The Journal of Laryngology and Otology
January 1991, Vol. **105**, pp. 41–43

Extramedullary nasal plasmacytoma

M. L. NAVARRETE, P. QUESADA, M. PELLICER, C. RUIZ (Barcelona, Spain)

Abstract
The literature on this rare tumour has been reviewed and three cases of nasal plasmacytoma are described. Immunohistochemistry demonstrated cytoplasmic IgA and Kappa determinants in all cases. Two patients are disease-free at the present time, the third developed an IgG-k multiple myeloma, previously not described in the literature.

Introduction

Plasmacytomas are either medullary or extramedullary. The latter is more common in the upper respiratory tract (Batsakis, 1983). Extramedullary plasmacytoma (EMP) represents less than 1 per cent of all head and neck malignancies (Kyle, 1975) and less than 0.4 per cent of upper respiratory malignancies (Webb *et al.*, 1962). Characteristically, EMP of the upper airways presents as a submucosal mass or swelling with a polypoid configuration often without destruction of the bone and causing nasal or pharyngeal symptoms. Microscopically, EMP consists of sheets of plasma cells which may be monomorphous or pleomorphic. Since the plasma cells are derived as a clonal proliferation from B-lymphocytes they may be characterized by immunohistochemical demonstration of cytoplasmic immunoglobulin determinants in the neoplastic cells. Immunoperoxidase methods are convenient in routinely processed histological specimens. By immunohistochemical demonstration of a monoclonal staining pattern, consisting of one light chain type and one heavy chain class, most EMPs can be distinguished from reactive plasma cell infiltrates with polyclonal staining patterns.

Case reports

Case 1

A 45-year-old man presented with a five month history of right-sided nasal obstruction and three episodes of epistaxis. On clinical examination, there was a deviated nasal septum to the left side. A pedunculated mass was seen in the right nasal fossa. The rest of the ENT examination was normal. Physical examination and laboratory data were normal. By biopsy, a histological diagnosis of plasmacytoma was made. Immunohistochemical staining showed Ig A-kappa phenotype (Fig. 1). Serum and urine electrophoresis failed to demonstrate any myeloma component or Bence-Jones protein. All other screening tests to rule out multiple myeloma were negative. The diagnosis of solitary plasmacytoma was confirmed. Surgical excision was performed and no recurrence has occurred in the three-year follow-up period.

Case 2

A 57-year-old man presented with a one year history of right sided nasal obstruction, right sided episodes of epistaxis and hyposmia. On clinical examination, there was a mass in the right nasal fossa. A right carotid angiogram was performed because of the clinical suspicion of an angioma, which showed vascularization by the internal maxillary artery. A CT scan showed invasion of the ethmoid bone. Surgical excision of the tumour was performed by a paralateral rhinotomy approach with previous ligation of the right external carotid artery.

Histological diagnosis of a plasmacytoma was made. Immunohistochemistry demonstrated an IgA-kappa phenotype (Fig. 2). Iliac crest bone marrow biopsy, bone survey, serum protein electrophoresis, urine analysis and chest X-rays were all normal and multiple myeloma was also excluded. No recurrence has occurred in the three year follow-up period.

Case 3

An 80-year-old woman presented with a one-year history of epiphora and right sided nasal obstruction. On clinical examination, there was a mass of benign appearance in the right nasal fossa. A CT scan showed an expansive process in the right nasal fossa by a tumour on the inferior concha, which was removed. Histological diagnosis of a plasmacytoma was made. Immunohistochemistry demonstrated an Ig A-kappa phenotype. Iliac crest bone marrow biopsy, bone scan, serum protein electrophoresis, urine analysis and chest X-rays were all normal and multiple myeloma was excluded. Radiation therapy (Co^{60}) was

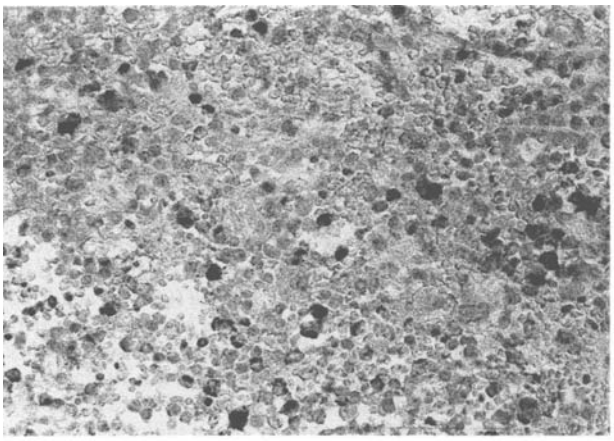

FIG. 1
Immunoperoxidase staining positive for intracytoplasmic alpha-heavy chains in a plasmacytoma.

