Primary plasmacytoma of the thyroid

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Summary: Primary plasmacytoma of the thyroid is a very rare tumour. We report a case and emphasize the importance of excluding multiple myeloma which may present as an extramedullary plasmacytoma. Histological diagnosis has been simplified by the use of immunohistochemical techniques.

Introduction

Extra-medullary primary plasmacytomas are uncommon and their occurrence in the thyroid is rare.^{1,2} Only two cases have been reported from Britain.³ Histological diagnosis has been made much easier by the demonstration of a monotypic population of plasma cells, using immunohistochemical techniques. However, its presence as an early manifestation of multiple myeloma must be excluded. A primary plasmacytoma of the thyroid is described supported by immunohistochemical and ultrastructural examination. The presentation and management of these tumours is discussed.

Case report

A 78 year old woman presented with a 6-month history of a swelling in the left side of her neck. Her only symptom was mild dysphagia. Physical examination revealed a firm, smooth, non-tender, mobile enlargement of the left lobe of the thyroid with deviation of the trachea. Thyroid function, haemoglobin, serum proteins and electrolytes were normal. Chest X-ray showed marked tracheal displacement and an ultrasound scan confirmed the presence of a solid mass in the left lobe of the thyroid.

At operation, a well encapsulated tumour mass (8 cm × 4 cm × 3 cm) was found in association with enlarged lymph nodes lying both anterior to the gland and also within the left deep cervical chain. A left thyroid lobectomy with lymph node biopsy was performed.

Light microscopy showed thyroid and lymph node tissue diffusely infiltrated by sheets of mature

plasma cells (Figure 1). Immature and atypical forms of plasma cells were infrequent and mitoses rare. There were some residual thyroid follicles with marked lymphocytic infiltration suggestive of an underlying chronic thyroiditis.

Immunohistochemistry was performed using the unlabelled antibody (PAP) immunoperoxidase method with trypsinization for immunoglobulins (IgG, IgM and IgA) and for light chains (kappa and lambda). Plasma cells showed a striking intracytoplasmic staining for IgG and kappa light chains in both the thyroid and lymph node tissue. Staining for lambda light chains, IgM and IgA was negative. Electron microscopy confirmed the neoplastic features of the plasma cells, including well developed and stacked rough endoplasmic reticulum and numerous mitochondria. These are the features of a plasmacytoma.

Further investigations were performed to determine whether the tumour was solitary or a manifestation of multiple myeloma. The serum protein level was 82 g/l with an albumin/globulin ratio of 1.1:1 and a normal immunoglobulin profile. Protein electrophoresis showed a broad atypical band

Figure 1 Sheets of mature plasma cells with residual thyroid follicles containing colloid. Low power × 80.

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Accepted: 29 January 1990

in the gamma region which was identified by immunoelectrophoresis as IgG kappa. Protein electrophoresis and immunoelectrophoresis reverted to normal within 4 months of the operation. Bone marrow, skeletal survey and isotope bone scan were normal and Bence Jones protein negative.

Treatment was completed by local irradiation with 4000 cGy to the thyroid region and adjacent lymph nodes. The patient is well 18 months later with no evidence of local or distant recurrence.

Discussion

Extra-medullary plasmacytoma occurs either as a manifestation of multiple myeloma or as a solitary primary tumour. Oberkircher *et al.*⁴ found that 17% (24 of 141) of patients with multiple myeloma presented with extra-medullary tumours but the incidence of thyroid involvement has been reported to be only 2.6% (7 of 272) among extra-medullary plasmacytomas.⁵ Primary plasmacytoma of the thyroid is very rare. Hazard and Schildecke¹ found two cases following 14,000 thyroid operations performed over a 22-year period and MacPherson *et al.*² discovered one among 870 thyroid tumours. A review of the 28 cases of primary plasmacytoma of the thyroid which have been reported has shown that most patients present in the fifth to seventh decades and that there is no sex difference.

The diagnosis of primary plasmacytoma can be made only after the exclusion of multiple myeloma. Investigations should include pre- and post-operative serum electrophoresis, bone marrow examination and a skeletal survey or bone scan. Preoperative serum electrophoresis demonstrated an M-protein in 11 cases which by immunoelectrophoresis has shown four, including our own, with IgG kappa chains.^{2,4,6} In all cases, serum proteins reverted to normal within 2 to 9 months of removal of the tumour. Three of these patients had Bence

Jones proteinuria which also reverted to normal after treatment.⁷⁻⁹ Bone marrow examination was normal in all cases.

The histological distinction of primary plasmacytoma from plasma cell granuloma is difficult.¹⁰ Tissue immunostaining for immunoglobulins, by the PAP technique, has simplified the diagnosis since the demonstration of a monotypic population of plasma cells establishes the neoplastic nature of the lesion and correspondence with the immunoelectrophoretic characterization of serum protein establishes the site of production. Our case demonstrated a striking intracytoplasmic staining of IgG and kappa light chains in both the thyroid and lymph node tissue and corresponded with the serum immunoelectrophoretic finding of a raised IgG kappa protein.

Primary plasmacytomas of the thyroid appear to have an excellent prognosis even when there is involvement of adjacent lymph nodes. Patients have been followed for a median of 3 years (range 1-23 years). There has been no mortality from recurrent disease and the two tumours that recurred responded well to further treatment with either radiotherapy or chemotherapy.^{1,11} Hemi- or total thyroidectomy with regional lymph node biopsy is the initial treatment of choice. Local irradiation is probably an advisable adjunct to surgery when the juxtathyroid lymph nodes are involved by tumour. Further surgical clearance of these lymph nodes may be considered, although radical block dissections of the neck seem to be unnecessary because of the excellent prognosis of the tumour despite lymph node involvement. The rarity of plasmacytoma of the thyroid makes it difficult to determine optimum management for these patients.

Acknowledgements

We would like to thank Mr S.E. Knight, Consultant Surgeon and Dr M.H. Ali, Consultant Histopathologist, for their permission to report this case.

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