever been proven[10]. Our patient had no history of exposure to toxic substances.

The clinical signs are the result of a mass effect produced locally by the tumor and, depending on its location, they can be summarized: swelling (80%), obstruction of the airways (35%), epistaxis (30%), pain (20%), proptosis (15%), rhinorrhea (10%), regional lymphadenopathy and cranial nerve palsies[11]. In our case, the patient presented with swelling of the left nasal region with nasal obstruction, associated with moderate episodes of epistaxis.

The CT appearance is that of a discreetly heterogeneous mass, with moderate to intense enhancement, frequently associated with bone lysis (like our case, but without bone lysis). The appearance is not specific and resembles that of nasopharyngeal lymphomas and carcinomas. MRI shows a tumor in T1 iso signal with massive contrast enhancement and central inhomogeneity[12].

However, whether solitary or multiple, plasmacytomas must be considered in the differential diagnosis before any invasive lesion of the naso-sinus cavities. The information provided by CT investigations on bone involvement and by MRI on the tumor signal and contrast enhancement can sometimes guide the diagnosis. However, diagnostic certainty can only be histological[13,14] and the ENT specialist is most often confronted with the diagnosis of solitary plasmacytoma in front of the unexpected result of a tumor biopsy[7].

The histological diagnosis of extramedullary plasmacytoma is particularly difficult, because plasma cells are cells that are found in abundance in various pathologies such as chronic inflammation, sarcoidosis, syphilis, neuroblastomas or certain sarcomas[15,16].

The definitive diagnosis of an extramedullary plasmacytoma is based on the anatomopathological analysis of biopsies performed under local or general anaesthesia. The anatomopathological study and immunohistochemistry are studied by high-resolution electron microscopy with haematoxylin-eosin staining, to determine the degree of plasma cell differentiation. Immunohistochemical techniques (immune labelling by antibodies, immune peroxidase) have been applied in order to identify the kappa or lambda light chains and the heavy chains IgG, IgA and IgM (rare IgD and IgE) of intracytoplasmic immunoglobulins[17]. In our case, the cell population presents an intense membrane marking in network by the CD138; it expresses the Kappa light chains.

In fact, neither the symptoms nor the macroscopic appearance of the tumor had immediately pointed towards a plasmacytoma. The diagnosis of solitary extramedullary plasmacytoma is affirmed by the normality of biological examinations, bone marrow biopsy and radiological examinations, thus eliminating any metastatic dissemination[7]. A classification of extramedullary plasmacytoma has been established, depending on tumor extension[8]: Stage I – tumor limited to a single site, Stage II – locoregional tissue and/or lymph node invasion and Stage III – extension so-called metastatic (myelomatosis).

Once the diagnosis of plasmacytoma has been established, its "extramedullary" and isolated character still needs to be proven. All the authors are unanimous as to the assessment to be made to eliminate multiple myeloma[9]. This assessment includes complete blood count, a blood ionogram, calcemia, phosphoremia, electrophoresis of blood proteins, uremia, creatininemia, search for proteinuria, bone-medullary biopsy, X-rays of the entire skeleton. Some authors complete this assessment with a myelogram, a whole-body MRI[10]. In short, to confirm the diagnosis of an extramedullary plasmacytoma, the following 4 criteria must be present:

1. An isolated lesion of soft tissue with evidence of clonal plasma cells proven by biopsy.
2. Normal bone marrow with absence of clonal plasmacytosis.
3. Absence of skeletal abnormality of the spine and pelvis.
4. Absence of CRAB criteria (C: normal calcemia, R: correct renal function, A: Anemia "no anemia", B: Normal bone marrow biopsy).

The presence of a monoclonal paraproteinemia in the serum and/or in the urine, as in our case, is not a reason for excluding the diagnosis of extramedullary plasmacytoma, because the 4 criteria are confirmed[11].

Dissemination generally occurs within two years of the initial diagnosis and rarely beyond[12]. This tends to integrate extramedullary plasmacytoma in a continuum with multiple myeloma, especially as other authors[13] made the same findings in patients who developed multiple myeloma 22 and 36 years after the diagnosis of extramedullary plasmacytoma. Non-supporters of this theory argue that a disseminated extramedullary plasmacytoma does not give the same lesions as multiple myeloma[14] and has a much more favourable prognosis with 31% survival at 5 years compared to 18% for "de novo" multiple myeloma[15]. It is therefore difficult to conclude imperatively on the interrelationships extramedullary plasmacytoma / multiple myeloma, but it seems, by comparing the different experiences of the literature, that there is a continuity between the two, where the solitary plasmacytoma can be the inaugural manifestation of a multiple myeloma, with generalization from the outset or delayed by a few months or years of evolution. This transformation is seen in 5 to 32% of the cases[16,17].

The treatment of extramedullary plasmacytoma remains the subject of much debate in the literature and it is difficult to obtain significant statistics.

Therapeutic modalities include surgery, radiotherapy or a combination of the two. Still, others have recently attempted electrocoagulation[5] of an endonasal extramedullary plasmacytoma or the KTP/532 laser[18]. Chemotherapy (particularly alkylating agents) is reserved for major recurrences and disseminations[19].

Extramedullary plasmacytomas are tumors known to be very radiosensitive[20]. In addition, these are submucosal lesions of the upper aero-digestive tract that do not lend themselves to wide surgical resection due to the proximity of vital structures. Surgery can thus lead to functional disorders or aesthetic damage[21]. In our case, surgery was ruled out to avoid functional and aesthetic consequences (secondary palatal prosthesis with feeding difficulties and aesthetic damage). Our patient received exclusive radiotherapy.

The majority of radiotherapists agree to treat small extramedullary plasmacytomas with 30 to 40Gy and go up to 50-60Gy for extramedullary plasmacytoma with bone or muscle invasion[22]. Finally, salvage surgery is always possible, just as radiotherapy can complete an insufficient resection[23]. It seems judicious to us to temper this debate according to the location of extramedullary plasmacytomas[24]. Faced with locally advanced endonasal extramedullary plasmacytoma (like our case), we start with radiotherapy at 47Gy and go up to 60Gy, then we discuss surgery[25].

Long-term follow-up is therefore essential, including an ENT examination to detect local recurrences and a haematological assessment to detect dissemination[26,27].

The prognosis of solitary plasmacytoma essentially depends on the risk of transformation into multiple myeloma, the size of the tumors and lymph node invasion. Its 10-year survival is 50-80%[28].

CONCLUSIONS

Solitary or multiple plasmacytoma must be considered in the differential diagnosis of any invasive lesion of the sinonasal cavities[28]. The clinical presentation is non-specific. The information provided by imaging helps to guide the diagnosis. Histological certainty must be obtained.

Radiotherapy is the reference treatment regardless of the initial surgical procedure[29].

The prognosis of plasmacytoma is dominated by the risk of developing multiple myeloma.

Regular clinical and biological follow-up is necessary.

Conflicts of interest: The authors declare there is no conflict of interest.

Contribution of authors: All the authors have equally contributed to this work.

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