Eur Arch Otorhinolaryngol (2003) 260 : 229–231
DOI 10.1007/s00405-002-0531-5

MISCELLANEOUS

F. Hostalet · D. Hellin · J. A. Ruiz

Tumefactive fibroinflammatory lesion of the head and neck treated with steroids: a case report

Received: 21 June 2002 / Accepted: 26 July 2002 / Published online: 14 November 2002

Abstract Tumefactive fibroinflammatory lesion is an idiopathic fibrosclerosing disorder of the head and neck region that clinically simulates a malignant process, but is histologically benign. This lesion is believed to be part of a broader fibrosclerotic syndrome that includes idiopathic mediastinal and retroperitoneal fibrosis, sclerosing cholangitis and Riedel's thyroiditis. The tumefactive fibroinflammatory lesion differs from other fibrosing conditions encountered in the head and neck region: fibromatoses, nodular fasciitis and fibrosarcomas. Although there is no optimum treatment, steroid therapy is suggested as the first line of management. Our patient was treated with corticosteroids and had a favourable response, supporting this approach as the initial treatment.

Keywords Fibrosis · Inflammatory · Lesion · Head and neck · Steroid therapy

Introduction

Tumefactive fibroinflammatory lesion is a term that has been applied to describe fibrosclerosing disorders occurring in the head and neck [15]. Rice et al. [10] are thought to have described the first case of fibroclerotic lesion in the soft tissues of the neck in 1975 and called it sclerosing cervicitis. Histologically identical lesions were subsequently reported involving the parotid gland [6], the nasal and paranasal sinus [11], maxillary antrum [9] and infratemporal fossa [7]. This lesion clinically simulates a malignant neoplasm, but is histologically benign. Its histopathologic appearance is nearly identical to that of fibloesclerotic lesions of the mediastinum and retroperitoneum and Riedel's thyroiditis [15]. The aetiology is unknown and the optimal treatment remains controversial. We report an additional case of tumefactive fibroinflammatory lesion of the head and neck, which responded dramatically to corticosteroid therapy.

F. Hostalet (✉) · J.A. Ruiz
Department of Pathology, Hospital "Vega Baja" de Orihuela,
Ctra Orihuela-Almoradi S/N, 03314 San Bartolome-Orihuela,
Alicante, Spain
Fax: +34-6-6776060

D. Hellin
Department of Otolaryngology,
Hospital "Vega Baja" de Orihuela, Alicante, Spain

Case report

A 74-year-old white female was referred to our institution for evaluation of a painless mass in the right side of the neck that had been slowly growing for 3 months. Hoarseness from vagal nerve paralysis was also a presenting complaint. The patient had no history of recent or remote trauma, she took no medication known to be associated with fibrosclerosing lesions in other locations and had never received radiation therapy.

Clinical examination showed a large and firm mass occupying the right side of the neck. The mass was fixed to the underlying structures, but the covering skin was freely mobile. Based on the symptoms and the clinical findings, the initial impression was of an underlying malignancy. Computed tomography (CT) scan demonstrated the obliteration of the normal anatomy with encasement of the great vessels by a large, ill-defined tumour, which strongly suggested a malignant process (Fig. 1).

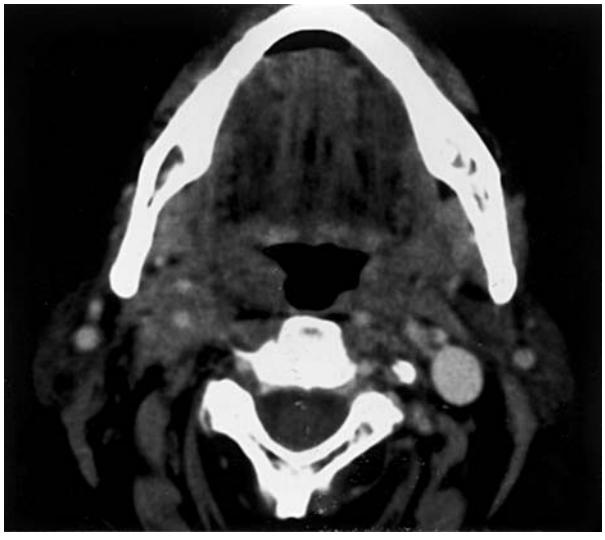

Fig. 1 CT Scan of the neck, showing a large, ill-defined mass encasing the great vessels in the right side of the neck

