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Auris Nasus Larynx 35 (2008) 288–290

AURIS NASUS LARYNX
INTERNATIONAL JOURNAL OF ORL & HNS

www.elsevier.com/locate/anl

Thyroid cartilage involvement in patient affected by IgA multiple myeloma: Case report

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Received 1 November 2006; accepted 22 April 2007
Available online 6 September 2007

Abstract

Neoplasms originating from plasma cell are rare in the head and neck region. A correct clinical evaluation is very important in order to formulate a differential diagnosis as well as to distinguish local from metastatic disease. We report a case of larynx involvement by an IgA multiple myeloma in a 69-year-old female diagnosed in October 2004 and treated with chemotherapy; the 1 year control do not show progression of disease and the laryngeal lesion is unchanged. We highlight the radiological findings and clinical features to suspect plasma cell tumors in cases with a similar presentation.

Keywords: Multiple myeloma; Thyroid cartilage; Plasma cell tumor of larynx

1. Introduction

Neoplasms originating from plasma cell are rare in the head and neck region. They arise as a monoclonal proliferation of plasma cells during their various stage of differentiation (classified as a peripheral B-cell neoplasm in a revised European-American classification [1]). The clinical classification divides these neoplasm into three types: multiple myeloma (MM), solitary plasmacytoma of bone (SPB) and extramedullary plasmacytoma (EMP).

A correct clinical evaluation is very important in order to make the correct diagnosis as well as to distinguish this kind of tumor and the extent of disease, to institute the correct treatment and to estimate survival time. A plasma cell tumor classified as a solitary plasmacytoma of bone if evidenced in bone; in soft tissue as a extramedullary plasmacytoma, while the systemic form of the disease is called multiple myeloma.

These three clinical entities are considered as representing distinct manifestations of a continuum of disease. About 20–30% of EMP may progress to a MM [2].

We report the clinical and radiological features of a patient affected by IgA MM with laryngeal involvement, treated by chemotherapy; we outline the importance of differential diagnosis considering also this rare entity.

2. Case report

A 69-year-old woman affected by high-plasma level of IgA was diagnosed with a monoclonal gammopathy of undetermined significance (MGUS). She experienced progressive hoarseness and slight dysphagia. The patient did not complain of dyspnea nor did she have risk factors for laryngeal cancer (smoking and alcohol). She had no previous intubation, trauma or surgical procedures. She was a type II diabetic with hypertension, both efficiently controlled by medical therapy.

Indirect laryngoscopic examination revealed sub-mucosal swelling of the right ventricular fold (false cord), ventricle and true vocal cord, reduction of right side motility, normal mucosa (Fig. 1). A mass in the right thyroid ala was found during neck examination; after a CT scan without contrast enhancement of the neck, a structural alteration within thyroid cartilage in right ala with cortical thinning and right supraglottic and concentric cricoid ring thickness was evidenced (Fig. 2).

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doi:10.1016/j.anl.2007.04.011

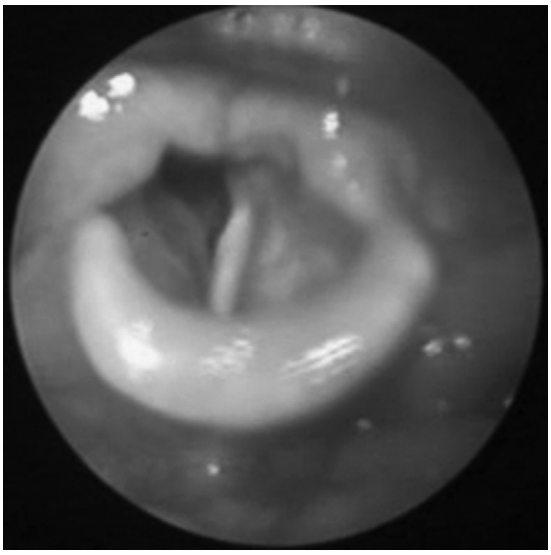

Fig. 1. Indirect laryngoscopic examination show a sub-mucosal swelling of right false cord, ventricle and true vocal cord and normal mucosal surface.

Biopsy of the mucosa and sub-mucosal layer was performed under direct microlaryngoscopy. Histopathology demonstrated a diffuse massive infiltration of plasma cells.

