OLGU SUNUMU CASE REPORT

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An Uncommon Orbital Neoplasm: Solitary Fibrous Tumor: Case Report

Ender Görülen Bir Orbita Neoplazmı: Soliter Fibröz Tümör

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ABSTRACT The purpose of this case report is to present an intraconally located orbital solitary fibrous tumor, an uncommon fibroblastic tumor of the orbit which has diagnostic challenges. A 33 years old female patient presented with three years of slowly progressive mass and proptosis of the left eye. Magnetic resonance imaging demonstrated a well-defined, strongly enhancing intraconal orbital mass. Histopathological and immunohistochemical analysis following en - bloc removal of the lesion revealed a mesenchymal tumor showing strong positivity for CD34 and Vimentin. The diagnosis was solitary fibrous tumor. Although uncommon, solitary fibrous tumor should be kept in mind in the differential diagnosis of a well-defined orbital mass associated with slowly progressive proptosis.

Keywords: Exophthalmos; solitary fibrous tumors; antigens, CD34; immunohistochemistry; orbital neoplasms

ÖZET Bu makalede, yavaş ilerleyici propitozisin az rastlanan bir sebebi olan intrakonal yerleşimli bir orbital soliter fibröz tümör (SFT) olgusunu sunmayı amaçladık. Otuz üç yaşında kadın hasta, sol gözünde 3 yıldır yavaş büyüyen kitle ve proptozis şikayeti ile başvurdu. Magnetik rezonans görüntülemede, iyi sınırlı, yoğun kontrast tutan intrakonal kitle tespit edildi. Kitle lateral orbitotomi yoluyla bütünüyle eksize edildi. Çıkarılan 31x31x35 mm boyutlarındaki kitle, immunohistokimyasal analizi de içeren histopatolojik inceleme sonucunda, güçlü CD-34 ve Vimentin pozitifliği gösteren mezenkimal tümör olarak tanımlandı. Bu bulgular ışığında olguya benign orbital SFT tanısı konuldu. Yavaş ilerleyici proptozise sebep olan iyi sınırlı orbital kitlelerin ayırıcı tanısında SFT akılda tutulmalıdır.

Anahtar Kelimeler: Ekzoftalmus; soliter fibröz tümörler; antijenler, CD34; immünohistokimya; orbita neoplazileri

Solitary fibrous tumor (SFT) was first described as a primary spindle cell tumor of the pleura in 1931 and has been subsequently reported in extra-pleural sites.[1-4] Orbit is an uncommon location for SFT, where diagnosis is challenging and requires immunohistochemical analysis along with clinical, radiological and pathological evaluation. Besides, the nomenclature of specialized fibroblastic tumors of the orbit has changed recently. CD34 immunopositive fibroblastic neoplasms, which were previously classified as separate entities, are now comprised as SFT.[5] In this case report, we present a case of an orbital SFT treated with complete surgical excision.

CASE REPORT

A 33 year old woman presented with three years of slowly progressive mass and proptosis of the left eye associated with occasional left periorbital pain and diplopia. She did not have any additional health problems. Ophthalmological examination revealed best corrected visual acuities of 20/20 and 20/32 in the right and left eye respectively. The biomicroscopic anterior segment and fundoscopic findings were normal in both eyes. Motility examination revealed diplopia on left gaze and a mild left hipertropia. The left globe was superomedially displaced. Magnetic resonance imaging demonstrated a well circumscribed 31x31x35 mm solid intraconal mass in the left orbit which displaced the left optic nerve laterally. The mass showed hypointensity on T1 and heterogeneous mixed intensity on T2 weighted images. It also contained a signal void tubular structure. On post contrast T1 images there was marked enhancement of the mass. No sign of bony destruction was noted (Figure 1). Considering these findings, surgical removal of the tumor was planned.

The lesion was approached through an S shaped rim incision starting from the upper skin crease extending beyond the lateral canthus. Following tissue dissection, the periost of the lateral orbital rim was incised and elevated. With the bone drill device, the lateral rim was trimmed. The tumor was visualized and isolated from surrounding tissues. En bloc excision was achieved after the tumor was secured with 2-0 silk sutures. Postoperatively, there was marked improvement in proptosis, globe displacement, ocular motility restriction and diplopia.

