An Uncommon Case of Orbital Myofibroma in Adult: case report

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ABSTRACT

Introduction: Myofibroma was initially described by Williams Schrum in 1951 as Congenital Fibrosarcoma. This is an uncommon benign tumor of mesenchymal cells exhibiting myofibroblastic differentiation that occurs mainly in the first decade of life. Case Presentation: A 44-year-old male patient presented to the neurosurgery clinics with a history of diplopia and bulging of his left eyeball, his visual acuity was normal, and no other abnormalities were found on ophthalmological examination. The Magnetic Resonance Imaging of the head was performed with intravenous contrast. It showed a well-defined lesion measuring 3.3 × 2.4 cm in T2-weighted sequence in the left orbital and intracranial region involving the left eyeball and extending to the ethmoid sinus, frontal bone, medial rectus muscle and optic nerve. Subsequently, the patient underwent a left fronto-orbital craniotomy for tumor resection. The microscopy and immunohistochemistry study are most consistent with the diagnosis of an orbital myofibroma/myopericytoma. Discussion and Conclusion: We reported a rare occurrence of adult myofibroma in the orbital region. The patient had excellent postoperative recovery and is under ambulatory follow-up.

Keywords: Myofibroma; Myofibromatosis; Eye; Neoplasms; Adult

RESUMO

Introdução: O miofibroma foi descrito inicialmente por Williams Schrum em 1951 como fibrossarcoma congênito. É um tumor benigno incomum de células mesenquimais exibindo diferenciação miofibroblástica que ocorre principalmente na primeira década de vida. Relato do caso: Paciente do sexo masculino, 44 anos, apresentado ao ambulatório de neurocirurgia com histórico de diplopia e abaulamento do globo ocular esquerdo, acuidade visual normal e nenhuma outra anormalidade foi encontrada no exame oftalmológico. A ressonância magnética da cabeça foi realizada com contraste intravenoso, apresentando lesão bem definida medindo 3,3 × 2,4 cm em sequência ponderada em T2 na região orbitária esquerda e intracraniana envolvendo o globo ocular esquerdo e estendendo-se até o seio etmoidal, osso frontal, músculo reto medial e nervo óptico. Posteriormente, o paciente foi submetido à craniotomia fronto-orbital esquerda para ressecção tumoral. Foi realizada uma orbitotomia da borda orbitária superior esquerda para a excisão tumoral. O estudo de microscopia e imuno-histoquímica é mais consistente com o diagnóstico de miofibroma/miopericitoma orbital. Discussão e Conclusão: Relatamos uma ocorrência rara de miofibroma adulto na região orbital. O paciente apresentou excelente recuperação pós-operatória e está em acompanhamento ambulatorial.

Palavras-chave: Miofibroma; Miofibromatose; Olho; Neoplasia; Adulto

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INTRODUCTION

Myofibroma was initially described by Williams Schrum in 1951 as Congenital Fibrosarcoma. This is an uncommon benign tumor of mesenchymal cells exhibiting myofibroblastic differentiation that occurs mainly in the first decade of life. It can be classified as solitary, multicentric or generalized. However, myofibroma in adulthood is usually solitary and superficial. Though the most common site of occurrence is the head and neck region, myofibroma of the orbit is a very rare site of involvement and its solitary occurrence in adult patients is even more rare. In our report we presented an adult patient who had a solitary myofibroma with cranial and orbital involvement. The presentation of this disorder as shown here has few cases described in the literature.

CASE PRESENTATION

A 44-year-old male patient presented to the neurosurgery clinics with a history of diplopia and bulging of his left eyeball (proptosis), his visual acuity was normal, and no other abnormalities were found on ophthalmological examination. There was no significant family or past medical history.

The Magnetic Resonance Imaging (MRI) of the head was performed with intravenous contrast. It showed a well-defined lesion measuring 3.3 × 2.4 cm in T2-weighted sequence in the left orbital and intracranial region involving the left eyeball and extending to the ethmoid sinus, frontal bone, medial rectus muscle and optic nerve (Figure 1).

