Synovial cell sarcoma of the neck. Case report and review of the literature

Il sarcoma sinoviale del collo. Esposizione di un caso clinico e revisione della letteratura

F. BERTOLINI, B. BIANCHI, A. PIZZIGALLO, A. TULLIO, E. SESENNA
Dept. of Maxillofacial Surgery, University Hospital of Parma
Dept. of Maxillofacial Surgery, University of Sassari, Italy

Summary

Synovial sarcoma is an uncommon neoplasm accounting between 8 and 10% of all soft tissue malignancies. Involvement of the head and neck region is rare. So far, fewer than 100 cases have been reported in the literature and only a few originated in the soft palate, tongue, larynx, hypopharynx, and cervical oesophagus. We report the case of a synovial sarcoma of the parapharyngeal space in a 72-year-old female, and experience in the behaviour and surgical treatment of this neoplasm is described. The patient has remained disease free for 28 months, with regular follow-up. Based upon this case and the others reported in the literature, synovial sarcoma does not have a good overall prognosis, and on account of its rarity and unpredictable biologic behaviour, surgical excision, and regular clinical and radiographic follow-up for at least 3 years are strongly recommended.

Introduction

Synovial sarcoma (SS) is an uncommon neoplasm accounting between 8 and 10% of all soft tissue malignancies. The tumour originates primarily in the extremities, in close relation to tendon sheaths, bursae, and joint capsules. Involvement of the head and neck region is quite rare. The first documented report of head and neck synovial sarcoma was described by Jernstrom in 1954 in a case involving the pharynx. Since this time, fewer than 100 cases have been reported in the literature. The most commonly involved cervical sites are the neck, the retro- and the para-pharyngeal spaces. Only a few SS have been described originating in the soft palate, the tongue, the larynx, the hypopharynx, and the cervical oesophagus. A case of synovial sarcoma of the para-pharyngeal space is described and personal experience concerning the behaviour and treatment of this neoplasm are reported.

Materials and Methods

In November 1999, a 72-year-old female was referred to our department due to a slowly expanding, painless mass in the left cervical area that she had first noticed two years previously. She also complained a progressive dysphagia, change in her voice, and respiratory dyspnoea over the last month. The clinical examination revealed a left cervical oval mass, about 12x4 cm in size, firm-elastic in consistency, which displaced the laryngo-tracheal axis. There was no regional lymph node involvement. Endoscopy revealed an oval mass extending from the rhino-pharynx to the hypo-pharynx and displacing...
medially the left lateral pharyngeal and hypopharyngeal wall. The tumour was covered with normal mucosa.

A MNR examination with contrast revealed a large peri-pharyngeal and peri-laryngeal tumour mass with clear contours, extending from the naso-pharynx to C6-C7 (Figs. 1, 2). The incisional biopsy did not yield a definitive diagnosis, but showed a vascularized lesion.

A left external carotid artery angiogram demonstrated an area of tumour blush in the region of the lesion. The patient underwent pre-operative embolization of the left pharyngo-palatine artery and the left superior thyroid artery with polyvinyl alcohol particles (PVA) (150-250 micron) to minimize the risk of severe intra-operative bleeding.

The patient underwent tracheostomy and excision of the mass via a left cervicotomy and a midline mandibulotomy approach to reach the upper part of the lesion (Fig. 3). The definitive diagnosis of the lesion was “monophasic synovial sarcoma with haemangiopericytoma-like vascular structures”.

Macroscopically the mass was lobulated, 12x4x3 cm in size, and covered by a thin, but solid pseudo-capsule, with some necrotic areas (Fig. 4). The lesion was hypercellular with up to 12 per high-power field (HPF).

Histologically, SS showed a biphasic cellular pattern consisting of a stroma of fibroblast spindle-cells presenting scattered pale epithelial-like cells arranged in glandular formations, nests, or cleft-like spaces (Fig. 5). Complications occurred in the post-operative period, due to an ab-ingestis pneumonia with deterioration of the general conditions; moreover, due to persistent dysphagia, the patient required placement of a gastrostomy tube for a period of 6 months. During this period, she received radiation therapy (51 Gy) locally.

