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Case Report

Rare neurohypophyseal tumor presenting as giant pituitary macroadenoma with cavernous sinus invasion – A case report and review of literature

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ABSTRACT

Background: Granular cell tumors (GCTs) of the pituitary are rare tumors of posterior pituitary that can present as giant pituitary macroadenoma due to the slow indolent growth of the tumor. We are reporting this case due to the rarity of GCT and usually these tumors are confined to the suprasellar region since they are arising from the pituitary stalk. GCTs that attain such giant size with cavernous sinus invasion are still rarer.

Case Description: A 38-year-old female who presented with progressive deterioration of vision and on evaluation by magnetic resonance imaging showed a giant pituitary macroadenoma with bilateral cavernous sinus invasion. The patient underwent pterional craniotomy and near-total excision of the lesion was done due to high vascularity and firmness of the tumor. Histopathology examination of the lesion showed spindle to globular cells with granular cytoplasm and was reported as GCT of the pituitary.

Conclusion: GCTs are WHO grade1 non-neuroendocrine tumors arising from neurohypophysis and infundibulum. Complete excision is usually difficult due to the high vascularity, firm consistency, and local invasion of the tumor to the cavernous sinus and optic apparatus.

Keywords: Cavernous sinus invasion, Giant, Granular cell tumor, Non-neuroendocrine tumor, Pituitary

INTRODUCTION

Granular cell tumor (GCT) of the pituitary is a rare benign tumor arising from the pituicytes of the neurohypophysis. These are the most common benign tumors of the neurohypophysis. These tumors are characterized by their slow growth and present usually with the visual symptoms due to compression of the optic chiasm.[1] The incidence of symptomatic GCT is extremely rare and accounts for 0.5% of the sellar tumors. However, asymptomatic granular cell tumorlets have been described in 17% of autopsies.[2] There had been only two reported cases of such giant GCT of the pituitary with cavernous sinus invasion.[3,4]

CASE REPORT

A 38-year-old female presented with progressive deterioration of vision in both eyes for 3 years. It started as blurring of vision in the left eye which progressed to involve the right side also for

which she was prescribed spectacles by an ophthalmologist, but later she started to have difficulty seeing the outer part of her visual field. There was no history of headache, vomiting, sudden deterioration of vision, or seizures. There was no history suggestive of hyper or hypopituitarism. On examination, she was conscious oriented her visual acuity was 6/60 in the left eye and 6/12 on the right, her optic fields showed field loss on the right half on the right eye and almost complete loss of field in the left eye except for a small island in the center and she had partial optic atrophy of both eyes. Pupillary reflexes were normal in the right eye but her left eye had Grade 1 relative afferent pupillary defect. She was evaluated with magnetic resonance imaging brain with contrast which showed a well-enhancing sellar, suprasellar lesion around 4 cm × 4.2 cm × 2.5 cm with mass effect on optic chiasm and tracts. The lesion was invading bilateral cavernous sinus with encasement of the right internal carotid artery (ICA) and abutting the left ICA. There was no evidence of hydrocephalus [Figure 1]. Pituitary hormones were evaluated and showed low serum cortisol, the rest of the pituitary hormones were within normal limits.

We proceeded with right pterional craniotomy, trans-sylvian approach, and decompression of the lesion. The lesion was very vascular, firm, and not suckable. Near-total excision of the lesion was done, the optic apparatus was decompressed.

