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Giant Solitary Fibrous Tumour of The Orbit Resected via Subconjunctival Approach

Orbital Dev Soliter Fibroz Tümörün Subkonjunktival Yaklaşımla Rezeksiyonu

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ABSTRACT

Solitary fibrous tumours (SFTs) are uncommon neoplasms originating primarily in the pleura but occasionally in the orbit. Despite their generally benign nature, they can exhibit aggressive behaviour. This study focuses on a rare case of a giant SFT located in the orbit, emphasizing diagnostic challenges and treatment considerations. A 69-year-old male with a history of progressive eyelid swelling, proptosis, and vision loss underwent a subconjunctival approach for the removal of an orbital SFT. Histopathological analysis, along with immunohistochemistry markers, confirmed the tumour's identity. Successful tumour resection was achieved through the subconjunctival approach, highlighting the efficacy of this method for orbital SFTs. The histopathological examination revealed typical features of SFT, characterized by spindle cells and unique architectural patterns. Immunohistochemistry further supported the diagnosis. This case underscores the importance of considering orbital SFTs in the differential diagnosis of orbital tumours, particularly when confronted with proptosis and visual disturbances. Surgical excision, guided by radiological imaging and immunohistochemically analysis, remains the primary therapeutic approach. However, due to the potential for aggressive behaviour, further research is needed to optimize management strategies, especially in cases of incomplete resection or atypical behaviour.

Keywords: Solitary fibrous tumour, orbit, subconjunctival approach, proptosis, differential diagnosis

ÖZ

Yalnız fibrous tümörler (YFT), genellikle plevradan kaynaklanan nadir neoplazmlardır, ancak nadir durumlarda orbitada da görülebilirler. Genel olarak benign nitelik taşımasına rağmen agresif davranışlar sergileyebilirler. Bu çalışma, orbitada yer alan nadir bir dev YFT vakasına odaklanarak tanısal zorlukları ve tedavi düşüncelerini vurgulamaktadır. İlerleyici göz kapağı şişliği, proptozis ve görme kaybı öyküsüne sahip 69 yaşındaki erkek hasta, orbitadaki bir YFT'nin çıkarılması için subkonjonktival yaklaşım uygulandı. Histopatolojik analiz ile immünohistokimya işaretleri, tümörün kimliğini doğruladı. Başarılı tümör çıkarılması, subkonjonktival yaklaşımın orbitadaki YFT'ler için etkililiğini göstermektedir. Histopatolojik inceleme, iğsi hücreler ve benzersiz mimari desenlerle karakterize edilen tipik YFT özelliklerini ortaya çıkardı. İmmünohistokimya, tanıyı daha da destekledi.Bu vaka, özellikle proptozis ve görme bozuklukları ile karşılaşıldığında orbital tümörlerin ayırıcı tanısında orbital YFT'leri düşünmenin önemini vurgular. Radyolojik görüntüleme ve immünohistokimyasal analiz rehberliğinde cerrahi çıkarılma, temel tedavi yaklaşımını oluşturur. Bununla birlikte, agresif davranış potansiyeli nedeniyle, eksik çıkarılma veya atipik davranış durumlarında yönetim stratejilerini optimize etmek için daha fazla araştırmaya ihtiyaç vardır.

Anahtar Kelimeler: Soliter fibrous tümör, orbita, subkonjonktival yaklaşım, proptozis, ayırıcı tanı

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INTRODUCTION

Solitary fibrous tumours (SFTs) are rare neoplasms that primarily originate in the pleura but can also occur in the orbit, although their occurrence in this location is uncommon. [1] Typically, patients affected by orbital SFTs present with progressive swelling of the eyelids, proptosis, and visual disturbances [2]. Unlike SFTs occurring in pleural sites, orbital SFTs do not exhibit systemic symptoms such as hypoglycaemia, arthralgia, or pleural or peritoneal effusion. [2] However, it is important to consider the possibility of the meningeal variation of solitary fibrous tumours with orbital involvement in the differential diagnosis of aggressive dural-based lesions. [3]

