Clinical and histopathological features of orbital granular cell tumor: case report

Características clínicas e histopatológicas de tumor de células granulares da órbita: relato de caso

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ABSTRACT

A 53 year-old woman presented with a slowly progressive, painless proptosis OS. Computed tomography disclosed a round, homogeneous, well-delimited lesion in the inferior-temporal orbit. The tumor was composed of round cells with eosinophilic granular cytoplasm. Some of the cells had larger eosinophilic granules surrounded by a clear halo; known as pustulo-ovoid bodies of Milian or Bangle bodies. The diagnosis of a granular cell tumor was then established and confirmed by immunohistochemistry. Granular cell tumors are uncommon benign soft tissue neoplasms that have a predilection for the head and neck region. Awareness of the typical histopathological features is crucial for the correct diagnosis.

Keywords: Orbital neoplasms/diagnosis; Exophthalmos; Tomography, X-ray computed; Case report

RESUMO

Mulher de 53 anos apresentou proptose lentamente progressiva no olho esquerdo. Tomografia computadorizada mostrou uma lesão na região temporal inferior da órbita esquerda, bem delimitada, arredondada, homogênea. O tumor era composto de células com citoplasma granular eosinofílico. Algumas das células possuíam grandes grânulos eosinofílicos circundados por um halo claro, conhecidos como corpos ovoides-pustulares de Milian or corpos de Bangle. O diagnóstico de tumor de células granulares foi estabelecido, confirmado pela imuno-histoquímica. Tumor de células granulares são neoplasias incomuns com predileção da região da cabeça e pescoço. O conhecimento das características histopatológicas típicas são cruciais para o correto diagnóstico.

Descritores: *Neoplasias orbitárias/diagnóstico; Exoftalmia; Tomografia computadorizada por raios x; Relato de caso*

INTRODUCTION

Granular cell tumors (GCT) are uncommon benign soft tissue neoplasms that have a predilection for the head and neck region. Abrikossoff, in 1926, was the first to histopathologically describe it as a myoblastoma[(1)]. The combined input from immunohistochemistry and electron microscopy has clarified the morphology of this lesion, which is probably derived from a Schwann cell[(2,3)]. GCTs are found in various locations, usually as a small, solitary, benign lesion. GCTs of the eye and ocular adnexae are rare, but have been described in the orbit, periorbital skin and eyelids, extraocular muscles, lacrimal sac, ciliary body, conjunctiva, and caruncle[(4)].

CASE REPORT

A 53 year-old woman presented with a slowly progressive, painless proptosis OS. During clinical examination, an upward displacement of the left globe was seen, although the exam was otherwise unremarkable. Visual acuity was 20/20 in both eyes and intraocular pressure was 16/16 mmHg. Computed tomography disclosed a round, homogeneous, well-delimited lesion in the inferior-temporal orbit, indenting the globe without invading any orbital structure (Figure 1). An excisonal biopsy was performed and the surgery was uneventful.

The tumor was solid and entirely encapsulated. It was composed of round cells with eosinophilic granular cytoplasm. Some of the cells had larger eosinophilic granules surrounded by a clear halo; known as pustulo-ovoid bodies of Milian or Bangle bodies. No mitosis or areas of necrosis were seen. Immunohistochemistry was performed and the tumor was positive for vimentin, S-100, NSE and CD 68 while SMA, actin, desmin, EMA, cytokeratins, chromogranin, and HMB45 were negative. The diagnosis of an orbital granular cell tumor was then established (Figure 2).

DISCUSSION

Although there are no unique clinical or radiological features distinct from other benign orbital tumors, GCT is easily recognized by routine light microscopy. The diastase-resistant, PAS-positive cytoplasmic granularity is typical of GCT. The granules are believed to be lysosomes or a component of the Golgi apparatus. In some cells, the granules aggregate to form the pustulo-ovoid bodies of Milian. A small number of cases can be less differentiated and, in those, immunohistochemistry in a valuable tool. GCTs are usually positive for vimentin, S-100 protein, NSE, CD 57 and CD 68, while negative for SMA, actin, desmin, EMA, cytokeratins, chromogranin and HMB 45.

Some GCTs may present atypical features and are further termed Malignant GCT. The distinction is done on histopathological grounds and the features are: Necrosis, nuclei spindling, vesicular nuclei with large nucleoli, increased mitotic activity (> 2 mitoses/10 HPF at 200x), high nuclear to cytoplasmic ratio and nuclear pleomorphism. The presence of 3 or more of these features correlates with rates of local recurrences and metastasis of 32% and 50%, respectively[(5)].

