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A rare orbital tumor: benign isolated orbital schwannoma

Nadir bir orbita tümörü: benign izole orbital schwannom

Özet

Schwannomlar sinir kılıfından köken alan, orbitada nadir görülen tümörlerdir. Orbitada lokalizasyonuna ve orijin aldığı sinire göre pek çok klinik prezentasyonla karşımıza çıkabilir. Bu nedenle orbita hastalıklarında ayırıcı tanı açısından önem arz eder. Bu çalışmada, izole orbital schwannom olgusu literatür ışığında tartışıldı. 41 yaşındaki bayan hasta sol alt orbita kenarına lokalize ağrısız kitle ile başvurdu. Kitle dışında göz muayenesi normaldi. Bilgisayarlı orbita tomografisinde 15X10 mm boyutlarında orbita inferolateralinde ekstrakonal yerleşimli iyi sınırlı solit yumuşak doku lezyonu izlendi. Total olarak çıkarılan kitlenin histopatolojik incelemesinde tanı benign schwannom olarak rapor edildi. Hastanın 1 yıllık takibinde herhangi bir komplikasyonla karşılaşılmadı.

Anahtar kelimeler: Orbital schwannom, infraorbital sinir, ayırıcı tanı, görüntüleme

Abstract

Schwannomas, arising from the nerve sheath, are rare tumors of the orbit. They may present with various clinical presentations depending on localization of tumors in the orbit and the nerve from which they arise. Therefore, they should be considered as differential diagnosis of orbital disease. In this study, the case of isolated orbital schwannoma is discussed in the light of the literature. A 41 year old female patient was referred to the hospital with a painless mass localized near the inferior part of the left orbit. Eye examination of the patient was normal except for the mass. Orbital computerized tomography showed a well-bordered, solid soft- tissue lesion measuring 15x10 mm in size, located in the extraconal space of the inferolateral orbit. As a result of the histopatholgical examination of the totally excised mass, the diagnosis was established as benign shwannoma. No complications were observed during the one-year follow-up of the patient.

Key Words: Orbital schwannoma, infraorbital nerve, differential diagnosis, imaging

* Özlem Yalçın Tök
Levent Tök
** Nisa Ünlü
*** Nilgün Kapucuoğlu
* Yavuz Bardak

* Süleyman Demirel Üniversitesi, Tıp Fakültesi, Göz Hastalıkları AD, Isparta, Türkiye
** İntegra Görüntüleme Merkezi, Ankara, Türkiye
*** Süleyman Demirel Üniversitesi, Tıp Fakültesi, Patoloji AD, Isparta, Türkiye

Yazışma Adresi:
Özlem Yalçın Tök
Turan Mah. Mehmet Bilginer Sitesi
C Blok No: 9 Gökçay/Isparta -Turkey
Tel: 90 505 4862260
e-mail: esattok@yahoo.com

TOD 44. Ulusal Oftalmoloji Kongresinde poster sunumu yapılmıştır.

Introduction

Schwannomas, also known as neurilemmomas, are peripheral nerve tumors, arising from Schwann cells. These tumors are typically slow-growing and painless (1). They constitute 1-8% of head-neck tumors and 1-6% of orbital tumors and are usually unilateral (2). These tumors are most commonly observed in the second or fourth decades of life and are benign lesions, the borders of which are surrounded by well-defined capsules in general (1,3,4). The differential diagnosis of orbital tumors is of great importance. Diagnosis is mainly established by histopathological examination (1,2).
In the present study, we discuss clinical findings and treatment of a case of schwannoma located in the inferior orbit.

Case Report

A 41 year old female patient was referred to the hospital with a 4-month history of painless mass lesion located in the lower margin of the left orbit (Figure 1). She had no complaint of double-vision and no history of trauma. The lesion, which was solid, painless, partially mobile and which did not cause proptosis or a marked displacement of the eye, was palpated. There were no changes in Valsalva maneuver, eye movements, posture and the size of the lesion. No pulsation was felt and no transillumination was observed in the lesion. In the eye examination, the visual acuity was 20/20 in the left and right eyes. The anterior and posterior segment examination was normal. Pupillary reflex and eye movements were also normal. The physical examination revealed no palpable skin lesion, cafe au lait spots or lesions such as axillary or inguinal speckling, suggestive of neurofibromatosis. Orbital computerized tomography (CT) showed a well-circumscribed, solid soft- tissue lesion measuring 15x10 mm in size, located in the extraconal space of the inferolateral orbit (Figure 2).

Figure 1: Preoperative image showing swelling of the lesion near the orbit and the lower eyelid.

