Multiple Myeloma Revealed by a Plasmacytoma of the Sphenoid Sinus in a Young Subject: Case Report

Wafaa Bennane¹,²,*, Hanaa Bencharef¹,², and Bouchra Oukkache¹,²

ABSTRACT

Extramedullary plasmacytomas are a rare entity. They represent less than 1% of malignant head and neck tumors. We report the case of a 42-year-old patient complaining of headache, ptosis of the right eye and hypoesthesia of the face evolving for a few weeks. MRI showed a mass in the sphenoid sinus with local extension. The myelogram objectified the presence of 60% dystrophic plasma cells, which confirms the diagnosis of multiple myeloma. The patient received chemotherapy with a good course. Here, we report a rare case of sphenoidal sinus plasmacytoma that evolved into multiple myeloma.

Keywords: Multiple myeloma, plasmocytoma, sphenoid sinus, tumour.

1. Introduction

Plasmacytoma is a rare tumor characterized by the neoplastic proliferation of a single clone of plasma cells, which may be associated with myeloma. Extramedullary plasmacytomas represent less than 1% of malignant head and neck tumors. The location in the sphenoidal sinus is rare and constitutes only 1.6% of extramedullary plasmacytoma. Only a few cases were reported in the literature. We report an exceptional case of sphenoidal plasmacytoma revealing multiple myeloma in a young subject.

2. Case Report

A 42-year-old man, with no particular pathological history, presented to the emergent department with intracranial hypertension and a ptosis of the right eye, epistaxis and hypoesthesia of the right hemiface. The neurological examination found hypoesthesia in the territory of V2 and V3 nerves with ptosis of the right eye. The rest of the examination was normal. A cerebral Magnetic resonance imaging (MRI) confirmed the presence of a voluminous expansive process in the sphenoid sinus that was isointense on T1-weighted images and hypointense on T2-weighted images (Fig. 1), with homogeneous and intense gadolinium enhancement. This mass lesion invaded the cavernous sinus and encompassed the internal carotid artery on the right, the sella turcica above and the cavum below. In addition to that, multiple cranial, orbital and facial osteolytic lesions with the same characteristics have been found. These lesions suggested, among other things, a plasmacytoma. A chest computed tomography (CT) performed showed multiple costal osteolytic lesions, some of which are complicated by fractures.

Biological tests showed severe anemia at 4.1 g/dl. It was normochromic, normocytic and non-regenerative. The patient also had renal failure with creatinine at 88 mg/l, hypercalcemia at 120 mg/l and a high C-reactive protein (CRP) value at 185 mg/l. The myelogram revealed the presence of 60% of dystrophic plasma cells (Fig. 2). Serum protein electrophoresis found a monoclonal peak with, on immunoabsorption, the presence of an IgG Lambda immunoglobulin. All these results confirmed the diagnosis of multiple myeloma. The biopsy of the mass was not performed because of the local invasion and the depth of the anemia.

Therapeutically, the patient received chemotherapy with a good evolution.

3. Discussion

Plasmacytomas are rare malignant tumours, of which there are two clinical forms: bone or intramedullary plasmacytoma and extramedullary plasmacytoma.
The bony forms are the most frequent and mainly affect the vertebrae. Extramedullary plasmacytomas represent only 10% of solitary plasmacytomas and 1% of all tumors [1]. They preferentially reach the submucosa of the upper airways and represent 0.4% of malignant tumors of the head and neck [2].

Location in the sphenoid sinus is extremely rare and constitutes only 0.1% of sphenoid tumors with only twenty cases reported in the literature [1]. We recall that the sphenoid sinus is surrounded by critical structures such as the optic, abductor, ophthalmic and maxillary nerves as well as the division of the trigeminal nerve and the carotid arteries. These reports largely explain the difficulty of surgically approaching and histologically diagnosing a pathological process when it is located in the sphenoid sinus with extension to surrounding structures, as is the case for our patient [3].

The average age at the time of diagnosis of the plasmacytoma is 40 to 50 years old, such is the case for our patient who was young [3] whereas it is most often between 50 and 80 years old, with a peak at age 60 years in case of multiple myeloma [4].

Symptoms are usually mild or nonspecific until the tumor spreads to the walls of the sinus and surrounding structures, consisting primarily of visual loss, diplopia, and facial pain indicative of tumor extension to the cranial nerves II, VI and V. Other signs may reveal the disease such as epistaxis, rhinorrhea or even nasal obstruction [1]. On MRI, the plasmacytoma appears isointense on T1-weighted images with enhancement after injection of gadolinium [5].

In all cases, biological and radiological follow-up is necessary since more than 85% of cases of plasmacytoma develop true multiple myeloma [6]. The main tests that need to be done are the blood count, the erythrocyte sedimentation rate, the electrophoresis of serum and urinary proteins, and the myelogram, which confirms the diagnosis by showing the presence of more than 10% of dystrophic plasma cells [7]. In our patient, the myelogram showed that 60% of dystrophic plasma cells were present, which constitutes both an important diagnostic and prognostic element.

Current treatment for multiple myeloma varies considerably. Multimodal treatment, encompassing chemotherapy,
radiation therapy, targeted therapy and stem cell transplantation, has been recognized as an important approach to improving overall survival [3]. Complete excision of the tumor is not recommended because it has never been validated in terms of tumor control and survival. For solitary forms, targeted radiotherapy on the body of the sphenoid bone is the treatment of choice because it allows satisfactory local tumor control [8].

The prognosis of multiple myeloma depends on several factors: those related to the patient (age, WHO performance index status), factors related to the disease, in particular renal function, tumor mass as well as certain biological parameters such as CRP, Albumin, percentage of dystrophic plasma cells etc., [3].

The prognosis is generally good for solitary plasmacytomas and bad for plasmacytomas accompanying myeloma multiple with survival not exceedingly not 18 months [5].

4. Conclusion

Plasmacytoma of the base of the skull revealing multiple myeloma is a rare entity, but one that must be considered among the diagnoses to be considered in the presence of any invasive and lytic lesion of the sphenoid sinus. However, it is necessary to complete a myelogram in order to rule out systemic multiple myeloma.

Conflict of Interest

Authors declare that they do not have any conflict of interest.