From Department of Otorhinolaryngology, Autonomous University, Barcelona, Spain.
Accepted for publication: 10 September 1990.

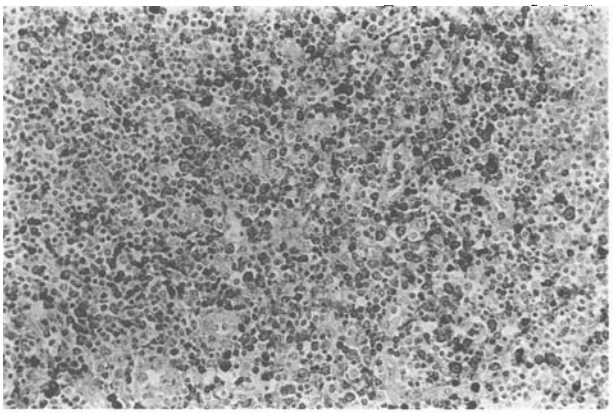

FIG. 2
Some cells positive for kappa light chains. (P.A.P. ×25)

given post-operatively to the nasal pyramid region (total dose: 40 Gy).

One year later, the patient complained of pain in the right fifth rib and left hip. A bone scan showed an increased uptake in multiple areas in the pelvis, spine, ribs and lytic lesions in the skull. Urine protein electrophoresis demonstrated Bence-Jones proteinuria with a narrow spike in the region of kappa light chains. Serum protein electrophoresis showed a gamma-M component of IgG-kappa. Chemotherapy was begun with melphalan and prednisone. Two years later, the patient suffered a stroke with a left hemiplegia. Deterioration was progressive and the patient died four months later from septicaemia.

Discussion

Eighty per cent of extra-medullary plasmacytoma (EMP) occur in the upper respiratory tract; 10 per cent are multiple myelomas (Kapadia *et al.*, 1982). There is a preponderance of men 3:1 (Waldron and Mitchell, 1988) and the average age of presentation is 60–70 years (Wiltshaw, 1976; Maniglia and Xue, 1983).

Most cases present in the nasal cavity, the most frequent single location (Dolin and Dewar, 1956; Webb *et al.*, 1962). It is usually a single tumour but multiple foci may occur (Stout and Kenney, 1949). EMP is a rare tumour of the head and neck region that can only be diagnosed by biopsy and histological examination (Chaudhuri *et al.*, 1988). Usually the diagnosis of plasmacytoma is an unsuspected result of biopsy (Sadek *et al.*, 1985) as in our cases. Clinical and histological diagnosis is difficult. Histologically it must be differentiated from multiple myeloma (MM) and polyclonal infiltrates of plasma cells. Plasmacytoma has a monoclonal origin as demonstrated by immunohistochemistry. The diagnosis of EMP can only be confirmed after the exclusion of systemic disease by serum and urine protein electrophoresis and immunoelectrophoresis, skeletal survey, bone scan, and marrow biopsy (Waldron and Mitchell, 1988). Some authors stipulate a disease-free interval of three years to qualify as a solitary plasmacytoma (Corwin and Lindberg, 1979). According to the spread of the disease EMP can be scored (Waldron and Mitchell, 1988). Localized disease may be treated by surgery or radiotherapy (Booth *et al.*, 1973; Corwin and Lindberg, 1979; Woodruff *et al.*, 1979). There are no significant differences between surgery alone, radiotherapy alone or combined surgery and radiotherapy (Helmus, 1964). The tumour should be treated as malignant with about a 50 per cent five-year survival rate (Chaudhuri *et al.*, 1988). The incidence of dissemination of EMP varies in different series from 21 to 75 per cent (Fishkin *et al.*, 1970; Mill and Griffith, 1980).

Multiple myeloma (MM) will develop in 17 to 33 per cent of patients and the necessity for lifelong follow-up is emphasized (Pahor, 1978). The true nature of primary EMP and its relationship to MM is controversial, some authors suggest they are separate entities while others that they form a continuous spectrum of plasma cell dyscrasias (Fu and Perzin, 1978; Corwin and Lindberg, 1979; Kapadia *et al.*, 1982). Disseminated disease is treated by chemotherapy (Fu and Perzin, 1978). One of our cases of Ig A-kappa plasmacytoma evolved in one year to an Ig G-kappa multiple myeloma. The presentation of an Ig G-kappa MM in a patient who previously had an Ig A-kappa plasmacytoma supports the hypothesis that EMP and MM are related diseases belonging to a continuous spectrum of plasma cell dyscrasias and suggests a failure in the gene codifying for the heavy chain protein.

