Solitary fibrous tumor of the orbit- A case report

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Abstract

Intracranial solitary fibrous tumor (SFT) / Hemangiopericytoma (HPC) is a rare mesenchymal tumor. The occurrence in the orbit is quite rare, with only 2/244 cases in the Japanese population of primary tumors in the orbit. Herein, we report the case of a 54-year-old woman who presented with right visual disturbance. The vision had worsened and was lost after a year. Three years later, magnetic resonance image (MRI) revealed a right intraorbital tumor, and 16 years later, swelling of the right eyelid and exophthalmos appeared. Computed Tomography (CT)scan revealed a well-defined homogeneous mass in the orbito-cranial communicating region, and MRI revealed a 30 mm mass lesion compressing the right optic nerve, presenting with low signal intensity on T1-weighted image (T1WI) and T2-weighted image with heterogeneous contrast effects. Also, dynamic contrast-enhanced T1WI revealed slow signal enhance effect. The lesion was surgically resected via right frontotemporal craniotomy. Histopathological examination of the lesion revealed SFT. Immunopathologically, the tumor was negative for S-100, glial fibrillary acidic protein, epithelial membrane antigen, synaptophysin, while was positive for cluster of differentiation 34, B cell/chronic lymphocytic leukemia lymphoma 2. Moreover, signal transducer and activator of transcription 6 was positive in intranuclear, and Ki-67 was negative in most of the cells. The postoperative course is uneventful without recurrence after 4 years. SFT of the orbit should be considered as a differential diagnosis although it is extremely rare disease. It is similar to meningioma and schwannoma and we should take Dynamic-Contrast-Enhanced MRI for preoperative diagnosis. Careful postoperative follow-up is needed because some cases report recurrence and malignant transformation.

Introduction

Intracranial solitary fibrous tumor (SFT) / Hemangiopericytoma (HPC) is a rare mesenchymal tumor, which is commonly found in posterior cranial fossa, falx cerebri, convexity, and spinal dura mater. The occurrence in the orbit is quite rare[1,2], with only 2/244 cases in the Japanese population of primary tumors in the orbit[3]. Most of them are tumors with a slow-growing tendency and lack of clinical symptoms, and are identified when they present with local pressure symptoms and intracranial hypertension[4]. In this report, we describe a case of SFT / HPC that developed in the orbit and slowly enlarged over a long period of time.

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Case Report

The patient is a 54-year-old woman with past surgical histories of cervical cancer and gastric cancer. She visited ophthalmologist at our hospital with a chief complaint of decreased vision in the right eye and was treated as a retrobulbar neuritis. The vision had worsened and was lost after a year, while no abnormal findings were observed on magnetic resonance imaging (MRI). The patient was referred to our department for visual impairment of unknown cause, and the plan was to monitor her condition as an outpatient at the neurosurgery department. Three years later, MRI revealed a right intraorbital tumor, but MRI follow-up was performed as the patient had already lost her sight. The tumor continued to grow gradually, and 16 years later, swelling of the right eyelid and exophthalmos appeared. Computed Tomography (CT) scan revealed a well-defined homogeneous mass in the orbito-cranial communicating region (OCCR) (Fig. 1).

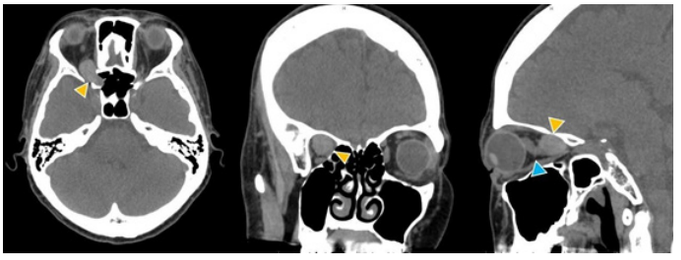

Figure 1: Plain CT revealed a uniform, mildly hyperdense lesion (yellow arrows) extending from the right orbit to the optic canal, compressing the optic nerve (blue arrow) downward and forward.

MRI revealed a 30 x 12 x 12 mm mass lesion in the same area compressing the right optic nerve, presenting with low signal intensity on T1-weighted image (T1WI) and T2-weighted image (T2WI) with heterogeneous contrast effects. Also, dynamic contrast-enhanced T1WI revealed slow signal enhance effect. Then, surgical resection was planned with the provisional diagnosis of optic sheath meningioma (Fig. 2).