Although solitary fibrous tumours are generally considered benign neoplasms, they can exhibit aggressive behaviour, including high rates of local recurrence and the potential for late-term metastasis to the bones and lungs. [4] These tumours have been known to extend from the orbit to adjacent structures such as the paranasal sinuses, extradural anterior and middle cranial fossas, cavernous sinus, and other cranial regions through the orbital fissures or foramina. Despite their rarity, it is crucial to recognize the clinical features and potentially aggressive nature of orbital solitary fibrous tumors. Complete surgical excision of the tumour is the primary treatment approach, which can significantly reduce the associated clinical symptoms. However, in cases where orbital involvement of meningeal solitary fibrous tumours is suspected, a careful differential diagnosis is essential due to their potential aggressiveness. [3]

CASE PRESENTATION

A 69-year-old male patient presented with a six-year history of swelling in the right upper and lower eyelids, accompanied by eye extrusion and vision loss. The patient had previously undergone two surgeries due to a right frontonasal tumour that extended into the orbit. **(Figure 1 A and B)** Total vision loss was noted in the right eye. External examination revealed increased periorbital pigmentation and 7 mm of exterior displacement of the right globe. Surgical excision scars from the previous transcranial procedures were observed in the right frontozygomaticoorbital region. A computed tomography (CT) scan demonstrated a solid extra-conal mass measuring 4×3×4 cm, adhering to the lateral-superior wall of the right orbit and the orbital roof. The tumour extended into the left lateral subfrontal space. Based on a provisional diagnosis of a soft tissue tumour, the patient underwent a right subconjunctival approach for tumour resection. **(Figure 1 C and D, Figure 2)**

Under general anaesthesia, the patient was fully prepped and draped, and traction sutures were placed in the center of the tarsal plates of both upper and lower lids. A lid speculum was used to keep the globe exposed and the eyelids opened. A superolateral conjunctival peritomy was performed around the frontozygomatic hemi-limbus, and Tenon's capsule was dissected free from the globe until the tumour mass was reached in the superolateral region. Careful blunt dissection was then performed around the tumour, utilizing the best available tumour tissue plane. The tumour's capsule, which adhered to neighbouring tissues, was reached and a grayish, soft tumour mass was resected and cauterized using bipolar cautery. The tumour was then freed from the lateral orbit and completely resected. A gross examination of the tumour revealed a well-circumscribed and pseudo-encapsulated mass with dimensions ranging from 3x4x4 cm. Upon cut section, the tumour displayed a grayish and fleshy appearance with areas of haemorrhagic foci. Macroscopically, the tumour had a soft-elastic consistency and a grayish surface. The patient was followed up for six months, and a second operation was planned to achieve total resection of the tumour using a sub-conjunctival approach.

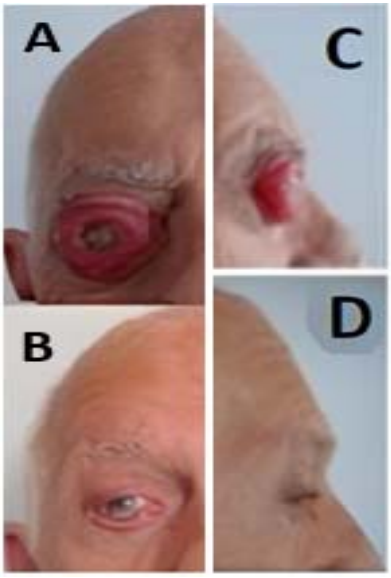

Figure 1: External photograph showing a superior-temporal displacement of the globe with a firm lobulated mass felt along the inferonasal orbit and conjunctiva, with scarring of the cornea.

