Giant neck neoplasm. Case report

Neoplasia maligna gigante laterocervicale. Descrizione di un caso clinico

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Key words

Neck neoplasms • Malignant giant neoplasms • Case report

Parole chiave

Neoplasie latero-cervicali • Neoplasia maligna gigante • Caso clinico

Summary

Giant latero-cervical neoplasias usually originate in the parotid gland, as slow-growing adenomas, in subjects who take little care of their personal health. Giant adenomas of submandibular gland are very rare. These neoplasias involve prevalently male sex (male/female ratio: 2/1) and usually occur between 20 and 40 years of age. Signs of malignant transformation may be observed in the adenomatous epithelial component in a percentage ranging from 1% to 10% of cases. The case is reported of a giant malignant latero-cervical neoplasia originating from a pleomorph adenoma of the submandibular gland. The diagnostic work-up and treatment protocol are described.

Riassunto

Le neoplasie giganti latero-cervicali originano generalmente dalla ghiandola parotide, come adenomi a lento accrescimento, in soggetti con scarsa cura della propria salute. Gli adenomi giganti della ghiandola sottomascellare sono molto rari. Queste neoplasie interessano prevalentemente il sesso maschile (rapporto maschi/femmine: 2/1) e si manifestano, generalmente, tra i 20 e i 40 anni. Aspetti di trasformazione maligna si possono osservare a carico della componente epiteliale adenomatosa in percentuali variabili dall'1% al 10%. Viene descritto un caso clinico di neoplasia maligna gigante latero-cervicale ad origine da un adenoma pleomorfo della ghiandola sottomascellare, e discussa la strategia diagnostico-terapeutica adottata.

Introduction

Giant latero-cervical neoplasias usually originate in the parotid gland, as slow-growing adenomas, in subjects who take little care of their personal health [1-3]. Giant adenomas of submandibular gland are very rare [4], as also those located in the accessory salivary gland [5]. These neoplasias involve prevalently the male sex (male/female ratio: 2/1) and usually occur most frequently between 20 and 40 years of age [6]. In the cases described in the literature, the size and the weight of these neoplasms vary from 1 kg to > 25 kg [1-3 6].

Signs of malignant transformation may be observed in the adenomatous epithelial component in 1% to 10% of cases and this transformation is mainly related to the time taken to grow [1 3 6-8], with a risk of malignant degeneration which considerably increases after 10-15 years [8].

A case of giant malignant latero-cervical neoplasia originating from a pleomorphic adenoma located in submandibular gland is described. The diagnostic work-up and treatment protocol are discussed.

Case report

V.A., a 56-year-old housewife, came to our observation, in November 2001, presenting a giant esophitic neoplasia located in the left latero-cervical area, >30 cm in diameter, covered with distrophic and hyperaemic skin and with a large ulcerated area, which bled easily and was foul smelling (Fig. 1). As far as concerns the case history, the patient reported the presence of a neoplasm in the left submandibular site since the age of 16 years, which had grown very slowly and not exceeding 3-4 cm, for some 40 years. Ultrasonography examinations, periodically performed, indicated a submandibular gland origin, revealing adenomatous-like morphological features; cytological examination on fine needle biopsy confirmed the diagnosis of a pleomorphic adenoma of the submandibular gland. Over the last few years, the neoplasia had begun to grow more rapidly. Growth of the mass had become enormous and uncontrollable due to an accidental facial trauma which led to ulceration. The patient had been submitted elsewhere to a biopsy and computed tomography (CT) evaluation.

Fig. 1. Pre-operative appearance of neoplasia.

Fig. 3. Macroscopic appearance of the mass after surgical removal.

Histological examination revealed a poorly differentiated squamous cell carcinoma whilst the CT study revealed spread of the mass to the submandibular cavity and the left lateral and median region of the neck with apparent infiltration of the internal jugular vein. The mass appeared adhered to the hyoid bone, the larynx and mandibular body. The lymph nodes appeared to be hyperplastic and were widespread in the entire latero-cervical area as well as in the left parotid cavity (Fig. 2). The patient had been considered, elsewhere, as not amenable to surgery and was submitted to chemotherapy with cisplatin. Treatment was withdrawn after the third cycle due not only to intolerance but also to lack of effect on the neoplasia. Upon hospitalisation, in our division, the patient's general conditions were poor. Both eating and rest had become difficult on account of the position, the large size and foul smell of necrosis of the mass. The patient complained of a constant pain in the neck which spread in the left shoulder, the face and nape of the neck, exacerbating with displacement of the mass, she was in a state of severe depression and had suicidal intentions. Blood tests revealed severe anaemia (red blood cells (RBC) 2.6 mil/ml, haemoglobin (Hgb) 6.7 g/dl) and the patient was, therefore, submitted to blood transfusion resulting in a marked improvement (RBC 3.5 mil/ml, Hgb 10.1 g/dl).

