

Solitary nasopharyngeal plasmacytoma in an elderly woman

Abstract

Extramedullary solitary plasmacytomas are very rare entities representing only 3% of all plasma cell malignancies, and with a clear male predominance. All tissues can be affected, but the most frequent locations are the head and neck, lungs, and gastrointestinal tract. While not the rarest entity, solitary plasmacytomas still represents a small minority of nasopharyngeal malignancies: 4–6.2%.

We report an original case of solitary extramedullary plasmacytoma of the nasopharynx in a 76-year-old Tunisian woman with no significant past medical history investigated for nasal obstruction, vertigo, and upper (oropharyngeal) dysphagia that had been progressing for approximately two months.

As rare as it is, this diagnosis should, however, be considered in the differential diagnosis for any nasopharyngeal tumor.

Keywords: nasopharynx, solitary plasmacytoma, ent, extramedullary plasmacytoma, tumor

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Introduction

Solitary plasmacytomas are very rare entities. Their cumulative incidence is estimated at 0.15/100,000 to 0.191/100,000 with a clear male predominance.^{1–3} They are classically intraosseous (medullary or bone plasmacytoma) or, more rarely, extraosseous (extramedullary plasmacytoma): 70% vs. 30%.³ All tissues can be affected, but the most frequent locations are the head and neck, lungs, and gastrointestinal tract.^{1–7}

While not the rarest entity,^{2,3} solitary plasmacytomas still represents a small minority of nasopharyngeal malignancies: 4–6.2%.^{4,5} We report an original case of solitary extramedullary plasmacytoma of the nasopharynx in a 76-year-old Tunisian woman.

Case presentation

A 76-year-old Tunisian woman with no significant past medical history was investigated for nasal obstruction, vertigo, and upper (oropharyngeal) dysphagia that had been progressing for approximately two months. Her general condition was good, and the physical examination showed no significant abnormalities. The blood pressure profile was correct, as was the specialist neurological examination. Basic laboratory tests were within normal limits: complete Blood count, C-reactive protein, erythrocyte sedimentation rate, plasma protein electrophoresis, glucose, transaminases, creatinine, plasma ionogram, calcium, uric acid, lipid profile, and thyroid function. Chest radiography was normal.

ENT examination revealed a polypoid swelling of the nasopharynx. Axial slices as well as sagittal reconstruction of the cervicofacial CT-scan showed a polypoid, solid mass measuring 29×26×22mm without locoregional lymphadenopathy. Biopsy showed a diffuse infiltrate of monomorphic plasma cells, some of which were dystrophic. Immunostaining was positive for anti-CD138 and negative for anti-CD3, anti-CD20, and anti-CK antibodies. Anti-Ki67 antibody staining showed a proliferation index of 3 to 5%.

Further investigations ruled out a possible underlying multiple myeloma: plasma and urine protein electrophoresis, quantitative immunoglobulin assay, plasma and urine immunoelectrophoresis, myelogram, and bone marrow karyotype.

Abdominal ultrasound, thoraco-abdomino-pelvic computed tomography, X-rays of the bones, and cerebrospinal magnetic resonance imaging did not reveal any other tumor locations.

Following these results, the diagnosis of solitary nasopharyngeal plasmacytoma was made, and the patient was referred for radiotherapy.

Discussion

First described by Schridde H in 1905⁸ plasmacytoma is a localized, monoclonal plasma cell tumor.^{1–3,6–8} Extramedullary forms are exceptional, representing only 3% of all plasma cell malignancies.^{5,9} These tumors can occur at any age,^{1–3,9} but classically in the elderly,³ with a clear male predominance.^{1–3} Their occurrence in women, as in our case, remains much rarer. Indeed, in the large Swedish series by Nahi H et al, the global incidence of solitary plasmacytoma was 0.191/100,000 for males and 0.090/100,000 for female patients.²

Cervicofacial involvement is common but often underdiagnosed and plasmacytomas represent approximately 4% of all nasal cavity tumors.⁵ Diagnostic of solitary nasopharyngeal plasmacytoma requires the histological confirmation of an isolated plasma cell lesion on the nasopharynx, the presence of less than 5% plasma cells infiltration on myelogram, and the absence of specific end-organ damage of MM.^{3,5} Therefore, systemic investigations to rule out a possible underlying multiple myeloma must be performed before concluding to solitary plasmacytoma.^{1,3,5}

The clinical symptoms associated with solitary plasmacytoma of the nasopharynx are nonspecific. They may include: Upper (oropharyngeal) dysphagia, difficulty in swallowing, nasal obstruction, vertigo, shortness of breath, pain symptoms, and taste disturbance.^{3,5,6,10} However, this location can remain completely asymptomatic,⁶ and can mask concomitant laryngeal involvement.⁷

Exact pathogenesis of nasopharyngeal plasmacytoma remains unknown. Clonal plasma cell proliferation in the nasopharynx initiated by inhaled irritants or viral infection have been suggested to explain this location.^{5,10}

This location often responds to radiotherapy;^{1–3,5} however, exceptionally refractory forms are observed, requiring endoscopic resection with photodynamic therapy of the tumor remnant.¹¹ Microsurgery can also be effective as a radical treatment for small plasmacytomas.³ Chemotherapy does not appear to yield good results for these neoplasms, and its indication remains unclear and highly controversial.^{3,12}

Solitary plasmacytomas usually have a very good prognosis, particularly extramedullary ones. However, prolonged monitoring is recommended given the potential risk of secondary transformation into MM.^{13,14} This risk of progression to MM is estimated at 8 to 31% of cases,^{12,14} and appears to be higher if the proliferation index is high (greater than 10–30%).¹⁵

Conclusion

Solitary plasmacytomas of the nasopharynx are far from rare, but they are often underdiagnosed and poorly understood. They can remain asymptomatic for a long time and are frequently discovered incidentally on imaging. Their prognosis is generally good. Their occurrence in women, as in our case, remains exceptional. This diagnosis should, however, be considered in the differential diagnosis for any nasopharyngeal tumor.

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None.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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