Histopathological examination of the resected tumour revealed a dense proliferation of spindle cells with variable architectural patterns. There were areas with a fascicled pattern as well as areas with a random and "pattern-less" cell arrangement. The spindle cells showed uniform, oval-shaped nuclei with finely dispersed chromatin.

Immunohistochemistry analysis demonstrated strong and diffuse positivity for CD34, CD99, and bcl-2 in the tumour cells, while S-100 protein and CD45 were negative. **(Figure 3, 4)**

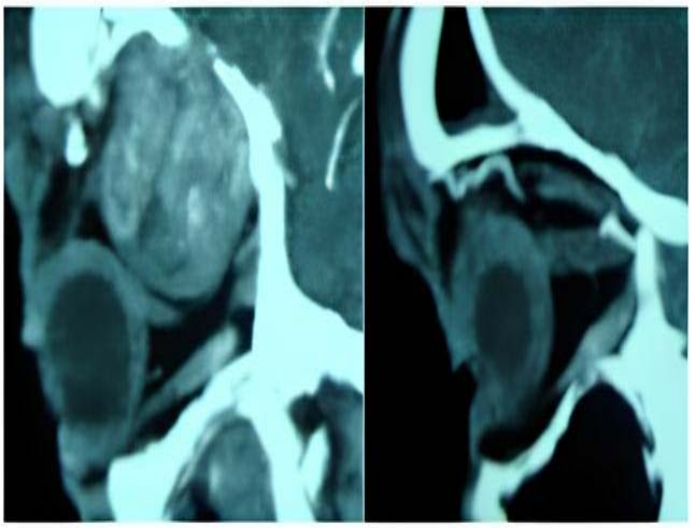

Figure 2: Computerized tomography of the orbit revealed an intensely enhancing well-circumscribed large orbital mass lesion measuring 5 × 4.8 × 4.5 cm and post-operative image.

DISCUSSION

Solitary fibrous tumours (SFTs) primarily originate from the pleura but can also occur in the orbit as rare spindle cell neoplasms. While their occurrence in the pleural region has been extensively studied, orbital SFTs are relatively uncommon, and there have been limited case reports and series documenting their characteristics. [1] Orbital SFTs present progressive swelling of the upper eyelid and proptosis, with accompanying visual disturbances. Although generally benign in nature, SFTs can exhibit aggressive behaviour, necessitating thorough evaluation and appropriate differential diagnosis for aggressive dural-based lesions. [3]

The differential diagnosis of orbital SFTs includes hemangiopericytoma, among other conditions. This section elaborates on the distinguishing features of these conditions, guiding clinicians in accurate diagnosis. The similarities and differences between these entities are crucial for proper therapeutic planning.

The age range of patients with orbital SFTs typically spans from 33 to 75 years. Surgical resection is the primary treatment modality, with various approaches employed depending on the tumour location. Complete excision of the tumour is associated with favorable outcomes, while incomplete resection is a significant risk factor for tumour recurrence. The role of postoperative radiotherapy in managing atypical or incompletely resected central nervous system SFTs is still under investigation and requires further study. [5]

Radiological imaging techniques, such as computed tomography (CT) and magnetic resonance imaging (MRI), play a crucial role in diagnosing and evaluating orbital SFTs. These tumours often display marked enhancement on postcontrast CT and MR images. Dual-phase CT scans may reveal rapid enhancement followed by early washout of contrast material. Furthermore, cerebral angiography can provide insight into the rich vascular supply feeding the tumour through branches of both the external and internal carotid arteries. [6]