Haematological consultation was sought in order to further assess the nature of the disease. Serum and urine electrophoresis was performed which confirmed the previous findings of a high-plasma level of monoclonal IgA; bone marrow biopsy revealed plasma cell infiltration of 20%. Routine laboratory tests showed anaemia (haemoglobin 8.9 g/dl; RBC 2,980,000/mmc; WBC 3200/mmc; plasmatic albumin 3.5 g/dl, beta-2-microglobulin 5.6 g/dl). All these findings affirmed the diagnosis of MM stage IIIa. The patient underwent chemotherapy with VAD protocol (vincristine, doxorubicin and dexamethasone) for a total of 4 cycles; 1-year clinical and radiological follow-up did not show progression of disease with the laryngeal involvement unchanged.

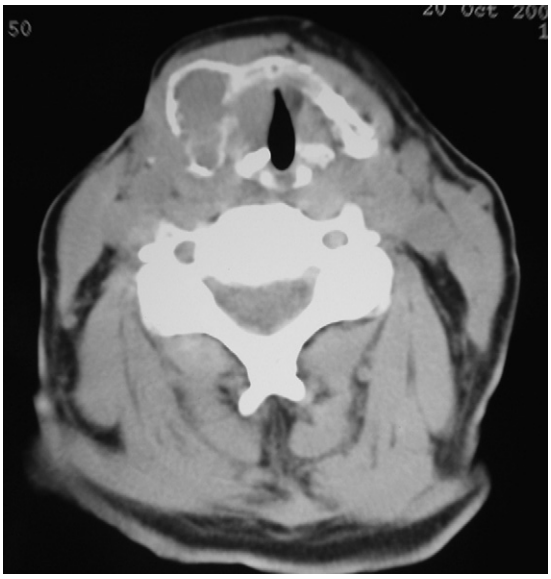

Fig. 2. CT scan of larynx without contrast enhancement showing a structural alteration within thyroid cartilage in right ala with cortical thinness.

3. Discussion

Laryngeal involvement in patients affected by MM is rare and few cases are reported in literature [3–9].

Two pathogenetics mechanisms are postulated by several authors to explain the involvement of laryngeal cartilage: (1) cartilage may be involved by an adjacent plasmacytoma; (2) cartilage, particularly in older people, may undergo to osseous metaplasia with formation of central marrow space, therefore a plasmacytoma may directly originate from this bone marrow [10]. Thyroid cartilage expansion and destruction evidenced in the CT scans suggests as in our case a myelomatous involvement of extraskeletal bone marrow formed by osseous metaplasia [8].

Consideration of this process is mandatory considering that laryngeal involvement in EMP is between 5% and 18% [11–13] with different treatment and with a better prognosis than MM. As a matter of fact EMP may evolve to MM many months or years later after diagnosis [14,15]. When a plasma cell tumor involving the larynx is diagnosed the next step is to exclude systemic disease. The patient must undergo detection of monoclonal gammopathy in serum or urine, a bone marrow aspiration biopsy and complete radiological examination in order to demonstrate osteolytic bone lesions.

Diagnosis of MM is based on: histological evidence of plasmacytoma or plasmacytosis in bone marrow; clinical signs of disease such as bone pain, anaemia and renal failure; detection of the monoclonal gammopathy in serum or urine and demonstration of osteolytic bone lesion. EMP and SBP are diagnosed if plasmacytoma is present in soft tissue or bone with no dissemination of disease.

Involvement of the thyroid cartilage is very rare [8,9]. CT examination can demonstrate a typical pattern: thinning and expansion of the lamina of the thyroid ala (as in our case), thus confirming its origin within the thyroid structure caused by osseous metaplasia of the cartilage [8]. Treatment of different form of plasma cells tumors is related to their extension: EMP and SBP can be treated by external radiation therapy with 40–50 Gy for a 4-week period including cervical lymph nodes in order to prevent lymph node recurrences [16]. Surgical removal is a second line treatment particularly in case of salvage therapy after radiation failure.

The treatment of symptomatic patients affected by MM, even with larynx involvement, involves chemotherapy or bone marrow transplantation. Surgery is mainly diagnostic in the majority of cases and tracheotomy is performed in case of airway obstruction.

Usually plasma cell neoplasms appear in the elderly while MM variant about 10 years later than the others two. After diagnosis MM have a mean survival of about 2–3 years, while EMP and SPB have a better prognosis.

4. Conclusion

Plasma cell neoplasms are unusual in the head and neck region and are histologically indistinguishable because they represent a continuum disease, as illustrated by progression of EMP and SPB to MM. The diagnosis is made on the basis of clinical, radiological and pathological findings, and the distinction between EMP, SPB and MM is critical for treatment and survival. In the case of typical CT scan larynx pattern a plasma cell tumor must be suspected for correct management.

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