Inflammatory myofibroblastic tumor of the pituitary: A case report

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ABSTRACT

Introduction: Inflammatory myofibroblastic tumor (IMT), a subtype of inflammatory pseudotumor, is a rare cause of non-neoplastic sellar mass. IMTs have been reported in various anatomical locations of the body, but their presence in the Central Nervous System (CNS), especially in the pituitary gland, is rare. The objective of our case report is to analyze the basis of their mimicry, discuss their differential diagnosis, and review the literature related to inflammatory pseudotumors of the pituitary.

Case presentation: A 45-year-old male presented in our hospital with chief complaints of severe headache since two months ago and blurring of vision since two weeks ago. On investigation, Magnetic Resonance Imaging (MRI) revealed a pituitary macroadenoma, which was compressing the optic chiasma and encasing the left internal carotid artery. The patient underwent endonasal endoscopic transsphenoidal excision of the pituitary lesion and

became asymptomatic in the postoperative period. Histopathology reported the presence of spindle-shaped cells, inflammatory cells, respiratory epithelium lined tissue, and areas of necrosis suggestive of an inflammatory myofibroblastic tumor.

Conclusion: In the following communication, we discuss this esoteric pseudotumor of the pituitary and its peculiarities in diagnosis and management as it can easily disguise itself as a pituitary macroadenoma on radiological investigations, thus hoodwinking the physician of its actual pathology. We also review the literature to perceive the best management protocols and prognostic factors by drawing parallels with IMT in other parts of the CNS. The differential diagnosis of inflammatory myofibroblastic tumors of the CNS is difficult and relies mainly on histological analysis.

Keywords: Inflammatory myofibroblastic tumor, pituitary neoplasm, plasma cell granuloma, pseudotumor

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INTRODUCTION

Inflammatory Myofibroblastic Tumors of Central Nervous System (IMT-CNS), have been reported in literature sporadically over the last few decades, but their presence in the pituitary gland was first reported in 2001. Although the pulmonary system is the most frequently involved site, the CNS, gastrointestinal tract, tonsils, urogenital tract, heart, and orbit may also be affected. Concomitant involvement, such as that of the lung and CNS, might be encountered as well. Only a few cases of Inflammatory Myofibroblastic Tumors of the pituitary gland (IMT-Sella) have been reported.

We discuss a case of pituitary IMT that mimicked pituitary macroadenoma but was later diagnosed on histopathology. The objective of our case report is to analyze the basis of their mimicry, discuss their differential diagnosis and review the literature for their behaviour, management protocols and prognosis by searching PubMed using the keywords such as “inflammatory myofibroblastic tumor”, “plasma cell granuloma”, “inflammatory

pseudotumor”, “pituitary” and references cited in identified PubMed articles.

CASE PRESENTATION

A 45-year-old male presented with complaints of moderate to severe intensity, bifrontal headache for two months and blurring of vision in the temporal side of visual field since two weeks ago. Neurological examination revealed bitemporal hemianopsia. His blood work was suggestive of panhypopituitarism. The patient was evaluated radiologically, and MRI revealed a large well defined T1 hyperintense lesion involving sella extending to the suprasellar region. The lesion measured 2 x 2.1 x 1.6 cm. It appeared hyperintense in T1W1 and hyperintense in STIR with small hypointense areas. The lesion had multiple CSF signal intensity areas suggestive of cystic change. The T2W1 lesion had well defined peripheral hyperintense rim. The lesion extended into the suprasellar region. The optic chiasm appeared compressed by the mass lesion. Laterally, on the left side of the lesion was seen encasing the

Figure 1. Brain MRI. Sagittal view showing hyperintense lesion on T1 image extending into the suprasellar region and compressing optic chiasm.

Figure 2. Microscopy showing spindle cells with moderate cytoplasm and elongated nuclei.

Figure 3. Immunohistochemistry showing Vimentin positivity.

Figure 4. Immunohistochemistry negative for ALK.

internal carotid artery (**Figure 1**).

A working diagnosis of pituitary adenoma was made, and the patient was taken up for endonasal endoscopic transsphenoidal total excision of pituitary adenoma. Intraoperatively, the tumor was firm to hard in consistency, grey-white in color and moderately vascular. In the post-op period, there were no complications such as CSF leak, bleeding, diabetes insipidus, and neurological deficit. It was uneventful, and the patient was relieved of his symptoms.