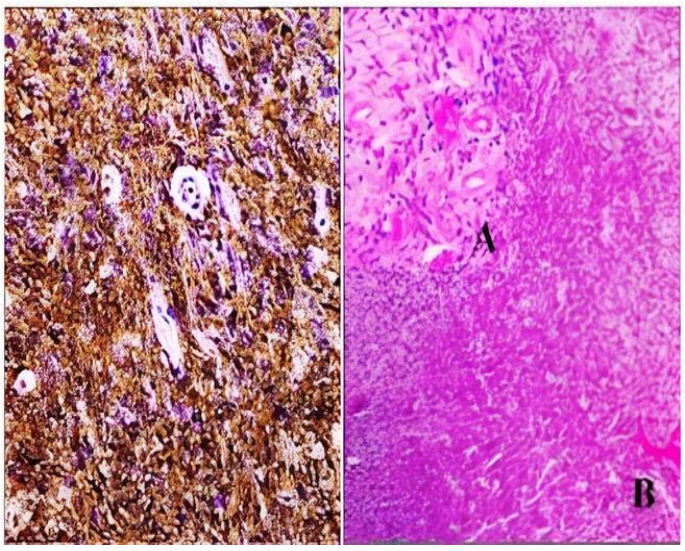

Figure 3: Microscopic aspect of a solitary fibrous tumor: Plump spindle cells with round or oval vesicular nuclei and prominent vascularity.

Histologically, orbital SFTs exhibit high cellularity and significant pleomorphism. Immunohistochemically analysis is essential for confirming the diagnosis and differentiating SFTs from other neoplasms. CD34 and vimentin are commonly employed markers, displaying diffuse positive staining in SFTs. Conversely, SFTs typically exhibit negative staining for cytokeratin, epithelial membrane antigen, glial fibrillary acidic protein, S-100 protein, and factor XIII. [7] While histologic appearance can vary, complete resectability remains the most critical prognostic factor for both orbital and pleural SFTs. [9]

Clinical awareness of orbital SFTs as a potential cause of unilateral proptosis is essential. These tumours can present with both benign and malignant behaviour. Malignant forms

may invade local structures, demonstrate recurrent growth, or metastasize, while benign lesions can often be effectively cured through surgical excision. Therefore, including orbital SFTs in the differential diagnosis of unilateral proptosis is paramount, along with the utilization of immunohistochemically markers to ensure accurate diagnosis. [8]

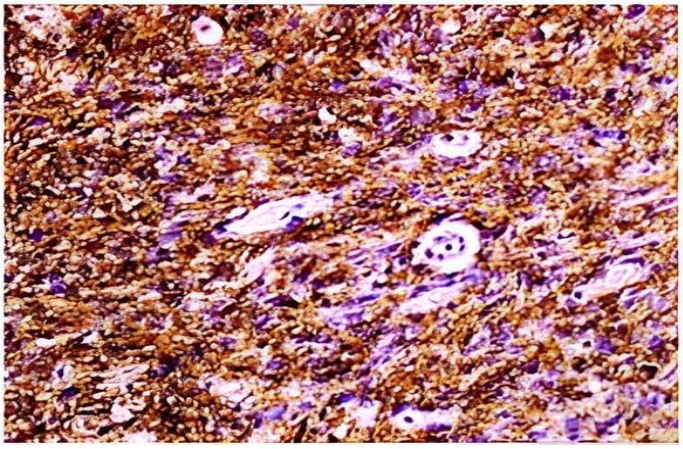

Figure 4: Immunohistochemically exam, showing strong positivity of tumour cells to CD34.

This case underscores the importance of considering a broad age range for patients with orbital SFTs. While the typical age range is 33-75 years, cases outside this spectrum are not uncommon. The role of advanced imaging techniques like MRI and comprehensive immunohistochemically analysis is crucial in the diagnosis and treatment planning of orbital SFTs. The study also discusses the potential for aggressive behaviour and recurrence, emphasizing the need for thorough surgical resection and close follow-up.

Orbital SFTs, while rare, should be considered in the differential diagnosis of orbital tumours across a wide age range. This case highlights the importance of modern imaging techniques and extensive immunohistochemically profiling in the management of these tumours. Further research is encouraged to deepen understanding and improve treatment strategies, particularly in cases of incomplete resection or atypical behaviour.

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