Case Report

Granular Cell Tumor in the Medial Rectus Muscle: A Case Report

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Keywords

Granular cell tumor · Orbit · Medial rectus muscle · Recurrence

Abstract

An 80-year-old female complained of diplopia after undergoing cataract surgery. On the first examination, adduction and abduction were slightly restricted. Magnetic resonance imaging revealed a mass in the medial rectus muscle. The results of pathological examination of a specimen harvested from the mass corresponded to granular cell tumor. Although we could not completely excise the mass because of firm adhesion of the mass to the muscle, there was no recurrence without any suspicious metastatic lesion at 2.5 years of follow-up.

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Introduction

Granular cell tumor is a soft tissue neoplasm of neural origin [1–4]. This develops in the head and neck regions in 70% of cases, and the tongue is the most common site; whereas, the orbital granular cell tumor is rare [1–4]. Here, we report a case of granular cell tumor in the medial rectus muscle.

Case Report

An 80-year-old female complained of diplopia after undergoing cataract surgery at another clinic. She did not have any history of systemic disease or family history.

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On the first examination, her best-corrected visual acuity was 0.8 in the right eye and 0.9 in the left eye. Intraocular pressure was 15 mm Hg in the right eye and 14 mm Hg in the left eye. The Hertel exophthalmometric examination revealed a 2 mm proptosis in the right eye (Fig. 1a). There was no palpable periocular mass. The Hess chart showed mild restriction of adduction and abduction (Fig. 1b). Magnetic resonance imaging (MRI) revealed a well-defined oval mass in the medial rectus muscle in the anterior orbit with iso-intensity to the gray matter on T1-weighted image and heterogeneous hypo-to-high intensity to the gray matter on T2-weighted image (Fig. 1c).

A biopsy of the mass was performed under general anesthesia via transconjunctival approach by one of the authors (Y.T.). As the mass was firmly adhered to the muscle and surrounding orbital soft tissue, it could not be completely excised. Pathological examination revealed tumor cells with oval nuclei and eosinophilic granules in the skeletal muscle (Fig. 1d). Immunohistochemical stainings for S-100 and CD68 were positive (Fig. 1e, f). The findings corresponded to granular cell tumor. The pathological findings did not meet all categories of Fanburg-Smith criteria (spindling, high nuclear to cytoplasmic ratio, vesicular nuclei with large nucleoli, pleomorphism, necrosis, and increased mitotic activity) [5].

At 2.5 years of follow-up, the residual tumor did not enlarge, which was confirmed on repeated orbital computed tomographic images. Exophthalmos and extraocular muscle motility did not deteriorate. Systemic computed tomographic images did not show any suspicious metastatic lesion.

Discussion

We report a case of granular cell tumor in the medial rectus muscle. Granular cell tumor developing in the orbit is rare, and there had been 60 reported cases of orbital granular cell tumor so far [1].

Our case was an 80-year-old female, and the mean age of presentation is found to be 44 years [1]. Overall, granular cell tumors predominantly occur in females [1–3], but there is an equal distribution between sexes in cases with orbital granular cell tumor [1].

Our patient showed restriction of adduction and abduction in accordance with involvement of the medial rectus muscle. In approximately 70% of the cases with orbital granular cell tumor, a lesion directly involves the extraocular muscles [1, 6]. As the tumors have neural origin and develop in association with small-to-medium-sized nerves, predilection for involvement of the extraocular muscles is due to dense neural supply to the muscles [1]. Positive S100 immunohistochemical stain implies the neurogenic nature of granular cell tumor [1]. The most frequently involved muscle is the inferior rectus muscle, followed by the medial and superior recti muscles [1, 6]. Similarly, the most common ocular symptom is diplopia [1, 6]. In regards to these points, our patient was a relatively typical case of orbital granular cell tumor. Preoperative visual acuity was unknown because the patient underwent cataract surgery at another clinic; however, it was possible that the patient noticed diplopia because of improvement of visual acuity after cataract surgery.

The symptoms of granular cell tumors depend on the tumor site, and other possible ocular symptoms are decreased vision, impairment of accommodation, and pain [1]. Granular cell tumors rarely involve the optic nerve and parasympathetic ciliary ganglion [1]. Tumors located in the superior orbit frequently induce pain [1]. In our case, the tumor was located in the medial rectus muscle, which did not cause those symptoms.

In this case, MRI revealed a well-defined oval mass in the medial rectus muscle with iso-intensity to the gray matter on T1-weighted image and heterogeneous hypo-to-high intensity to the gray matter on T2-weighted image. Although granular cell tumors generally show

Fig. 1. Case presentation. **a** A patient's face photo showing mild proptosis in the right eye. **b** The Hess chart showing mild restriction of adduction and abduction in the right eye. **c** A T2-weighted axial magnetic resonance image demonstrating a heterogeneous mass in the right medial rectus muscle. **d** A pathological photo showing tumor cells with oval nuclei and eosinophilic granules, and skeletal muscle fibers (hematoxylin and eosin staining; magnification, $\times 100$). **e, f** Immunohistochemical stainings for S-100 (**e**) and CD68 (**f**) are positive (magnification, $\times 100$).

hypo-intensity on T2-weighted images [1], the tumor in our case largely had the typical MRI findings of granular cell tumor.

Surgical resection is the standard treatment of choice for orbital granular cell tumor [1, 6]. However, even if these tumors are completely removed, diplopia does not improve in most of the cases [1, 5]. On the other hand, when tumors adhere to the surrounding tissue due to inflammatory response around the tumors or are located in the posterior orbit, complete removal of the tumors is difficult [1, 3, 6]. Although such tumors recur, these rarely show an aggressive behavior, including local invasion and metastasis [1, 3]. In our case, we could not completely excise the tumor because of its firm adhesion to the muscle and surrounding orbital soft tissue. However, the recurrence rate of cases undergoing a partial resection is relatively low (10%), and tumors located in the anterior orbit also have less risk of recurrence [1]. Although our case did not show any recurrence or metastasis after an incisional biopsy at 2.5 years of follow-up, we should carefully observe our patient, and if the tumor recurs or metastasizes, additional excision and/or particular radiation therapy such as gamma knife radiosurgery and proton beam radiation therapy are necessary for treatment [1].

In conclusion, we report a rare case of orbital granular cell tumor. Involvement of the medial rectus muscle and the complaint of diplopia in our case were relatively typical clinical characteristics of this tumor. We need to carefully follow-up the patient because of the possibility of recurrence and metastasis of the tumor.

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Statement of Ethics

The authors adhered to the tenets of the 1964 Declaration of Helsinki. We asked the institutional review board of Aichi Medical University Hospital and confirmed that the ethics approval for this report was not necessary on the basis of the ethical guidelines for medical and health research involving human subjects established by the Japanese Ministry of Education, Culture, Sports, Science, and Technology and the Ministry of Health, Labour, and Welfare. Written informed consent for the publication of this report and face photo was obtained from the patient.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All authors qualify for authorship based on contributions to the conception and design (Y.T.), acquisition and analyses of data (Y.T.), interpretation of data (T.Y., A.V., H.K., and Y.T.), drafting the article (T.Y. and Y.T.), revising the article critically for important intellectual content (A.V. and H.K.), final approval of the version to be published (T.Y., A.V., H.K., and Y.T.), and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved (T.Y., A.V., H.K., and Y.T.).

Data Availability Statement

All data are included in this article.

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