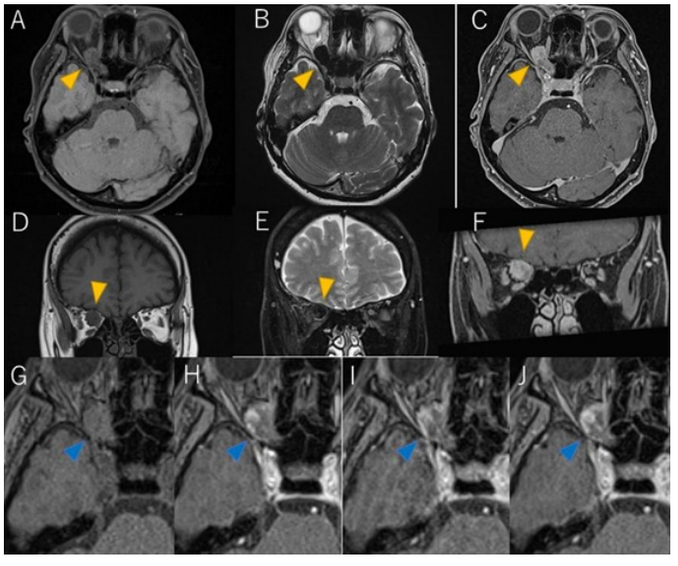

Figure 2: MRI revealed a 30x12x12 mm tumorous lesion that had extended from the right orbit through the optic canal to the intracranial space, and was compressing the optic nerve. T1- (A, D) and T2- (B, E) weighted images showed low signals and heterogeneous contrast effects with Gadolinium (C, F) (yellow arrows). Dynamic imaging taken every 30 seconds after gadolinium contrast injection showed that the tumor (blue arrows) was slowly enhanced (G, H, I, J).

The patient was placed in the supine position, and a frontotemporal craniotomy was performed with a microscope under general anesthesia. After retracting the right frontal lobe extradurally, orbital roof and anterior clinoid process were drilled off. After opening the right optic canal and incising the dura mater, a white elastically hard tumor was observed. When the intracranial part of the tumor was observed, it was strongly adherent to the right optic nerve, so the optic nerve was incised and the tumor inside the skull was removed. The tumor was macroscopically removed by entering the orbit from the lateral side of the superior rectus muscle and removing the tumor without damaging the trochlear nerve (Fig. 3).

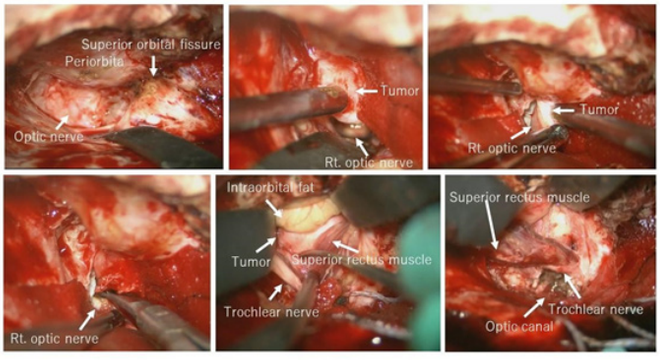

Figure 3: Intraoperative photographs. When the dura was incised, a white, elastic hard tumor was observed. When the intracranial tumor was observed, it was strongly adherent to the right optic nerve, so the optic nerve was incised and the tumor inside the skull was removed. The tumor was macroscopically removed by entering the orbit from the lateral side of the superior rectus muscle and removing the tumor without damaging the trochlear nerve.

Histopathologically, the mass consisted of collagen fibrous stromal tissue, presenting with poor-atypical columnar nuclei and rough proliferation of eosinophilic fusiform cells. Immunopathologically, the tumor was negative for S-100, glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA), synaptophysin, while was positive for cluster of differentiation (CD)34, B cell/chronic lymphocytic leukemia (CLL) lymphoma (BCL)-2. Moreover, signal transducer and activator of transcription (STAT)6 was positive in intranuclear, and Ki-67 was negative in most of the cells (Fig. 4). The postoperative course is uneventful without recurrence after 4 years (Fig. 5)