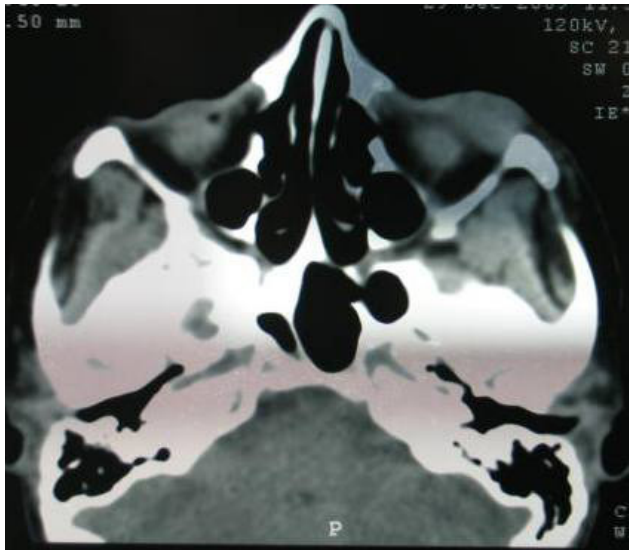

Figure 2: CT scans of a schwannoma demonstrate the well-defined extraconal tumor.

Inferior orbitotomy was performed with subsiliar insicion with the patient under anesthesia. The mass was excised totally (Figure 3). The histopathological examination of the mass revealed hypocellular (Antoni B) and hypercellular (Antoni A) areas, which consisted of spindle tumor cells with fusiform nucleus and marked nucleolus whose cytoplasms were not clearly defined.
Additionally, Verocay bodies with spindle cells displaying peripheral palisading were also noted (Figure 4). Histopathological diagnosis was reported as shwannoma. During one year follow-up of the patient, no relapse or complications were observed (Figure 5).

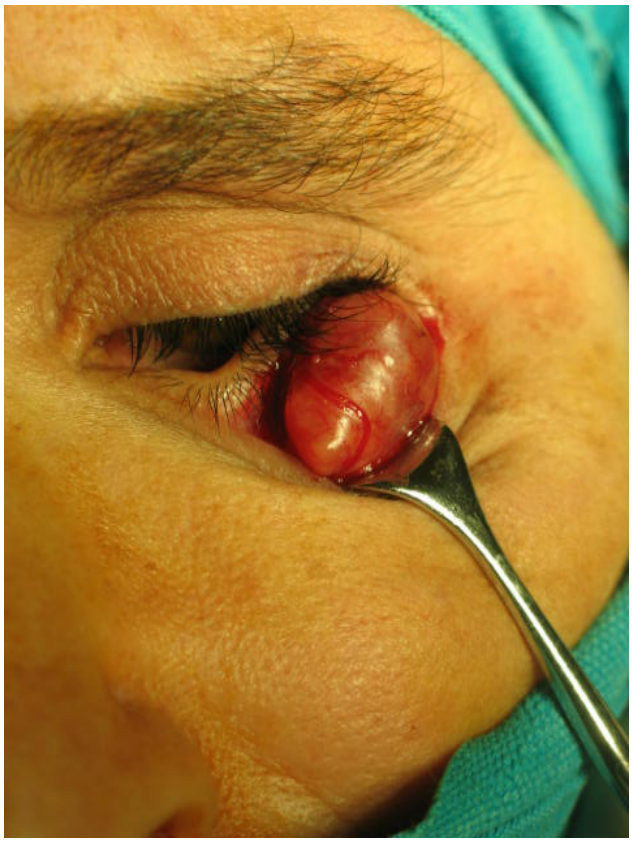

Figure 3: Intraoperative view of the mass.

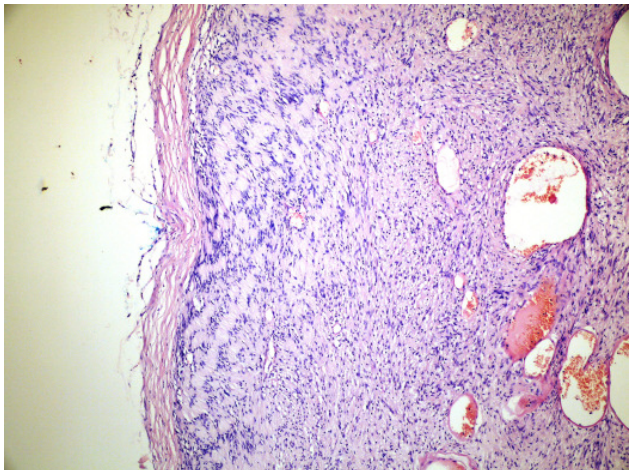

Figure 4: Verocay bodies, dilated vessels within well circumscribed lesion with fibrous capsule (x100, hematoxylin eosine).