On pathological examination, the specimen was an encapsulated nodular tissue measuring 36x25x24 mm, a solid gray-white mass when sectioned (Figure 2). Microscopic examination revealed a benign mesenchymal tumor consisting of small fusiform cells individually separated by thin bands of collagen fibers arranged in a storiform and fasicular pattern (Figure 3A). Tumor showed alternating hypercellular and hypocellular components represented by bluish perivascular tumor cell aggregation and pinkish hyalinized areas (Figure 3B). Slit like and stag horn-like vascular channels were identified scattered throughout the tumor. No mitotic figures, nuclear polymorphism or necrosis were noted. Reticulin stain showed moderately rich reticulin network (Figure 3C). Immunohisto-

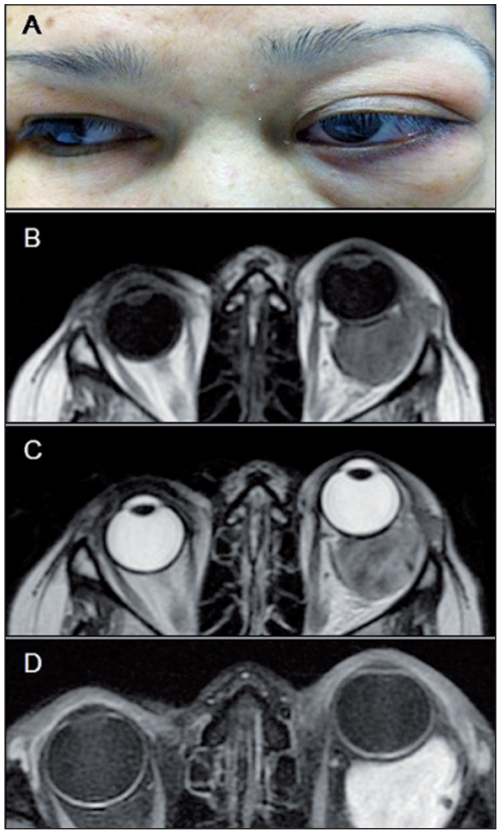

FIGURE 1: Preoperative findings: **(A)** Clinical image of the patient pre-operatively. **(B)** Left intraconal hypointense mass on T1 weighted MR image. **(C)** Heterogeneous hyperintense and hypointense signal on T2 weighted MR image **(D)** Intense enhancing of the mass on postcontrast T1 image.

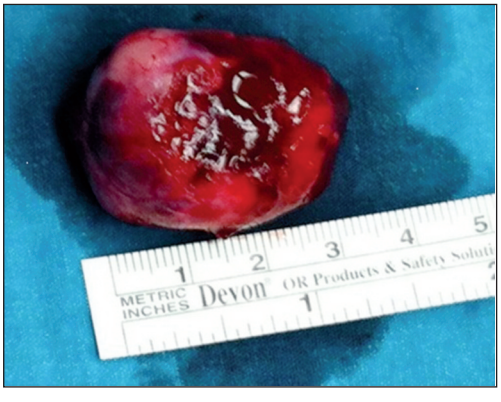

FIGURE 2: Macroscopic image of the resected tumor: 36x25x24 mm encapsulated nodular tissue.

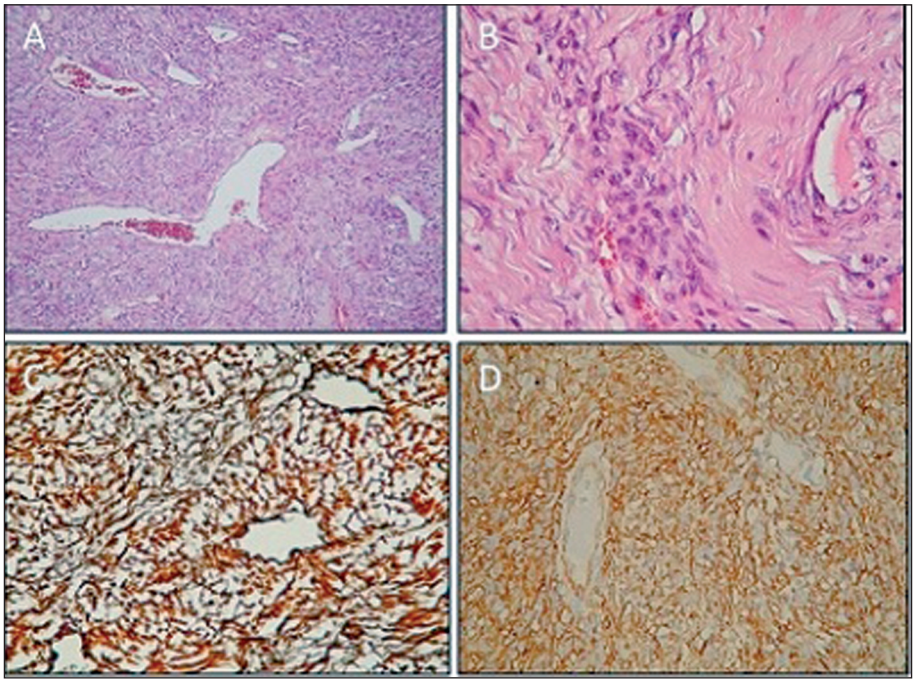

FIGURE 3: Microscopic and immunohistochemical findings: **(A)** x100, hematoxylin and eosin stain, benign fusiform cells composing storiform pattern. **(B)** x400, hematoxylin and eosin stain, hypocellular area composed of parallel arrays of collagen, separated by hypercellular fibroblastic nuclei. **(C)** x200, reticuline stain, rich reticuline network **(D)** x200, immunohistochemical staining, strong and diffuse positivity for CD34.

chemical work-up revealed strong positivity for CD34 (Figure 3D) and vimentin. Keratin, S100, desmin, CD99, CD117 was negative. These findings were consistent with a diagnosis of orbital SFT.

