Received: 26 October 2001
Revised: 22 March 2002
Accepted: 8 April 2002
Published online: 20 June 2002
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Abstract Background: Solitary fibrous tumor (SFT) is a rare spindle cell tumor that arises most often in the visceral pleura; however, a review of the literature shows at least 31 cases occurring in the orbit.

Methods: A retrospective case series of three patients with orbital SFT: a 50-year-old man, observed in 1997, with an angioma-like lesion in the upper half of the orbit causing osteolysis of the orbital roof; a 24-year-old man, observed in 1992, with a superotemporal mass in the right orbit occupying the lacrimal gland region, firstly diagnosed as schwannoma, recurring 4 years after dacryoadenectomy; a 70-year-old man, with a retrobulbar mass diagnosed on a biopsy as hemangiopericytoma, recurring and infiltrating the orbital roof 4 years after surgery.

Results: A review of the literature and presentation of three cases of SFT which showed infiltration of the orbital roof and/or recurrence.

Conclusions: Our cases provide evidence of how orbital SFT can behave aggressively and mimic other orbital tumors, thus making mandatory the consideration of this relatively new entity in common clinical practice as well as careful follow-up. Their aggressive growth is unusual, described in only 6 of the 31 cases so far reported in the literature. Immunohistochemistry is of importance for the diagnosis, since CD34 immunoreactivity is peculiar to SFT.

Introduction

Solitary fibrous tumor (SFT), a rare neoplasm first described in the pleura [4, 5, 6, 7] may occur at many different sites. We review the literature on the subject and report three new orbital cases: two were localized in the upper half of the orbit and one in the lacrimal gland fossa. They were characterized by unusually aggressive behavior, consisting of the infiltration of the orbital roof and/or recurrence, and created problems regarding differential diagnosis and treatment. We stress the diagnostic importance of CD34 immunoreactivity; it excludes other orbital soft tissue tumors, and in its absence the diagnosis may be erroneous.

Case reports

Case 1

In 1997 a 50-year-old man with a 9-month history of increasing swelling of the left eye presented with a well-circumscribed lesion in the upper half of the orbit on contrast-enhanced orbital CT; an angiomatous lesion was hypothesized (Fig. 1a). A transconjunctival approach disclosed a white, solid mass, which was biopsied. Histopathological diagnosis indicated a mesenchymal tumor, most probably an orbitally localized SFT.

Immediately, complete removal of the tumor was performed superotemporally by lateral orbitotomy. Osteolytic areas were noted on the orbital roof with leakage of cerebrospinal fluid; thus, an osteoplasty was performed.

The patient subsequently underwent radiotherapy. No recurrence was observed in the following 4 years.

Case 2

In 1992 a 24-year-old man presented with a 3-year history of a painless, palpable superotemporal mass in the right orbit leading
to progressive proptosis. Contrast-enhanced CT revealed a weakly hyperdense mass in the lacrimal gland region, with well-defined borders, causing exophthalmos and superficial bone erosion; radiologically, the mass had the appearance of a pleomorphic adenoma (Fig. 1b).

A complete dacryoadenectomy was performed and a histopathological diagnosis of schwannoma was made. After 4 years, the mass recurred and an extensive resection was performed. Histopathological and immunohistochemical examination of the material yielded the diagnosis of SFT. No recurrence was observed in the following 5 years.

Case 3

In 1994 a 70-year-old man presented with marked proptosis of the left eye since the age of 50 years. Contrast-enhanced CT revealed a retrobulbar mass which showed areas of relative hypodensity (Fig. 1c). Due to a recent mild increase in the proptosis, a transconjunctival biopsy of the mass was performed. A histopathological diagnosis of schwannoma was made. After 4 years, the mass recurred and an extensive resection was performed. Histopathological and immunohistochemical examination of the material yielded the diagnosis of SFT. No recurrence was observed in the following 5 years.

Methods

All the histologic slides from the excised lesions and recurrences were reviewed; new slides for routine stains and immunohistochemistry were obtained from paraffin blocks. The following monoclonal antibodies were used: CD34 (stem cells, endothelial cells; Dakopatts, Carpinteria, Calif.; diluted 1:100); α-smooth muscle actin (smooth muscle cells; Sigma, St. Louis, Mo.; 1:8,000); vimentin (mesenchymal cells; Dakopatts; 1:500), S-100 (S-100 protein; Biogenex, San Ramon, Calif.; 1:400), HHF35 (Total actin; Biogenex; 1:1), epithelial membrane antigen, EMA (epithelial cells; Dakopatts; 1:100), CD31 (endothelial cells; Dakopatts; 1:100), neurofilaments (neural cells, Dakopatts; 1:200), glio-fibrillary acid protein, GFAP (glial cells; Dakopatts; 1:200). After overnight incubation in the antigen, sections were stained by the alkaline phosphatase antialkaline phosphatase (APAAP) technique (Dakopatts), and colors were developed with new fuchsin. Appropriate positive and negative controls were used.

Pathological findings

All tumors were firm, with neither cystic spaces nor necrosis; those in cases 1 and 2 were gray-white in color, while the mass in case 3 was wine-red and showed hemorrhagic areas. Histologically, the tumors showed alternating hypercellular, dense areas without intermixed stroma and hypocellular areas with bundles of dense, sclerotic collagenous tissue around isolated spindle-shaped cells. A hemangiopericytoma-like pattern was detectable throughout various microscopic fields in case 3 (Fig. 2). Vascularity was less conspicuous in cases 1 and 2. Mitoses were less than 1 per 10 high-power fields in cases 1 and 2 and were 4 to 5 in case 3; atypical mitoses were absent. The final diagnoses of SFT were made on the basis of the above-mentioned findings and immunohistochemistry: CD34 and vimentin immunoreactivity was strong and consistent in the tumor cells, while muscular, epithelial, vascular, neuronal and glial markers were negative; S100 protein was weakly positive in case 2.

Discussion

Solitary fibrous tumors are rare neoplasms which arise most often in the pleura. Their intense immunoreactivity for CD34 [26] and the fibroblastic nature of tumor cells on electron microscopy [22] have definitely demonstrated their mesenchymal origin. This has opened the way for their description in many extrapleural sites not associated with serosal surfaces and has led to the abandon-