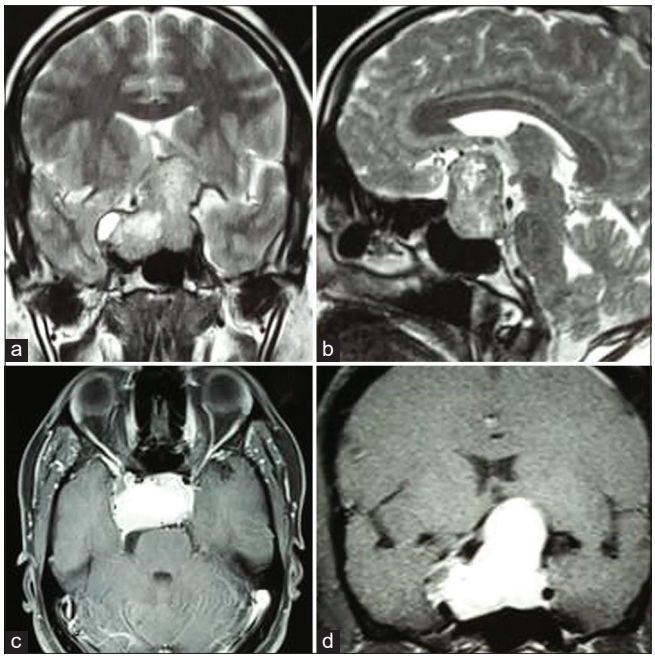

Figure 1: Magnetic resonance imaging of brain (a) coronal T2-weighted sequences showing the sellar-suprasellar lesion which invades both cavernous sinuses; (b) sagittal T2-weighted sequences; (c) axial T1-weighted contrast-enhanced sequences showing homogenous contrast enhancement; (d) coronal T1-weighted contrast-enhanced sequences.

Histopathology demonstrated large polygonal cells with eosinophilic granular cytoplasm. The tumor was positive for thyroid transcription factor 1 (TTF1) and focally positive for S100 [Figure 2] and CD68. MIB1 index is very low and GFAP was negative. The features were consistent with the GCT of the pituitary.

In the postoperative period, the patient developed transient diabetes insipidus which was managed with desmopressin nasal sprays and oral carbamazepine. On follow-up at 6 months, there was improvement in the visual acuity as well as the visual field.

DISCUSSION

GCTs of the pituitary are well-circumscribed benign tumors arising from the infundibulum or posterior pituitary. These tumors are characterized by their indolent growth and the incidence of symptomatic GCTs of the pituitary is extremely rare. Once they are becoming symptomatic these tumors would have grown to large proportions. There is a clear female predisposition (2:1). As the tumor is of significant

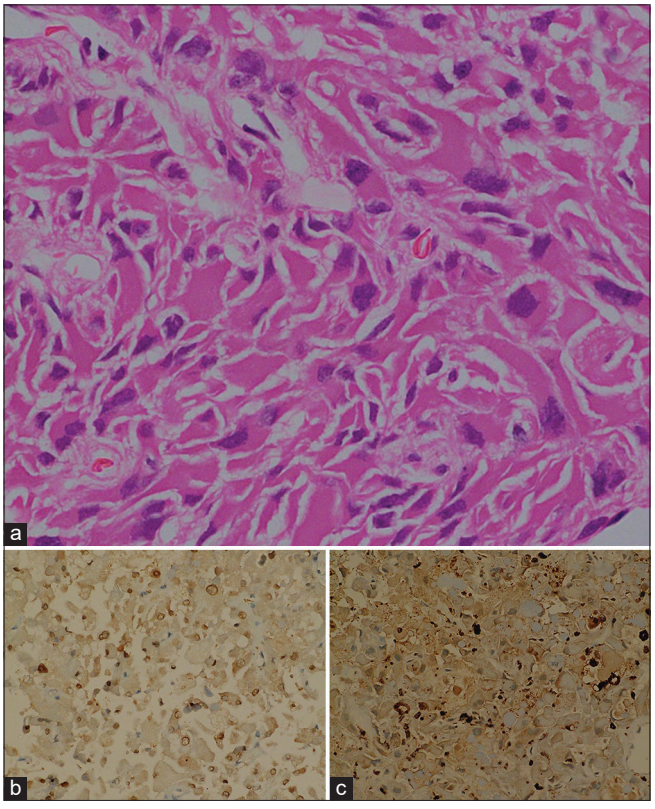

Figure 2: Photomicrographs of the tumor. (a) Hematoxylin-eosin staining (original magnification ×400) showing spindle to polyhedral cells with abundant granular eosinophilic cytoplasm; (b) immunohistochemistry (IHC), (original magnification ×400) showing TTF1 positivity; (c) IHC showing focal S100 positivity (original magnification ×400).