DISCUSSION

An orbital SFT has an indolent clinical course, usually presenting with slowly progressive proptosis and a non-tender, non-pulsatile mass, between the fourth and sixth decades of life. [6] Other symptoms include diplopia, reduced visual acuity and pain. Many other orbital neoplasms like cavernous hemangioma, schwannoma, neurofibroma, meningioma, leiomyoma, fibroma, fibrosarcoma, lymphoma and rhabdomyosarcoma share a similar clinical presentation. Computerized tomography or magnetic resonance imaging findings are also of limited value in distinguishing SFT from other well-defined orbital masses, but still can provide us some useful clues. SFT is usually seen as a well-defined ovoid mass that can be located in various sites in the orbit either extraconally or intraconally. Bone destruction is rare and only noted on malignant tumors. The common feature of SFT is homogeneous or heterogeneous enhancement on CT and MR images, reflecting the high vascularity of the tumor. Besides, signal void structures within the tumor have been reported.[7] This is an interesting and unusual finding which we also noticed in our case, possibly representing fast-flow vessels. The heterogeneous hypointense and hyperintense signal on T2 weighted MR images of SFT reflects different amounts of cellular components, collagen and fibroblasts as well as necrosis, degeneration or hemorrage within the tumor. Cavernous hemangioma which is the most common orbital tumor in adults can be differentiated from SFT by its hyperintensity on T2 images. Other tumors like schwannoma, neurofibroma and lymphoma show less noticable enhancement compared to SFT. [8]

Major histopathological differential diagnosis of orbital SFT includes hemangiopericytoma, schwannoma, fibrous histiocytoma, leiomyoma, leiomyosarcoma and optic nerve sheath meningioma. Although they have overlapping morphological features, a clear distinction can be made

among these tumors with immunohistochemical analysis. Strong and diffuse CD34 staining is characteristic for SFT. Hemangiopericytoma and fibrous histiocytoma reveal inconsistent and weak positivity for CD34. Schwannoma shows strong positivity for S-100 protein, leiomyoma and leiomyosarcoma show positivity to actin and desmin. Fibrous and transitional meningiomas are negative for CD34 and show positivity for epithelial membrane antigen (EMA).[9]

The morphological resemblance of such tumors may have led to the misdiagnosis of orbital SFT in the past. Recently, 41 cases of fibroblastic orbital tumors which were originally diagnosed as hemangiopericytoma (n=16), fibrous histiocytoma (n=9), mixed tumors (n=14) and giant cell angiofibroma (n=2), have been reclassified in a new spectrum under the encompassing terminology of orbital SFT.[5]

In conclusion, SFT should be kept in mind in the differential diagnosis of a well-defined orbital mass with an indolent clinical course. Strong CD34 staining is the most prominent diagnostic feature of SFT. Orbital SFT has a potential to recur and metastasize; therefore en-bloc and atraumatic excision of the tumor and careful postsurgical follow-up is recommended.

Conflict of Interest

Authors declared no conflict of interest or financial support.

Authorship Contributions

Idea/Concept: *Okan Toygar, Levent Akçay, Baha Toygar;* ***Design:*** *Okan Toygar, Levent Akçay, Özge Yabaş Kızıloğlu;* ***Control/Supervision:*** *Okan Toygar, Levent Akçay, Baha Toygar;* ***Data Collection and/or Processing:*** *Okan Toygar, Özge Yabaş Kızıloğlu, Özlem Yapıcıer, Mustafa Kemal Demir;* ***Analysis and/or Interpretation:*** *Okan Toygar, Özge Yabaş Kızıloğlu, Baha Toygar, Levent Akçay, Özlem Yapıcıer, Mustafa Kemal Demir;* ***Literature Review:*** *Okan Toygar, Özge Yabaş Kızıloğlu;* ***Writing the Article:*** *Okan Toygar, Özge Yabaş Kızıloğlu, Levent Akçay, Özlem Yapıcıer, Mustafa Kemal Demir, Baha Toygar;* ***Critical Review:*** *Okan Toygar, Baha Toygar, Özge Yabaş Kızıloğlu, Levent Akçay, Özlem Yapıcıer, Mustafa Kemal Demir;* ***References and Fundings:*** *Okan Toygar, Baha Toygar;* ***Materials:*** *Okan Toygar, Özge Yabaş Kızıloğlu, Levent Akçay, Özlem Yapıcıer, Mustafa Kemal Demir, Baha Toygar.*

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