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Case Report

Giant solitary fibrous tumor of the orbit with aggressive behavior

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ABSTRACT

Solitary fibrous tumor (SFT) is a rare spindle cell neoplasm that was initially described as a mesenchymal neoplasm in the pleura. SFT have subsequently been diagnosed in the pericardium, mediastinum, liver, breast, peritoneum, nasopharynx, paranasal sinuses, salivary gland, thyroid gland, and orbit. Histopathological and immunohistological features are crucial for a correct differential diagnosis, because the neoplastic cells of SFT express CD34, thus differentiating this tumor from other spindle cell neoplasms. Orbital SFTs usually present an indolent and benign course with unilateral painless proptosis, visual disturbance, palpable orbital mass, ocular mobility disturbance, hyperglobus or blepharoptosis from the secondary mass effect in the orbit. However, few cases have shown aggressive behavior with adjacent tissue invasion, recurrence after partial resection, or malignant transformation with metastasis.

The purpose of this article is to present and discuss a case of orbital solitary fibrous tumor with aggressive behavior due to intracranial invasion.

1. Introduction

Solitary fibrous tumor (SFT) is a rare spindle cell neoplasm that was initially described as a mesenchymal neoplasm in the pleura [1–5]. SFT have subsequently been diagnosed in the pericardium, mediastinum, liver, breast, peritoneum, nasopharynx, paranasal sinuses, salivary gland, thyroid gland, and orbit [1–3]. About 80 cases of orbital SFTs have been published in the English literature, although this seems to be an underestimated number, considering the advances that immunohistochemistry has provided for the diagnosis of this tumor [1–5].

In fact, histopathological and immunohistochemical features are crucial for a correct differential diagnosis, because the neoplastic cells of SFT express CD34, thus differentiating this tumor from other spindle cell neoplasms [1,2].

Orbital SFTs usually present an indolent and benign course with unilateral painless proptosis, visual disturbance, palpable orbital mass, ocular mobility disturbance, hyperglobus or blepharoptosis from the secondary mass effect in the orbit [1]. However, few cases have shown aggressive behavior with adjacent tissue invasion, recurrence after partial resection, or malignant transformation with metastasis [1–5].

The purpose of this article is to present and discuss a case of orbital solitary fibrous tumor with aggressive behavior due to intracranial invasion.

2. Case report

An 85-year-old woman presented with a 36 months history of progressive proptosis of the left eye (Fig. 1). She referred a current complete inability in vision of the left eye and nasal obstruction.

Past medical history included diabetes, hypertension, osteoporosis, previous multiple myocardial infarctions, and advanced Alzheimer disease.

On clinical examination, she presented left proptosis, left periorbital swelling, and a dilated and nonreactive left pupil. Visual acuity was below perception of light. No extraocular movement were present.

Computed tomography (CT) scan showed a giant ill-demarcated left orbital mass. The mass extended to the nasal fossa bilaterally and it showed intracranial invasion with destruction of the bone surrounding the orbit (Fig. 2). The left external carotid angiogram

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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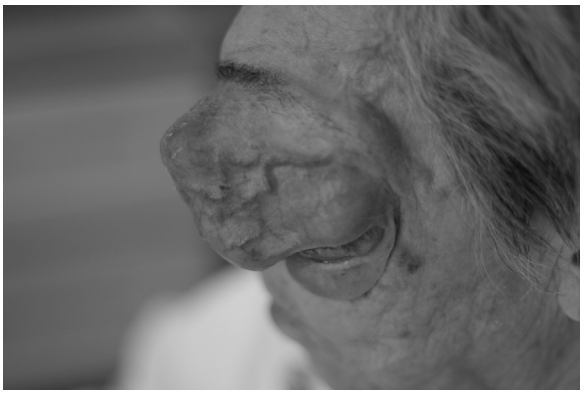

Fig. 1. Clinical appearance of the orbital lesion.

showed that the tumor was fed by the left internal maxillary artery, the left superficial temporal artery, and the left ophthalmic artery.

An incisional biopsy of the mass was performed under local anesthesia. Pathological examination disclosed a spindle-cell tumor in a dense fibrous tissue. The suspect of the aggressive nature of the lesion arose because of the findings of necrosis areas, nuclear atypia, and discrete mitotic index (till 5/10 high-power fields). Immunohistochemical stains of the tumor demonstrated a positive staining for Bcl-2 and CD34 but negative for CD-31, EMA, CK AE1-AE3, Actin, CD-117, GFAP and S-100 protein. The proliferation index Ki-67 was about 30%. Therefore, the tumor was finally diagnosed as a SFT.

In consideration of the poor general condition of the patient, the aggressiveness of the tumor, and the nasal and intracranial extensions of the lesion, the embolization of the tumor was decided to limit the growth of the SFT.