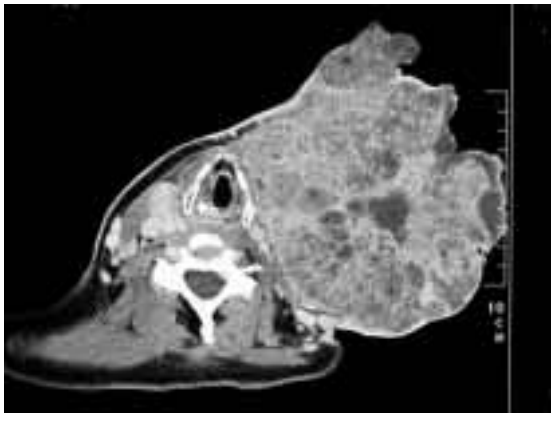

Fig. 2. Axial CT of neoplasia.

It was decided to proceed with surgical treatment for removal of the neoplastic mass, following selective embolisation, with catheterisation via the right retrograde femoral route, of the facial-lingual and upper thyroid trunks. Using this procedure, it was possible to achieve removal in toto of the neoplasm, which originated in the left submandibular gland, sparing the larynx, thyroid gland, hyoid bone and pre-laryngeal muscles which were not infiltrated. Furthermore, left radical neck dissection was performed with homolateral subtotal parothyroidectomy. Reconstruction of the large area of cutaneous exeresis was performed with a Bakamjiam delto-pectoral flap. The neoplasm was 35 cm in diameter and weighed 4600 g (Fig. 3). Histological examination confirmed the diagnosis of poorly differentiated squamous cell carcinoma with the base of attachment free from neoplastic infiltration. Within the neoplastic mass, areas were present which histologically referred to pleomorphic adenoma and the salivary origin of the neoplasia was confirmed by immunohistochemical evaluations carried out on the surgical specimen. The cervical and parotid lymph nodes were found to be inflamed.

The post-operative (p.o.) course was uneventful and the patient was discharged, completely healed, on the

12th p.o. day. At 6-month follow-up, the aesthetic outcome was good and the general psychological and physical conditions were good.

Conclusions

Only one case of giant malignant latero-cervical neoplasia of parotid gland origin has been described in the English literature [9]. Malignant transformation of pleomorphic adenomas of the submandibular gland is extremely rare [7 10].

The case described here is, to our knowledge, the first case, reported in the literature, of giant malignant latero-cervical neoplasia originating from a pleomorphic adenoma of the submandibular gland and, on account of the peculiarity of the clinical course, some considerations are worthwhile.

The extremely slow evolution of the neoplasia, ranging over a 40-year period, was characterised, in the year prior to surgery, by a sudden uncontrollable growth of the mass following trauma due to an accident. Trauma had probably accelerated the degeneration and the increase in size of the neoplastic mass. The greater risk of malignant degeneration due to accident trauma has been reported in the literature [1]. The enormous size of the mass and the presence of continuous pain, had a marked effect upon the quality of life of this patient, leading to a severe state of depression and rapid deterioration in general conditions. On the basis of clinical-instrumental findings, the mass had, at first, been considered not amenable to surgery and the patient was submitted to chemotherapy without any appreciable result. Surgical exeresis carried out, in our unit, initially for palliative purposes, revealed lack of malignant infilatration of the deep borders of the neoplasm and of latero-cervical lymph node metastases. These personal findings confirm, as already reported in the literature [1], that neoplastic degeneration of these giant masses, does not always involve the lesion in toto but only certain areas, mainly peripheral, without loco-regional features of aggressiveness, and suggest the need for surgical exeresis.

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Received: 15 October 2002
Accepted: 7 January 2003

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