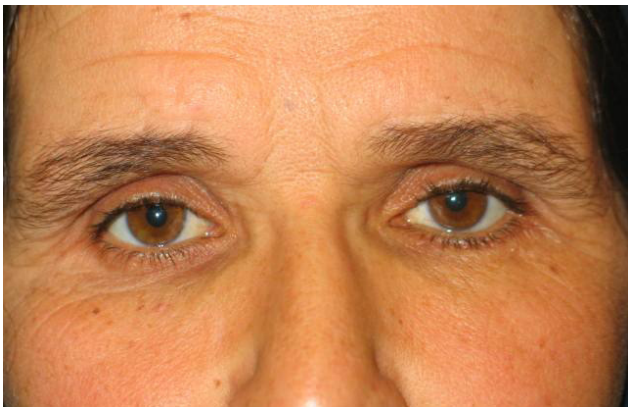

Figure 5: The view of the patient at postoperative 9 months.

Discussion

Schwannomas are rare tumors of neurogenic origin in the orbit (1). Usually they arise from sensory nerves, whereas those originating from motor nerves have also been reported (1,4,5). However, the nerve from which the tumor arises may not be identified in 50% of cases (2,5). They, in general, present with painless proptosis or displacement of the globe. Most shwannomas arise from branches of either supraorbital and supratrochlear nerve, thus producing a downward displacement of the globe. However, complaints on admission may vary depending on the localization of the tumor (1,2). As in this case, the tumor may rarely originate from the infraorbital nerve and produce an upward displacement of the eye. The tumor may reach a large size or originate from cranial nerves 3, 4, and 6, thus causing limitation of eye movements, strabismus and diplopia (2). In particular, lesions near the orbital apex may lead to decreased vision, narrowed field of vision and relative afferent pupillary defect. They may also cause atrophy of the optic disk, edema or retinal and choroidal folds. Clinical findings can be confused with retrobulbar neuritis and papillitis (2,5). Schwannomas may also affect one of the branches of the trigeminal nerve, causing pain and numbness and may be confused with the sinusitis. Numbness in the sensory field of the affected nerve can be a complaint on admission, or may be a complication due to nerve damage during surgery (6-8).

Benign schwannomas are usually sporadic and

solitary (1,2). However, several heritable diseases such as neurofibromatosis 2, schwannomatosis and Carney complex are associated with the development of multiple schwannomas. In 2-18% of cases, Von Recklinghausen's Neurofibromatosis can also be observed (2). CT and magnetic resonance (MR) imaging are of great assistance in the determination of the spread and localization of the tumor as well as its histopathologic nature (3,6). On CT, schwannomas appear as ovoid, smooth, solitary, orbital retrobulbar masses, most frequently in the superior orbit with the long axis in the same direction as the nerve, which is generally the anteroposterior direction. The tumor mass is often isodense or slightly hyperdense when compared to the brain, and following injection of intravenous contrast medium, it usually exhibits homogeneous or heterogenous moderate to marked contrast enhancement. Bony erosion may also occur and is more common in tumors that have some extraconal extent (3,6,8). MR imaging shows the tumor as an ovoid, well circumscribed mass, located in the extraconal or intraconal space. It is isointense in comparison to the extraocular muscle and cerebral gray matter on T1-weighted images and is hyperintense on T2-weighted images (3,6). In Antoni B areas, greater signal intensity is obtained on T2-weighted images when compared to the more cellular Antoni A portion (6). There may also be evidence of cavitary change on T2-weighted images, indicating a predominance of Antoni B pattern from a histopathological point of view. With gadolinium infusion, there is increased enhancement in myxoid Antoni B type areas as compared to the more cellular Antoni A type areas (6).

Surgery is the treatment of choice for schwannomas. Tumors that can not be excised because of their progressive growth, may lead to complications. Therefore, they should be excised totally. Incomplete excision may result in recurrence and intracranial spread. In cases with accompanying neurofibromatosis, there is the risk of recurrence and malignant transformation in aggressive lesions with myxoid matrix, hypercellularity and increased mitotic activity. They exhibit the same histopathologic features as schwannomas in other areas. Antoni A, Antoni B and Verocay bodies are characteristic histopathologic features of this tumor. Antoni type A areas are composed of well-differentiated spindle cells with ovoid nuclei and fine stippling of chromatin. These cells show a fascicular arrangement with a background of fibrillar cell processes. The nuclei palisade to create picket-fence like structures that have interdigitating cytoplasmic processes, thus creating a pattern known as Verocay bodies. In Antoni type B areas, bipolar and multipolar cells are embedded in a loose myxoid matrix. They may undergo secondary lipidization, which adds a yellow hue to the gross coloration. The consistency of the tumor mass depends on which one of these histologic patterns predominates, being more soft and cystic in tumors exhibiting predominance of Antoni B pattern. These tumors, which are moderately vascular, contain capillaries with thickened basement membranes. Degenerative changes such as hemorrhage, cyst formation and calcification may occur in long standing tumors that are immunohistochemically stained positive for S-100, vimentin and neuron-specific enolase (1,2,4,5,8).

Benign orbital schwannomas are rare. Because of their ability to mimic many clinical conditions depending on their localization or the nerve from which they arise, bening orbital schwannomas should be kept in mind in the differential diagnosis of orbital diseases .

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