Conclusion

Plasmacytoma must be ruled out whenever a chronic mass of benign appearance presents in the head and neck. Clinical and histological diagnosis of plasmacytoma is difficult. Immunohistochemistry (peroxidase antiperoxidase method) allows characterization of plasmacytoma phenotypes (light chain type and heavy chain class) and its differentiation from polyclonal plasma cell infiltrates. The diagnosis of EMP can only be confirmed after the exclusion of systemic disease (MM).

Acknowledgements

For their assistance in the preparation of this paper: E. Perello, F. Pumarola.

References

Batsakis, J. G. (1983) Plasma cell tumours of the head and neck. *Annals of Otology, Rhinology and Laryngology,* **92:** 311–313.

Booth, J. B., Cheesman, A. D., Vincenti, N. H. (1973) Extramedullary plasmacytoma of the upper respiratory tract. *Annals of Otology, Rhinology and Laryngology,* **82:** 709–715.

Chaudhuri, J. N. , Khatri, B. B., Chaterji, P. (1988) Plasmacytoma of the nose with intracranial extension. *Journal of Laryngology and Otology,* **102:** 538–539.

Corwin, J., Lindberg, R. D. (1979) Solitary plasmacytoma of bone vs. extramedullary plasmacytoma and their relationship to multiple myeloma. *Cancer,* **43:** 1007–1013.

Dolin, S., Dewar, J. P. (1956) Extramedullary plasmacytoma. *American Journal of Pathology,* **32:** 83–103.

Fishkin, B. G., Glassy, F. J., Hattersley, P. G. (1970) IgD Multiple myeloma: a report of five cases. *American Journal of Clinical Pathology,* **53:** 209–214.

Fu, Y. S., Perzin, K. H. (1978) Non-epithelial tumours of the nasal cavity, paranasal sinuses and nasopharynx: a clinicopathologic study (IX). Plasmacytomas. *Cancer,* **42:** 2399–2406.

Helmus, C. (1964) Extramedullary plasmacytoma of the head and neck. *Laryngoscope,* **74:** 553–559.

Kapadia, S. B., Desai, U., Cheng, V. S. (1982) Extramedullary plasmacytoma of the head and neck: A clinicopathologic study of 20 cases. *Medicine (Baltimore),* **61:** 317–329.

Kyle, R. A. (1975) Multiple myeloma. Review of 869 cases. *Mayo Clinic Proceedings,* **50:** 29–40.

Maniglia, A., Xue, J. W. (1983) Plasmacytoma of the larynx. *Laryngoscope,* **93:** 741–744.

Mill, W. B., Griffith, R. (1980) The role of radiation therapy in the management of plasma cell tumours. *Cancer,* **45:** 647–652.

Pahor, A. L. (1978) Plasmacytoma of the larynx. *Journal of Laryngology and Otology,* **92:** 223–232.

Stout, A. P., Kenney, F. R. (1949) Primary plasma cell tumours of the upper air passages and oral cavity. *Cancer,* **2:** 261–278.

Sadek, S. A. A., Dogra, T. S., Khan, M. K., Baraka, M. E., Shi-

dom, N. N. (1985) Plasmacytoma of the nasopharynx. *Journal of Laryngology and Otology,* **99:** 1289–1292.
Waldron, J., Mitchell, D. B. (1988) Unusual presentations of extramedullary plasmacytoma in the head and neck. *Journal of Laryngology and Otology,* **102:** 102–104.
Webb, H. E., Harrisson, E. G., Masson, J. K., Ramine, W. H. (1962) Solitary extramedullary myeloma of the upper part of the respiratory tract and oropharynx. *Cancer,* **15:** 1142–1155.
Wiltshaw, E. (1976) The natural history of extramedullary plasmacytoma and its relation to solitary myeloma of bone and myelomatosis. *Medicine (Baltimore),* **55:** 217–238.
Woodruff, R. K., Whittle, J. M., Malpas, J. S. (1979) Solitary plasmacytoma I. Extramedullary soft tissue plasmacytoma. *Cancer,* **43:** 2340–2343.

Address for correspondence:
M. L. Navarrete,
C. Roca Umbert 5, 3° 1°,
Hospitalet,
08907 Barcelona,
Spain.

https://doi.org/10.1017/S0022215100114781 Published online by Cambridge University Press