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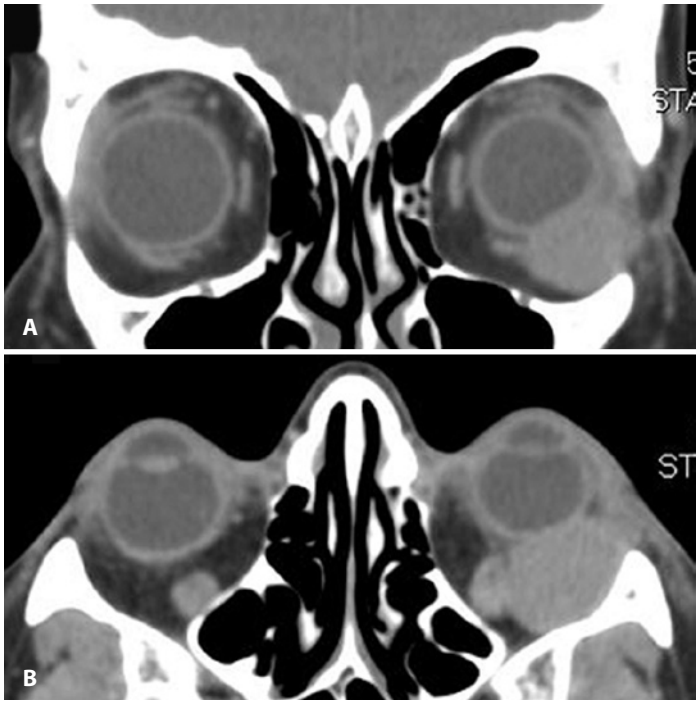

Figure 1. Coronal (A) and axial (B) computed tomography showing a well-defined lesion in the inferior temporal orbit, indenting the globe but without compromising any orbital structure.

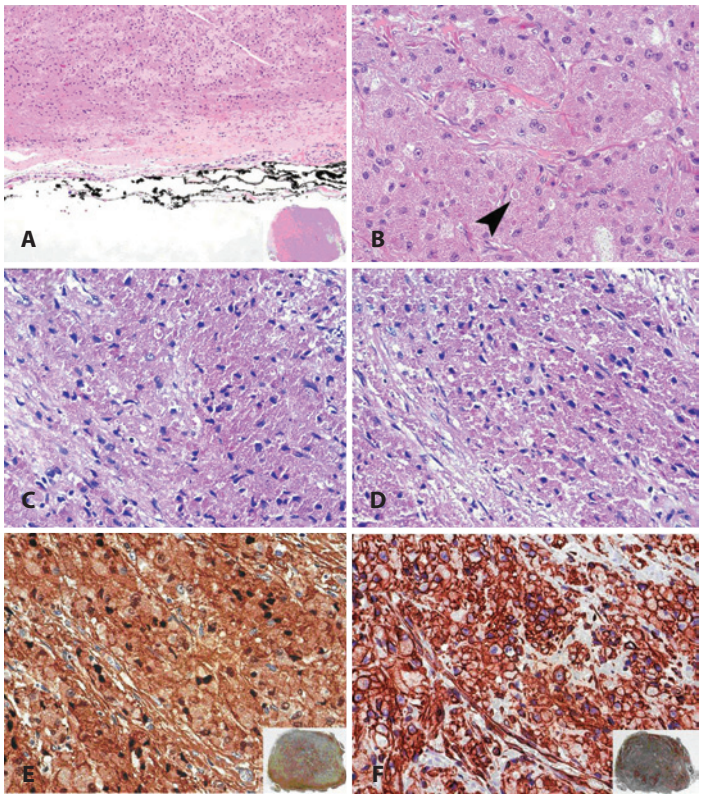

Figure 2. A) The tumor was solid (inset) and encapsulated (H&E, X200). B) The cells had a granular eosinophlic cytoplasm and presented the characteristic pustulo-ovoid bodies of Milian (arrowhead) (H&E, X400). C) The intra-citoplasmatic granules were PAS-positive (PAS, X400). D) The granules remained positive after treatment with diastase (PAS + Diastase, X400). E) Strong and diffuse (inset) immunostaining for S-100 (X400). F) Vimentin was also strong and diffusely (inset) positive (X400).

In summary, we presented histopathological and immunohistochemical findings of a rare orbital tumor. Awareness of the typical histopathological features is crucial for the correct diagnosis. Moreover, the criteria of malignancy must be well know in order to proper counseling and determining the prognosis of each patient.

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