Results

At follow-up (last examination August 2002), the patient is healthy, the gastrostomy tube has been removed, and she has no evidence of recurrence or metastatic disease (Fig. 6).
Synovial sarcoma of the head and neck region was first described by Jernstrom in 1954, reporting on a case of malignant synovioma of the pharynx. Fewer than 100 head and neck synovial sarcomas have been reported in the literature. The most quoted study is that by Roth et al. who reported 24 cases from the Armed Forces Institute of Pathology (AFIP) in 1975. These patients had a median age of 19 years (range 10 to 51) with a male-to-female ratio of 7:5, and these Authors concluded that patients are more frequently affected in the early years of life (second to third decade).

Like Batsakis et al., Roth et al. suggested that SS originates from undifferentiated or pluripotential mesenchymal cells, thus explaining their occurrence in areas devoid of normal synovial tissue. A progressively enlarged, but well-circumscribed, slow-growing, painless, and nodular neck mass of several months’ duration is usually the first complaint. The overlying skin or mucosa are generally normal.

Histologically, SS is composed of two morphologically different types of cells which form the characteristic biphasic pattern, consisting of a stroma of fibroblast spindle-like cells in which pale epithelioid cells are scattered, arranged in glandular formations, nests, or cleft-like spaces. SS consisting of only one cell type has been described, for the first time, by Hajdu et al., but not all pathologists agree that this variant does really exist. Micro-calcifications are reported to be present in 30% to 60% of all cases.

The diagnosis of SS is not usually difficult, if the lesion is classically biphasic. However, when the tu-
mour is predominantly mono-phasic, a diagnosis of fibrosarcoma is frequently made. The myriad of histologic features is responsible for the divergence in diagnosis and initial misdiagnosis: SS also may be confused with malignant hemangio-pericytoma, malignant schwannoma, or spindle cell carcinoma. The occasional presence of micro-calcifications may misleadingly point to a metastatic thyroid tumour. Synovial sarcoma with predominantly epithelioid or glandular components may be confused with adenocarcinoma or carcinoma arising within a mixed salivary gland tumor. Despite the protracted course of the tumour, the overall prognosis is not good. Wright et al. reported a 2-year survival rate of 55%, while only 38% of patients survived for 6 years, with approximately 50% of all patients developing metastatic disease.

The lung is the principle site of metastatic disease, followed by the lymph nodes and bone marrow, according to Enzinger and Weiss. In another study reported by Duvall et al., the overall 5-year survival rate was from 36% to 18% after 10 years. Roth et al. reported a 5-year survival rate of 47% and stated that it was comparable independently of the anatomic site.

On account of the rarity and unpredictable biological behaviour of SS, management still remains a controversial issue. It is generally accepted that the treatment of choice is radical surgical excision. SS tends to spread beyond the visible and palpable limits of the tumour. Radical procedures, to achieve adequate margins in the head and neck region, are difficult to perform without sacrificing important anatomical structures. For these reasons, local recurrence of SS is a frequent finding.

Adjuvant radiotherapy is advocated by many Authors, to improve local control rates, albeit most SS of the extremities do not respond to radiotherapy. Lymph node dissection has not been advocated in the absence of lymphadenopathy. Most metastases are blood borne, but up to 20% spread through lymphatic channels to the regional nodes. According to some Authors, post-operative chemotherapy reduces distant metastases, but its role remains to be established. Theoretically, the greatest potential of chemotherapy lies in the prevention or delay of distant metastases.

As in SS at other sites, the size of the primary tumour may be the most important prognostic factor: patients with a tumour larger than 5 cm have a less favourable outcome. The age, histological grade, as well as calcifications are other prognostic indicators.

Conclusions

In the case described here, the old age of the patient, the deep neck location, and the size of the lesion (11 cm) did not permit wide excision including the surrounding tissues, since surgery would have been too aggressive, albeit surgical excision was macroscopically radical. After 33 months, the patient is healthy without local or distant recurrence, confirmed both clinically and radiographically.

References


Received: November 11, 2002
Accepted: June 17, 2003

Address for correspondence: Dr. F. Bertolini, Cattedra e Divisione di Chirurgia Maxillo-Facciale, Ospedale Maggiore, Via Gramsci, 14, 43100 Parma, Italy Fax: +39 0521 703761. E-mail: prezzemolo21@hotmail.com