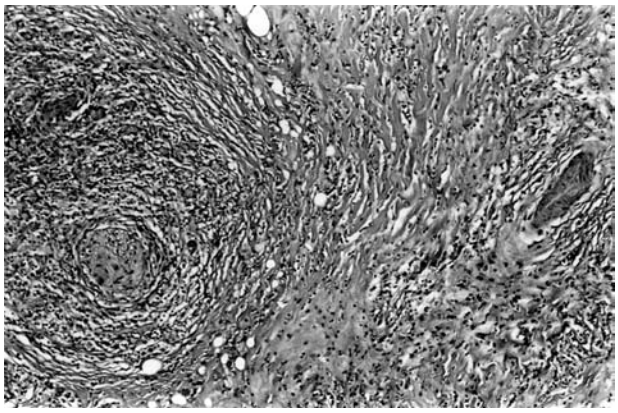

Fig. 2 Fibroinflammatory lesion shows proliferating fibrosclerotic tissue and prominent mononuclear inflammatory infiltrate with encasement of the neurovascular structures (HE, ×100)

Fig. 3 Typical collagenized stroma with scattered inflammatory cells, found within the lesion (HE, ×100)

An incisional biopsy of the mass was performed. Macroscopically, the biopsy tissue was firm and gray-tan. Histologically, the lesion was composed of proliferating fibrous tissue, bands of hyalinized collagen and prominent inflammatory infiltrate consisting mostly of lymphocites and plasma cells, which characteristically were encasing the vascular and neural structures (Fig. 2 and Fig. 3). There was mild cellullar pleomorphism, the mitoses were scarce and invasion of surrounding soft tissues was noted.

Immunohistochemical study showed that the proliferating spindle cells were reactive for vimentin, but they lacked muscle-specific actin, desmin, S-100 protein and Leu-7. The lymphocites expressed T and B cell markers, and the plasma cells displayed a polyclonal immunoglobulin staining pattern.

Silver chromate was used for identifying fungi and nite blue for acid-fast organisms, and tissue Gram's stains did not demonstrate organisms.

These histological and immunohistochemical features were consistent with tumefactive fibroinflammatory lesion of the head and neck. Subsequently, the patient underwent evaluation for other coexisting forms of fibrosclerosing lesions, but none were found.

The patient was treated with prednisone (80 mg per day) for 15 days and the hoarseness decreased. Subsequently, this dosage was gradually reduced over 3 months following clinical regression of the mass. When the treatment was finished, the patient was asymptomatic with no palpable mass. One month later a CT scan was performed showing clearly a pronounced shrinkage of the lesion. The corticosteroids were otherwise tolerated well; the only sequela was moderate hyperglycemia that required diet modification.

At follow-up 12 months after steroid therapy, the patient was well, without any complaints, and had no external evidence of recurrence.

Discussion

Tumefactive fibroinflammatory lesion of the head and neck is a rare disorder of obscure aetiology. This lesion is believed to be part of a broader fibloscleros ing syndrome that includes several lesions with similar histological appearance: retroperitoneal fibrosis, sclerosing cholangitis, mediastinal fibrosis and Riedel's thyroiditis. Inclusion of all these lesions in the syndrome implies a common causation and pathogenesis that is unproved. A periarterial location and encasement of neurovascular structures are seen in all three anatomic sites and point to a periarteritis as an underlying basis for the fibroinflammatory process [4]. The only reported example of tumefactive fibroinflammatory lesion in the upper extremity also had a neurovascular location [4, 12].

Histologic differential diagnosis of tumefactive fibroinflammatory lesion is primarily related to the more commonly encountered of the head and neck fibrous lesions. The fibromatoses [2, 3] that are encountered in the neck region are more cellular than the firm fibroinflammatory lesion. Although some fibromatoses may occasionally have reduced cellularity, they do not have the nest of inflammation seen in the fibroinflammatory lesion. Tumefactive fibroinflammatory lesion is not part of the spectrum of fibromatoses.

Nodular fasciitis also has been noted in the head and neck region [8]. This lesion is a pseudo-sarcomatous, self-limiting reactive process, which histologically can be distinguished from tumefactive fibroinflammatory lesion by the prominent whorled or feathery cellular pattern and lack of an inflammatory infiltrate.

