Infraorbital schwannoma. Case report

ABSTRACT

Background: Infraorbital schwannoma is a benign tumor of the peripheral nerve sheath composed of Schwann cells. It usually occurs between 20 and 70 years of age. Tumors are usually asymptomatic and can cause progressive, painless proptosis during growth.

Clinical case: A 32-year-old male was admitted to the ophthalmologic hospital with a painless, slowly progressive mass above the left lacrimal sac. During ophthalmologic examination, a tumor with a rubbery consistency and firmly attached to the surrounding structures was observed. Transillumination was negative. B-mode ultrasound disclosed a phakic eye as well as an infraorbital well-circumscribed homogeneous mass that was 19.7 mm at its largest diameter and without involvement of the lacrimal pathway. A-mode ultrasound showed medium-high reflectivity with small internal vascularity. Computed tomography showed a homogeneous well-circumscribed solid mass anterior and inferior to the left globe without bony erosion. The mass was excised using a subdermal incision. Histopathological diagnosis was infraorbital schwannoma.

Conclusions: Schwannoma is a rare benign orbital tumor. Few cases have been reported. Definitive diagnosis is made by histopathological findings with the presence of a true capsule, hyper- and hypocellular areas, thickening and hyalinization of the vessel walls. These findings should not be confused with other benign fusocellular tumors. We report a new case of infraorbital schwannoma and compared it with previously reported cases.

Key words: Schwannoma, orbital tumor, infraorbital nerve.

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Received: 4-10-2013
Accepted: 9-12-2013
INTRODUCTION

Schwannoma or neurilemoma is a well-differentiated and generally solitary benign tumor characterized by proliferation of Schwann cells from a peripheral nerve.\(^1\)\(^-\)\(^3\) Schwannomas comprise 1-8% of all tumors of the head and neck and 1-4% of orbital tumors. They usually appear between the second and fourth decades of life.\(^3\)\(^,\)\(^4\) Orbital schwannomas originate from the sensory nerves and, more frequently, from the supraorbital and supratrochlear nerves. They are solitary tumors with well-defined margins. The primary symptoms are retroocular sensation of pressure, proptosis and pain.\(^4\)\(^-\)\(^7\)

In the medical literature, only eight cases of schwannomas originating from the infraorbital nerve were found; two of them corresponded to the malignant variety.\(^5\)\(^-\)\(^7\)\(^,\)\(^13\) We report a new case of an infraorbital schwannoma and compared it with those previously reported.

CLINICAL CASE

We report the case of a 32-year-old male patient who presented to the hospital because of a non-painful growth of 4 years duration of the lacrimal sac. Ophthalmological examination found a well-circumscribed rubbery tumor in the left inferior nasal orbital rim, \(-19 \times 11\) mm, which was adhered to deep tissues and did not transilluminate (Figure 1). Eye movements were preserved. The differential clinical diagnoses considered were tumor of the lacrimal sac vs. dermoid cyst vs. chronic dacrocystitis.

B-mode ultrasound revealed a phakic ocular globe with an anterior infraorbital, homogeneous well-circumscribed tumor with its greatest diameter of 19.7 mm and apparently did not involve the lacrimal sac (data not shown). A-mode ultrasound revealed a medium-high reflectivity with poor internal vascularity.

Computed tomography (CT) of both orbits showed a well-circumscribed, homogeneous anterior tumor on the left orbital floor. It did not produce a scleral indentation and showed discrete displacement of the medial wall without affecting the lacrimal sac (Figure 2). An incisional biopsy of the tumor was carried out via a subdermal approach, freeing it from all planes and all adjacent attachments. Gross examination showed an oval-shaped well-circumscribed tumor, apparently encapsulated, with a homogeneous cut surface, yellow-gray in color and of medium consistency.
Microscopically, the tumor was composed of alternating hypercellular areas (Antoni A) and hypocellular areas (Antoni B) (Figures 3A and 3B). The former had spindle cells with indistinct cytoplasmic borders, with nuclear palisading reminiscent of Verocay bodies. Between the hyper- and hypocellular areas there were small blood vessels, some with thickening and hyalinization of their adventitia as well as partial obliteration of their lumen, most apparent with Masson’s trichrome stain (Figure 3C). At the periphery, the tumor was surrounded by a septum of thick connective tissue corresponding to a true capsule (Figure 3D). The histopathological diagnosis was benign schwannoma (neurilemoma).

**DISCUSSION**

Orbital schwannomas are rare tumors, which generally appear in young adults. Orbital schwannomas are associated with neurofibromatosis in 2-18% of the cases. Orbital schwannomas rarely originate from the infraorbital nerve. Currently, only eight cases have been reported. This case involved a 32-year-old male without any systemic disease or any disorder in another location.

The oculomotor, ciliar, lacrimal and temporal zygomatic nerves have been reported as the most common sites of origin in this location. The nerve of origin is not identified during surgery in 50% of the cases. In these cases the nerves from which the tumors originate may be identified according to the pre- or postoperative neural deficit. In our patient, the tumor was painless in the preoperative period and postoperatively had a discrete hypoesthesia of the left cheek, which confirmed the probable infraorbital origin of this tumor.

As the majority of these tumors originate from sensory nerves, they do not interfere with eye movements or vision unless they are located in the orbital apex or compress the optic nerve. However, although the most frequent symptoms of the tumors are pain and ocular displacement, in our patient the tumor was located anterior and inferonasal and did not displace the ocular globe.

In our patient, tomographic findings were very similar to those reported in the medical literature, presenting as a homogeneous infraorbital tumor of well-defined margins, isodense to the cerebral parenchyma without apparent cyst formation.

Magnetic resonance is another imaging study of great assistance in determining the anatomic location such as the internal consistency of this tumor. With this test, cystic changes or heterogeneous intensity of the T2 signal can be shown, which correlates with the histological pattern of Antoni B areas which, in turn, translates the

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**Figure 3.** (A) Hypercellular areas or Antoni A (H-E, x40). (B) Hypocellular areas or Antoni B (H-E, x40). (C) Blood vessels with thickening and hyalinization of its adventitia. (Masson’s trichrome x40). (D) Periphery of the neoplasm surrounded by a true capsule (Masson’s trichrome x10).
presence of friable or cystic areas that predispose to incomplete resections of the tumor and, as a result, development of recurrences.\(^3\)

Complete surgical resection of the tumor is the treatment of choice because it prevents recurrences. Our patient remained asymptomatic and without recurrence 4 months postoperatively.

Infraorbital schwannomas have the same histopathological characteristics of other head and neck schwannomas. They are generally encapsulated tumors composed of two areas: Antoni A or hypercellular where the Verocay bodies and Antoni B or hypocellular bodies can coexist, which can also have cystic areas. Other microscopic findings in these types of tumors are blood vessels with thickening and hyalinization of their adventitia and a true capsule.

Its main differential diagnosis should be with other tumors of the peripheral nerve sheath such as neurofibromas whose separation is predominantly based on the architectural and cytological findings. Immunohistochemical markers are of little use because both tumors express the S-100 protein in neoplastic cells (Schwann cells).\(^1,2,19\)

In conclusion, infraorbital schwannomas are rare tumors in this location that should be taken into consideration in the differential diagnosis of well-delineated, progressively slow-growing tumors in the orbital floor. The treatment of choice is complete surgical resection to avoid tumor recurrence and histopathological evaluation is mandatory in order to establish a definitive diagnosis.

REFERENCES