

# Inflammatory Myofibroblastic Tumor of the Cerebellar Hemisphere

## —Case Report—

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### Abstract

**A 60-year-old man presented with a rare cerebellar inflammatory myofibroblastic tumor (IMT) manifesting as gait disturbance and dysarthria. Brain magnetic resonance imaging demonstrated an intra-axial round-shaped isointense mass homogeneously enhanced with gadolinium in the right cerebellar hemisphere, as well as perifocal edema extending to the brain stem and right thalamus. The tumor was elastic hard and was resected en bloc with a clear margin. Histological examination revealed IMT with spindle cells and collagen, but negative for anaplastic lymphoma kinase expression. IMT most commonly affects the lung, but may involve many other parts of the body. There is some debate regarding the disease entity of IMT in the central nervous system (IMT-CNS) because of its rarity and high frequency of recurrence. IMT-CNS is an important differential diagnosis among tumor-like intracranial lesions and total resection is required.**

Key words: brain edema, central nervous system, cerebellar tumor, inflammatory myofibroblastic tumor, inflammatory pseudotumor

### Introduction

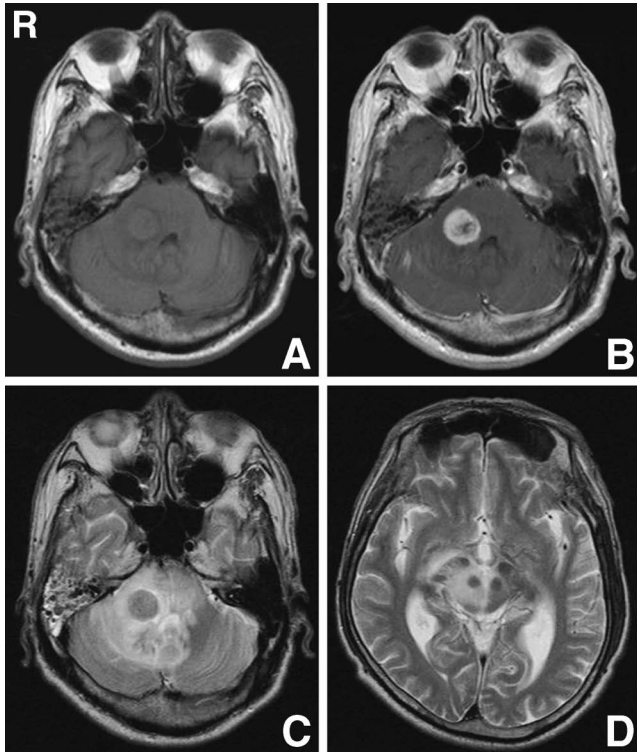
Inflammatory myofibroblastic tumor (IMT), or inflammatory pseudotumor, is a rare lesion consisting of myofibroblastic spindle cells and inflammatory cells that occurs primarily in soft tissue and the viscera of children and young adults.<sup>3,5</sup> IMT can affect any part of the body, but is most commonly seen in the lung, mesentery, and omentum, and usually follows a benign clinical course. Diagnosis depends on histological examination because the radiological and clinical findings are non-specific. IMT has been described under many different names, such as inflammatory pseudotumor and plasma cell granuloma, suggesting the complexity and variable histological characteristics of the tumor.<sup>8</sup> The etiology of IMT remains unknown, and both neoplastic and non-neoplastic processes have been proposed.<sup>11</sup> IMT in the central nervous system (IMT-CNS) is rare, and tends to arise from meningeal structures and recur. The disease entity of IMT-CNS has become contentious recently because of the high frequency of recurrence and malignant transformation compared with IMT not affecting the CNS (IMT-nCNS).<sup>6,10,11</sup>

Here we describe a case of cerebellar intraparenchymatous IMT in an adult, which seemed to exhibit the benign and inflammatory characteristics of IMT-CNS or inflammatory pseudotumor.

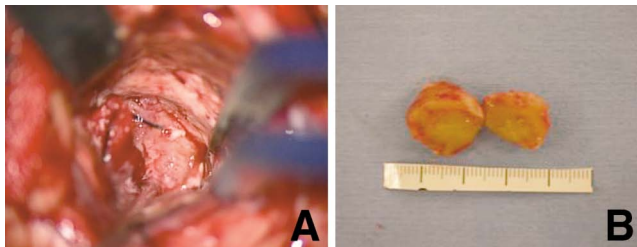
### Case Report

A 60-year-old man was admitted to our hospital complaining of rapidly progressing gait disturbance and ataxia. Neurological examination revealed right abducens palsy, right facial palsy (House-Brackmann grade III), dysarthria, and cerebellar ataxia. There was no past history except for diabetes mellitus. General physical examination was within normal limits. Results of blood examination were normal, including tumor markers (squamous cell carcinoma antigen, carcinoembryonic antigen,  $\alpha$ -fetoprotein, carbohydrate antigen 19-9, and prostate-specific antigen),  $\beta$ -microglobulin, and interleukin-2 receptor. Computed tomography showed a homogeneously enhanced, round-shaped intra-axial mass in the right cerebellar hemisphere compressing the brain stem. Brain magnetic resonance imaging demonstrated an isointense mass enhanced by gadolinium, with perifocal edema extending to the brain stem and right thalamus (Fig. 1). Cerebral angiography showed no abnormalities and whole-body gallium scintigraphy was negative.