Table 1: Comparison of various non-neuroendocrine tumors of pituitary.

	Pituicytoma	Granular cell tumor	Spindle cell oncocytoma	Sellar ependymoma
Mean age (years)	47	49	56	50
Sex predilection	Male	Female	Male	Male
Cell of origin	Major and dark pituicytes	Granular pituicytes	Oncocytic pituicytes	Ependymal pituicytes
Cavernous sinus invasion	Rare	Rare	3%	Rare
Histology	Elongated bipolar cells with eosinophilic nongranular cytoplasm arranged in fascicles	Sheets of polygonal granular cells with eosinophilic cytoplasm arranged in fascicles	Interlacing fascicles and poorly defined lobules of spindle cells with granular eosinophilic cytoplasm with mild-to-moderate atypia and pleomorphism	Broad fascicles of bipolar cells accompanied by typical ependymal features with perivascular pseudorosettes or true rosettes
Immuno histochemistry	TTF1, S100, GFAP	TTF1, S100, CD68, Alpha-1-antitrysin, Alpha-1-antichymotrysin.	TTF1, S100, GFAP	TTF1, S100, vimentin, GFAP, CD99
MIB index	0.5–2%	1–7%	1–8%	<1%

size once it becomes symptomatic, the major symptoms are attributed to mass effect from compression of the optic apparatus. Furthermore, there can be panhypopituitarism and very rarely diabetes insipidus. MR imaging usually shows a suprasellar mass with homogenous contrast enhancement. Calcifications are not common.[5] The tumor size at presentation varies from 1.5 cm to 6 cm. Radiologically, these tumors are difficult to differentiate from a pituitary adenoma, but in our case, the well-enhanced feature of the tumor was more in favor of a lesion arising from neurohypophysis. The cell of origin of this tumor is thought to be pituicytes since the tumor expresses nuclear TTF1 which is also seen in other tumors originating from pituicytes such as pituicytoma and spindle cell oncocytoma. Microscopically, the tumor shows densely packed polygonal cells with granular cytoplasm. On immunohistochemistry, the GCTs are variably positive for CD68, S100, alpha1 antitrypsin, and alpha1 antichymotrypsin. Most tumors are negative for GFAP.[10]

In the WHO classification of endocrine tumours (4th edition) which was released in 2017, the nonendocrine tumors arising from posterior pituitary were classified as distinct low-grade neoplasms that express TTF1. These include pituicytoma, GCT of the sellar region, spindle cell oncocytoma, and sellar ependymoma and constitute a morphological spectrum of a single nosological entity. The clinical and histological parameters which will aid us to diagnose these lesions are compared below [Table 1].[2,3,6,8] These tumors show overlapping histological features and IHC plays a significant role in diagnosis. Among these tumors, GCT of the sellar region appears to be the only lesion with relatively specific histological and IHC characteristics.[3,4]

Prognosis of these tumors depends on the extent of resection.[9] Highly vascular nature of the tumor (due to the surrounding vascular plexus) as well as the adherence of the lesion to the pituitary gland and infundibulum makes the gross total excision difficult.[1,7]

CONCLUSION

GCTs are very rare tumors confined to the suprasellar region since they are arising from the pituitary stalk. GCTs which arise from the posterior pituitary are still rarer. They grow to large proportions due to the slow indolent growth of the tumor. Even though these lesions are WHO Grade 1, complete excision is usually difficult due to the high vascularity, firm consistency, and local invasion of the tumor to the cavernous sinus and optic apparatus.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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