Therefore, the patient underwent embolization of the main feeding branches of the tumor that were the left internal maxillary artery, the left superficial temporal artery, and the left ophthalmic artery. The lesion was no more vascularized at the angiogram control (Fig. 3).

Then, the patient was dismissed and underwent a monthly follow-up.

The patient died 6 months later.

3. Discussion

Solitary fibrous tumor is an uncommon benign tumor that usually occurs in the pleura, mediastinum, or lung. Extrapleural sites include the peritoneum, pericardium, upper respiratory tract, nasal cavity, paranasal sinuses, parotid gland, mediastinum, liver, testis, thyroid gland, salivary glands, and orbit [1–5].

These lesions mainly present in the fifth decade, but range from the first to the eight decade, with a slight male predilection. Diagnosing SFT by clinical presentation alone is impossible and the lack of specific histological features initially pose problems for establishing a differential diagnosis with other mesenchymal tumors [4,5].

Clinically, SFT of the orbit usually present as slowly growing unilateral painless proptosis inducing severe facial deformity and may be associated with eyelid swelling, vision disturbances, tearing, and ptosis. Nasal obstruction is referred when there is a sinonasal involvement, as in our case. They less frequently may be associated with optic nerve dysfunction and extra ocular muscle impairment depending on their size and location [3–5].

On imaging, orbital SFT presents as a well or ill-defined soft tissue mass with strong enhancement on CT and MR imaging. The most common radiographic osseous finding on both CT and MR imaging is regressive remodeling of adjacent bone due to the long-standing pressure effect of the slow growing mass [3]. If extensive bone remodeling is present or a long standing lesion is noted, the possibility of malignant transformation should be suspected. On MRI, SFTs appear hypointense to isointense on T1-weighted images and iso/hypo-intense on T2-weighted images [1–5].

Histologically, SFT is a well-circumscribed, lobulated, rubbery mass that shows a patternless arrangement of alternating hypercellular and hypocellular regions of spindle cells separated by thick bands of hyalinized collagen of variable vascularity. In some cases, a focal hemangiopericytoma-like pattern of irregular branching vessels, fibrous histiocytoma-like storiform pattern, and synovial sarcomatous and neural-like pattern of palisading regional architecture can be observed [1–5]. This variability on histologic examination may lead to a mistaken diagnosis if limited tissue allows a single morphology to dominate the pathologic specimen [1–5].

The diagnosis of SFT can be confirmed by immunohistochemical analysis. In fact, CD34 is the most important marker to diagnose SFT, and its diffuse and strong immunoreactivity has been demonstrated in 79–100% of cases, as in our case [1–5]. However, varying degrees of CD34 reactivity can also be present in other orbital tumors, such as hemangiopericytoma, fibrous histiocytoma, schwannoma, neurofibroma, fibrosarcoma, giant cell angiofibroma, and giant cell fibroblastoma. In addition to CD34, SFT shows strong and diffuse positivity for B-cell lymphoma 2 (bcl-2) but negativity for keratin, cytokeratin, epithelial membrane antigen, S-100 protein, smooth muscle actin, factor VIII-related antigen, and desmin [1–5].

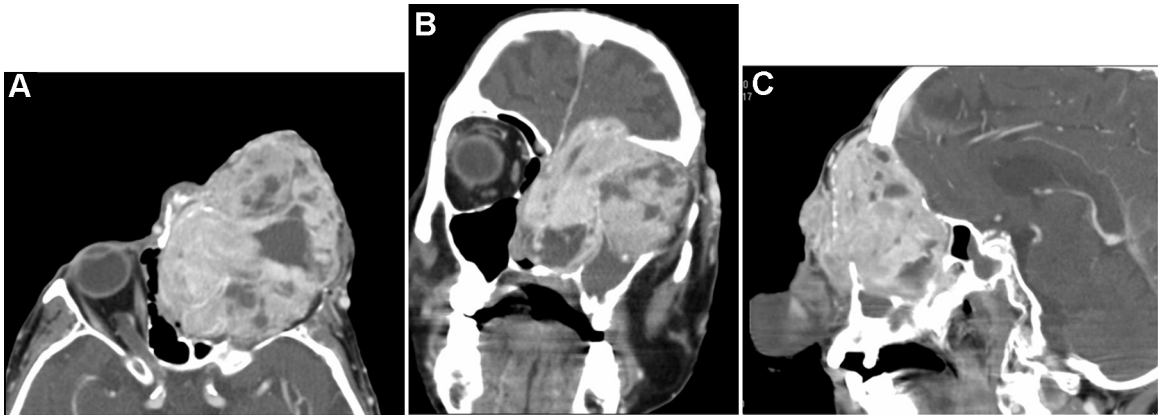

Fig. 2. Axial (A), coronal (B), and sagittal (C) views of the computed tomography showing a giant ill-demarcated left orbital mass, that presented both intracranial and sinonasal invasion with bony destruction.