Intraoperatively, a yellowish, well-demarcated tumor was identified using a navigation system (Fig. 2A). The tumor was elastic hard and fibrous, and approximately 18 mm in size. The lesion was resected en bloc (Fig. 2B). Histological examination revealed spindle cells, collagen, and central necrosis (Fig. 3A). Immunostaining demonstrated that the spindle cells reacted with smooth muscle actin and vimentin (Fig. 3B, C). The histological diagnosis



**Fig. 1** A: Axial T<sub>1</sub>-weighted magnetic resonance (MR) image demonstrating a round-shaped isointense mass in the right cerebellar hemisphere. B: Axial T<sub>1</sub>-weighted MR image after administration of gadolinium showing the mass is well demarcated and homogeneously enhanced. C, D: T<sub>2</sub>-weighted MR images showing the isointense mass with perifocal edema extending to the brain stem and the right thalamus.

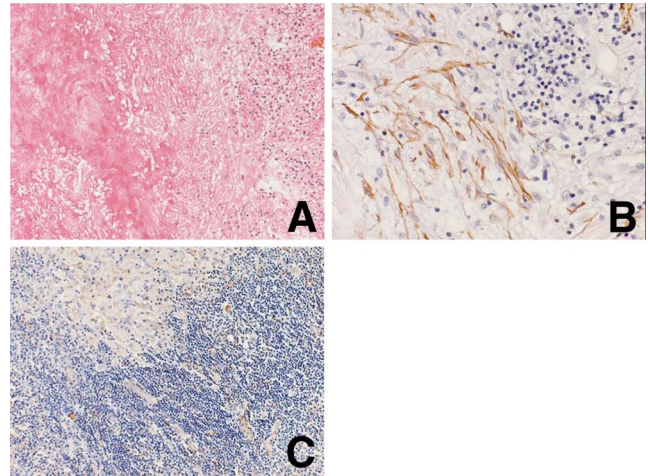


**Fig. 2** Intraoperative photographs showing a yellowish, well-demarcated tumor in the cerebellar parenchyma (A), and the resected mass is smooth and firm, and uniform throughout (B).

was IMT (inflammatory pseudotumor). No mitoses were detected, and the MIB-1 index was lower than 1%. Anaplastic lymphoma kinase (ALK) expression was negative. The postoperative course was uneventful and no recurrence was observed during the 8-month follow-up period.

## Discussion

IMT is a distinctive myofibroblastic spindle cell lesion accompanied by an inflammatory infiltrate of plasma cells,



**Fig. 3** A: Photomicrograph of the tumor showing spindle cells, collagen, and central necrotic zones. Hematoxylin and eosin stain, original magnification  $\times 40$ . B, C: Immunohistochemical examination showing the spindle cells are positive for smooth muscle actin (B) and vimentin (C).  $\times 40$ .

lymphocytes, and eosinophils.<sup>4)</sup> IMT-CNS is rare, with only approximately 70 reported cases.<sup>6,10)</sup> In the present case, positive immunohistochemical results for smooth muscle actin and vimentin agreed with previous reports of IMT.<sup>3,5,11)</sup> Localization of IMT can be divided into 5 types: intraparenchymatous, meningeal, mixed intraparenchymatous and meningeal, intraventricular, and extending to the cranial cavity/sphenoid sinus.<sup>6)</sup> The etiology of IMT-CNS is unknown, but 60% (34/57) of IMT-CNS tumors have arisen from the dural/meningeal structures and only 12% (7/57) from intraparenchymatous lesions.<sup>6)</sup> The present case had no attachment to dural/meningeal structures, and was classified as the intraparenchymatous type.

The neuroimaging findings and clinical course of IMT-CNS are non-specific, such that differential diagnosis is important in the case of meningeal lesions, especially with meningioma or sarcoma.<sup>1,2,9,12)</sup> Earlier cases likely included an indiscriminate mixture of neoplastic and non-neoplastic process. Of patients with extrapulmonary IMT, 15% (13/84) had one or more recurrences over a period of 1–24 months (mean 6 months),<sup>3)</sup> but patients with IMT-CNS had recurrence rate after incomplete resection of 40% within 2 years.<sup>6)</sup> Two of the three young patients with IMT-CNS extending from the meningeal structures into the brain tissue showed local recurrence after resection of the tumor and histological investigations revealed transformation into a semimalignant fibrohistiocytic tumor in one patient.<sup>6)</sup> Therefore, whether IMT-CNS is a different entity from IMT-nCNS remains controversial.<sup>6,10,11)</sup> The potential risk of malignant transformation and high risk of recurrence necessitate close follow up, particularly if resection was incomplete.<sup>7)</sup>

Staining of ALK on chromosome 2p23, which is well known from anaplastic large cell lymphomas, can be detected in approximately 50% of cases of IMT-nCNS and

correlates with the rearrangement of ALK.<sup>4,6,9,11</sup>) This suggests that some of IMTs with ALK activation could be of neoplastic origin.<sup>10</sup>) Three of four cases of IMT-CNS showed ALK protein overexpression immunohistochemistry.<sup>11</sup>) The present case was negative for ALK, and MIB-1 index was lower than 1%. However, necrosis was detected microscopically. Necrosis was detected in 6 of the 60 cases of IMT-CNS,<sup>6</sup>) and atypical forms may undergo malignant transformation.

Orbital pseudotumor extending into the cranial cavity is a well-described, distinct entity and does not require total resection. In contrast, there is some debate regarding the nature of IMT-CNS, such as whether this is a homogeneous entity, whether the tumor is neoplastic, and if neoplastic, the level of malignancy. No specific treatment protocol has not been developed for IMT-CNS because of its rarity, but total resection and close follow up seem essential because of the variable prognosis.

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