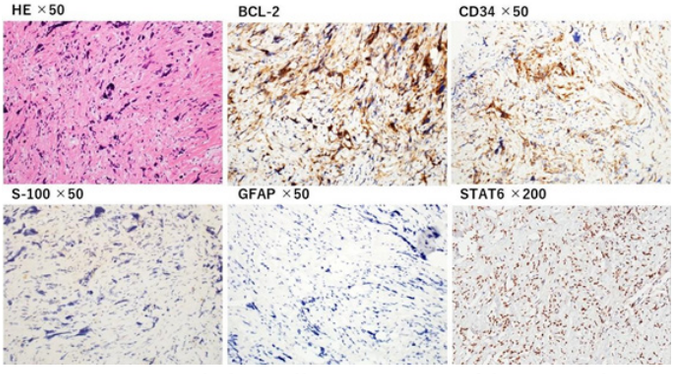

Figure 4: Histopathologically, HE staining of the resected tumor showed that it was composed of collagenous stromal tissue, with sparse proliferation of columnar nuclei and eosinophilic spindle-shaped cells with little atypia. Immunostaining showed negative results for S-100, GFAP, positive results for CD34 and BCL-2, and positive results for STAT6 in the nucleus.

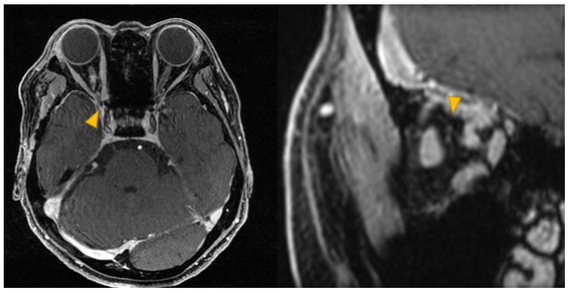

Figure 5: Postoperative MRI showed that the tumor had been completely removed, with no residual tumor.

This case report complies with the Declaration of Helsinki, and the corresponding author explained to the patient the significance of submitting this case to a medical journal as an academic paper, and written informed consent was obtained from the patient.

Discussion

SFT and HPC were considered separate diseases until the fact that gene fusion between nerve growth factor-induced protein A (NGFI-A) binding protein 2 (NAB2) and STAT6 being the driver mutation was discovered in 2013. Now SFT and HPC which has NAB2-STAT6 gene fusion are treated as a fibroblastic type of mesenchymal tumor from world health organization (WHO) classification of tumors of the central nervous system in 2016. Grade is classified into three categories, based on factors such as cell density, nuclear atypia, and mitosis. In addition, positive findings of NAB2-STAT6 gene detection and immunostaining with anti-STAT6 antibody are recommended to confirm the diagnosis[5,6]. Rare cases of strong positivity for progesterone receptors have also been reported[7]. Surgical removal is the mainstay of the treatment[8,9]. Some cases that have ended in partial resection report reccurrence or malignant transformation; affluent cellularity, pleomorphism, enlarged tumor size, and high levels of mitotic figures are implicated in the risk factors[4,10-18]. In the study of the risk of recurrence in orbit SFT, a higher recurrence rate was reported in OCCR / extraconal compared to intraconal[19]. Stereotactic radiotherapy and proton beam therapy are used for residual tumor and malignant SFT, although the efficacy of chemotherapy and radiation therapy has not been established[16,20]. Jackson et al reported in their 7 cases of SFC of orbit and 150 cases of literature review, that published rates of primary malignancy and recurrence across all histologic categories were 6% to 12% and 30% to 37%, respectively, and a treatment algorithm was predicated on the completeness of surgical excision and histological features[21]. In our case, histopathology revealed abundant collagen fibers, low cell density, poor nuclear atypia, and STAT6 positivity, suggesting grade 1 SFT, but the patient is under careful follow-up due to the tumor location.

The imaging findings of SFT are seen as a round or lobulated extraparenchymal mass with well-defined borders and heterogeneous iso- to hyper-density on CT scan, which may be accompanied by calcification and bone infiltration[11,22.] MRI shows a uniform or heterogeneous contrast effect, often with low to equal signal on T1WI and mixed low and high signal at T2WI[23]. Contrast effects similar to a dural tail enhancement along the adjacent dura mater may also be seen[11,16]. On preoperative MRI, the differential diagnosis of this tumor is sheath meningioma or optic schwannoma based on the site of origin, but the most likely differential diagnosis is meningioma, considering the frequency of occurrence and low signal in T1WI and T2WI, and the contrast findings. However, considering the strong low signal on T2WI and the slow contrast findings on dynamic contrast-enhanced MRI, fibrous tumors should also be considered for differential diagnosis.

Conclusion

SFT of the orbit should be considered as a differential diagnosis although it is extremely rare disease. It is similar to meningioma and schwannoma and we should take Dynamic-Contrast-Enhanced MRI for preoperative diagnosis. Careful postoperative follow-up is needed because some cases report recurrence and malignant transformation.

Conflicts of Interest Disclosure

The authors declare no conflicts of interest regarding this study.

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