The histopathology revealed neoplasm composed of spindle-shaped cells arranged in short bundles. Individual cells were found to be spindle with a moderate amount of eosinophilic

cytoplasm and elongated nuclei with tapered edges. Sprinkled in between these were epithelioid cells and lymphocytes. RBC's along with hemosiderin-laden macrophages, respiratory epithelium lined tissue, and few areas of necrosis were also noted (**Figure 2**).

Immunohistochemistry of spindle cells was positive for Smooth Muscle Actin (SMA), Vimentin, negative for Anaplastic Lymphoma Kinase (ALK), CD1(a) and S100 (**Figure 3 and 4**).

Histological diagnosis of IMT was made. On discharge, the patient was given a course of corticosteroids. Six months post-surgery, repeat MRI revealed no residual/recurrent tumor.

DISCUSSION

IMT is an arcane inflammatory pseudotumor first described by Bahadori and Leibow in 1973.¹ Eventually it was found to be ubiquitous occurring commonly in the lung, omentum, mesentery and urinary bladder of children and young adults.² IMT-CNS is a rare entity, with only 100 cases being reported so far.³ Most tumors arise from meningeal structures, though intracerebral and intraventricular lesions have been known to occur.⁴ From our literature search, only 7 cases of intrasellar IMT have been reported until now (Table 1).⁵⁻¹¹

The scarcity of literature available on intrasellar IMT compelled us to extrapolate data from the cases, occurring in other areas of the central nervous

system. Average age, median age and age range of IMT-CNS varies in different reports but that of IMT-Sella is found to be 44.8 years, 36 years and 18 – 62 years respectively. No gender predominance was seen in our review for IMT-Sella, which is incongruent with a male predominance of IMT-CNS.¹² (Table 2)

Etiopathogenesis of IMT remains unascertained with the ongoing debate between infective and autoimmune pathology. Clinically, IMT-Sella presents with features of headache, vision problems and hypopituitarism. Differential diagnoses of pituitary IMT are pituitary adenomas, inflammatory and granulomatous lesions of pituitary, meningioma and lymphomas. Radiologically, IMT-Sella appears hyperattenuated

Table 1. Summary of cases of inflammatory myofibroblastic tumors of the pituitary in the literature

Study	Sex/Age	Complaints	Therapy	Histology	Follow up	Outcome
Hansen et al. ⁶ (2001)	F/40	Headache, left-sided loss of vision, secondary amenorrhea, asthenia	Biopsy + corticosteroids + azathioprine + methotrexate	No type predominance indicated	2 years and 6 months	1 st recurrence on 16 months, 2 nd recurrence on 25 months, asymptomatic at the end of 2.5 years.
Murakami et al. ⁷ (2001)	F/34	Left-sided ptosis	Total gross resection	PCG	NM	Uneventful postoperative period
Al-Shraim et al. ⁵ (2004)	M/32	Polyuria, polydipsia, anorexia	Total gross resection + corticosteroids	FHC	NM	Patient responded well
Jochum et al. ⁸ (2004)	F/19	Headache, secondary amenorrhea	Biopsy + partial resection + stereotactic radiotherapy	FHC	NM	Tumor size kept increasing
Yamagami et al. ¹⁰ (2008)	M/18	Headache, nasal discharge, polyuria, polydipsia, short stature	Biopsy + corticosteroids	FHC	2 months	Headache and nasal discharge improved, no improvement in pituitary dysfunction
Kim et al. ⁹ (2011)	M/38	Anorexia, sexual dysfunction, ophthalmoplegia, diplopia	Partial resection + corticosteroids	FHC	3 years	MRI showed shrinkage of sellar mass
Gautam et al. ¹¹ (2017)	F/62	Right focal motor seizures	Total gross resection	NM	1 year	No residual or recurrent tumor, patient asymptomatic
Present study (2016)	M/45	Headache, blurred vision	Total gross resection + corticosteroids	FHC	1 year	No residual or recurrent tumor, patient asymptomatic

*NM: not mentioned; PCG: Plasma cell granuloma type; FHC: Fibrohistiocytic type

on CT and homogenous hypointense lesions on T2 weighted images because of the lack of mobile protons in the fibrotic background.⁹

On histopathology, IMT is a unique lesion composed of myofibroblastic spindle cells accompanied by inflammatory cells.¹⁵ IMTs were classified into three histological patterns by Coffin *et al.* based on pulmonary and extra-pulmonary location.² However, a classification relevant to IMT-CNS was proposed by Jeon *et al.* in 2005. They described a two-tiered classification system of Plasma Cell Granuloma (PCG) variant and fibrohistiocytic (FHC) variant.¹² The FHC variant is characterized by myofibroblastic proliferation with slight inflammatory cell component and the PCG like variant is the exact opposite, with distinct inflammatory cell component and limited myofibroblastic proliferation. In their study, Jeon *et al.* noticed, both variants presented equally, though in our review, FHC variant was predominant (**Table 1**). This clearly highlights the histopathological differences between the two entities, though its impact on disease progression cannot be stated,

without carrying out further long term studies.