Subsequently, the patient underwent a left fronto-orbital craniotomy for tumor resection. A left superior orbital rim orbitotomy for the tumor excision was performed. No significant postoperative complication was observed (Figure 2). Mild headache and enophthalmos were seen on postoperative follow-up.

On immunohistochemistry, the spindle cells were positive for smooth muscle Actin and Ki 67, however, it was negative for CD 34, Desmin, PGR 636, Cytokeratin 18, and E29.

Microscopy study showed a mesenchymal neoplasm with proliferation of spindle cells containing slightly irregular nuclei and ill-defined eosinophilic cytoplasm. There were areas of calcifications and fibroblastic star-shaped cells. There were no areas of necrosis and rarely mitotic activity was identified (Figures 3-6).

The microscopy and immunohistochemistry study are most consistent with the diagnosis of an orbital myofibroma/myopericytoma.

DISCUSSION

According to the World Health Organization (WHO) classification of tumors of soft tissue and bone, the myofibroma was classified...
as a perivascular tumor. Myofibroma of the ocular region (which includes the orbits and the eyelids) is very rare in the literature with majority of cases reported occurring in children. There are few cases reported in the literature of orbital myofibroma in adults with comprehensive immunohistochemical and microscopy study.

Immunohistochemistry is extremely important in diagnosis of a myofibroma. It generally shows positivity for actin and vimentin, but desmin, EMA, S100 and CK, are usually negative.

Myofibroma can be localized or generalized self-limiting benign processes represented by cells with characteristics of myofibroblasts and sometimes pericytes. Furthermore, myofibroma has other characteristics as plump spindled cells in a moderately cellular distribution dispersed amidst prominent intratumoral vascular channels. Although some mitoses can be seen, features of malignancy such as increased mitotic activity, atypical mitotic figures, necrosis, vascular invasion, or locally infiltrative growth are not identified. In some cases, it can be observed numerous mitoses as well as areas of necrosis. The differential diagnosis of myofibroma include leiomyoma, neurofibroma, nodular fasciitis, hemangiopericytoma, and fibrosarcoma due to their histopathologic similarities.

Soft tissue lesions are difficult to distinguish from other mesenchymal tumors and the findings of radiologic imaging can differ depending on the location of the lesion. Nevertheless, MRI and CT-scans represent imaging modalities that has proven useful in the diagnosis of myofibroma, because it typically shows non-homogeneous well-circumscribed masses, with moderate vascularity. Contrast-enhanced scans show variability in the enhancement pattern. On T1-weighted MRI, myofibromas appear as moderately low to low intensity images. On T2-weighted MRI, the lesions tend to appear as high intensity lesions. Ultrasonography, when performed, may yield a well-circumscribed lesion, which exhibits low internal reflectivity and

Figure 2. Postoperative CT-scan.

Figure 3. Classic zonation pattern showing a hypercellular area (left) and a myxoid, edematous, less cellular area (right). Vessels are prominent, sometimes dilated and often in a hemangiopericytomatos (staghorn) pattern (4x magnification).
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Case Report

Myofibromas can be homogenous or heterogeneous, with a heterogeneous internal structure. Additionally, myofibromas with increased vascularity enhance with gadolinium. Even with sophisticated imaging techniques, differential diagnosis can still be difficult hence histopathology is ultimately necessary to make a definitive diagnosis and to rule out a more aggressive neoplasm.

Albeit it’s more common in infants and childhood, it is important to remember myofibroma occurrence when a spindle cell lesion with a nodule, fascicular or whirling growth patterns and areas of hyalinization, which are admixed with blood vessels, is seen. Complete resection of the solitary orbital myofibroma is the preferred treatment modality. The recurrence rates for myofibromas are quite low, ranging from 0% to 12.5%. Recurrences can be attributed to tumors with difficult surgical access or incomplete removal.

CONCLUSION

A rare occurrence of adult myofibroma in the orbital region was reported. Surgical excision was the treatment of choice, once its recurrence is known to be uncommon after complete resection. The patient had excellent postoperative recovery and is under ambulatory follow-up.

REFERENCES

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