The last group of fibrous lesions more commonly seen in the head and neck area includes the malignant fibrous histiocytoma [14] and fibrosarcoma [13]. Tumefactive fibroinflammatory lesion is infiltrative, but it lacks the dense cellularity, cellular atypia and mitotic activity of a sarcoma.

The clinical diagnosis of fibroinflammatory lesion is difficult. The radiographic and clinical appearances can be misleading, and it is often thought to be a malignant tumour before biopsy. Furthermore, tumefactive fibroinflammatory lesion of the head and neck can be a localized manifestation of a systemic condition [1]. Accordingly, patients should be evaluated for fibrosclerosing lesions at the other sites.

There is no consensus regarding the optimal treatment of this lesion. Treatment approaches have included surgery, steroids and radiation therapy, used either alone or in combination with variable outcomes [5]. Olsen et al. [6] described a patient with cervical lesion that responded completely to treatment with high-dose steroid therapy. Our patient was treated with prednisone and had a favourable response, supporting this approach as the initial treatment. However, the dosage and duration of treat-

ment are not well established. Surgery is generally reserved for lesions that do not respond to steroid therapy; however, the location and extent of infiltration may not permit complete surgical removal. Radiation therapy has also been used for lesions that are unresponsive to treatment with steroids when the tumour's extent precludes safe surgical removal.

Patients with tumefactive fibroinflammatory lesion require long-term follow-up. Despite the nonmalignant nature of this lesion, recurrence and disease persistence are common. Recognising and following more cases will lead to a better understanding of the behaviour and treatment of this lesion.

Acknowledgements We are very grateful to Mr H. Garcia Gimenez, professor of English in the I. E. S. Thader, for advice in the writing of this manuscript.

References

1. Amorosa LF, Shear MK, Spiera H (1976) Multifocal fibrosis involving the thyroid, face and orbits. Arch Int Med 136: 221–223
2. Conley J, Haeley WV, Stout AP (1966) Fibromatosis of the head and neck. Am J Surg 112: 609–614
3. El-Sayed Y (1992) Fibromatosis of the head and neck. J Laryngol Otol 106: 459–462
4. Frankenthaler R, Batsakis JG, Suarez PA (1993) Tumefactive fibroinflammatory lesions of the head and neck. Ann Otol Rhinol Laryngol 102: 481–482
5. Laurenzo JF, Graham SM (1995) Tumefactive fibroinflammatory lesion of the head and neck: a management strategy. Ear Nose Throat J 74: 87–91
6. Olsen KD, DeSanto LW, Wold LE, Weiland LH (1986) Tumefactive fribroinflammatory lesions of the head and neck. Laryngoscope 96: 940–944
7. Patel PC, Pellitteri PK, Vrabec DP, Szymanski M (1998) Tumefactive fibroinflammatory lesion of the head and neck originating in the infratemporal fossa. Am J Otolaryngol 19: 216–219
8. Price EB Jr, Silliphant MM, Shuman R (1961) Nodular fasciitis: a clinicopathologic analysis of 65 cases. Am J Clin Pathol 35: 122–136
9. Prichard AJN, Colloby RPE, Heaton JM (1990) Tumefactive fibroinflammatory lesions of the head and neck. J Laryngol Otol 104: 797–800
10. Rice DH, Batsakis JG, Coulthard SW (1975) Sclerosing cervicitis: homologue of sclerosing retroperitonitis and mediastinitis. Arch Surg 110: 120–122
11. Said H, Hadi AR, Akmal SN, Lokman S (1988) Tumefactive fibroinflammatory lesions of the head and neck. J Laryngol Otol 102: 1064–1067
12. Savage PD, Wick MR, Thompson RC, Skubitz KM (1991) Tumefactive fibroinflammatory lesion of the extremity. Report of a case and review of the literature. Arch Pathol Lab Med 115: 230–232
13. Sessions DG, Ogura JH (1974) Fibrosarcoma of the head and neck. Ann Otol 184: 439
14. Weiss SW, Enzinger FM (1978) Malignant fibrous histiocytoma: an analysis of 200 cases. Cancer 41: 2250–2266
15. Wold LE, Weiland LH (1983) Tumefactive fibroinflammatory lesions of the head and neck. Am J Surg Patho1 7: 477–482