Immunohistochemically, these tumors are positive for SMA, muscle-specific actin, vimentin in up to 90% cases, desmin in up to 70% cases and negative for myogenin, S100 and Homatropine Methylbromide 45 (HMB45). In situ hybridization of FHC variant may often show clonal rearrangement in chromosome 2p23 leading to activation of the ALK gene. It has been reported that those IMT-CNS, which express ALK, have an aggressive course and are associated with high recurrence rates.¹⁶

Treatment modalities considered for IMT-CNS are surgical resection, corticosteroid therapy, radiotherapy and immunosuppressive therapy with ALK inhibitors. Treatment patterns mentioned in the literature and the present review are elaborated in **Table 3**. According to all of these studies, total surgical excision if achieved without much morbidity is the best modality. Corticosteroid therapy can be considered to avoid considerable surgical risk, especially in PCG like variants, which respond better to corticosteroid therapy than FHC variants.⁶

One recurrence and no cases of malignant transformation were noted in our study, although the average follow-up time was just 20 months, and maximum follow-up was carried out only for 3 years. Recurrence rates of 12.5% to 40% in IMT-CNS cases have been noted in the literature.¹⁷ (**Table 3**)

Gallago *et al.* studied all cases of recurrence in IMT-CNS and found 19 such instances in over 100 cases reviewed.³ Going through their data, we noted that the PCG variants had higher chances of recurrence than the FHC variants, and 11 out of 19 (57.8%) patients were females. Twelve of the cases (63%) who showed recurrence had not undergone gross total resection as their primary modality of management. The average time until recurrence for

Table 2. Demographic data of inflammatory myofibroblastic tumors of CNS and sella, as mentioned in literature.

Study	Average age (years)	Median age (years)	Range (years)	Sex ratio (M:F)
IMT-CNS				
Greiner <i>et al.</i> ¹³ (2003)	34.1	-	-	11:8
Buccolieri <i>et al.</i> ¹⁴ (2003)	-	32	5 – 76	7:3
Jeon <i>et al.</i> ¹² (2005)	43.7	42.5	24 – 65	3:2
IMT-Sella				
Present study	44.8	36	18 – 62	1:1

Table 3. Various treatment patterns and recurrence rates in inflammatory myofibroblastic tumors of CNS as described in the literature.

Study	TGR (%)	STR (%)	Biopsy (%)	Irradiation (+/- surgery) (%)	Corticosteroids (+/- surgery) (%)	Chemotherapy (+/- surgery) (%)	Recurrence rates (%)
Hausler <i>et al.</i> ⁴ (2003) (n=43)	65	25.5	37	30	25.5	0	25.5
Greiner <i>et al.</i> ¹³ (2003) (n=38)	60.5	24	16	24	0	0	-
Jeon <i>et al.</i> ¹² (2005) (n=10)	60	30	10	10	10	0	20
Present review (n=8)	50	12.5	37.5	12.5	62.5	12.5	12.5

TGR: Total gross resection; STR: Stereotactic radiotherapy

patients who underwent complete resection was 37 months, while the average time until progression/recurrence for the rest of the cases was 29 months. Hausler *et al.* also noted that the female gender and incomplete resection were independent risk factors for recurrence.⁴ Similarly, in our review, the only recurrence noted was in a female patient with incomplete resection.⁶

CONCLUSION

IMT-Sella is a rare neoplasm at a rare location. Histopathologically, these are FHC variant predominant lesions as compared to other CNS locations. Total surgical resection should be the target goals for treatment of all IMT. In case of difficult surgical access and contraindication to surgical procedures, treatment with corticosteroids can bring good results. The presence of PCG variant, female gender and incomplete resection are poor prognostic factors. It is important to differentiate the histopathological variant, so an early assessment can be made regarding the aggressiveness and malignant potential of the tumor and appropriate further management.

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CONFLICT OF INTEREST

No conflicts of interest arose.

AUTHORS' CONTRIBUTIONS

Sharma S is the corresponding author and was in charge of reviewing and editing the manuscript. Jyothish LS, Kutty R, and Peethambaran AK did the conception of the idea, reviewed and edited the manuscript. Nagar M and Jain S performed the literature search, reviewed and edited the manuscript.

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