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Case Report & Literature Review

Pituicytoma: A Rare Clinical Entity and Review of Literature

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ABSTRACT

Pituicytoma is an exceptionally rare low grade glioma (WHO grade I) of the neurohypophysis and infundibulum. It may effect neurohypophysis and infundibulum separately or together. The clinical presentation is similar to other pituitary tumours and imaging examination may suggest pituitary adenoma. Diagnosis is based on histopathological analysis. We present a pituicytoma case in a 48 years old female presenting as a focal lesion of the neurohypophysis. This case report reviews the clinical, neuroimaging and histopathological features of this rare tumour to diagnose it better. The pituicytomas may have originated in the pituitary stalk and the differential diagnosis should be compared with the pituitary stalk mass.

KEYWORDS: Histopathology, Immunohistochemisty, Neuroimaging, Pituicytoma.

INTRODUCTION

Pituicytoma is an extremely rare primary tumour of the sellar and suprasellar region that originated from pituicytes in the neurohypophysis or infundibulum[1]. It may effect neurohypophysis and infundibulum separately or together. The clinical presentation is similar to other pituitary tumours and imaging examination may suggest pituitary adenoma. Diagnosis is based on histopathological analysis. In 1955, Scothorne reported the first case[2]. Since then, only a few cases of true pituicytomas have been reported in the literature[3-14]. We report a rare case of pituicytoma and discuss the clinical, neuroimaging characteristics, surgical findings and histopathological features of this rare entity.

CLINICAL SUMMARY

48 years old female presented with history of visual disturbance since 8 months and recent onset headache for a period of 3 days. Visual disturbance was in form of inability to see side ways. Headache was gradual in onset, generalised, intermittent, throbbing in nature, mild to moderate in severity with no known aggravating or relieving factors. Visual field assessment revealed bitemporal hemianopia. Serum pituitary hormonal profile was grossly normal except Serum Prolactin level (96.82 ng /ml) which was slightly elevated. Serum sodium was 133 mEq / L. MRI brain with contrast revealed a well-defined, rounded T1 isointense (Fig 1), T2 and FLAIR hyperintense lesion measuring 1.4 x 1.5 cm in the pituitary fossa. (Fig 2) The pituitary gland was not separately made out from the lesion. The lesion showed homogenous post contrast enhancement and was compressing the adjacent optic chiasma. (Fig 3) She underwent neuronavigation guided endoscopic binostril transsphenoidal approach and excision of the lesion. Follow up after 4 months following surgery, she was completely asymptomatic and her visual symptoms impoved significantly.

PATHOLOGICAL FINDINGS

The tumour was soft, suckable and highly vascular. The histopathological examination revealed fragments of a neoplasm composed of cells arranged in fascicles and bundles. The cells are plump to elongated to spindly with moderate eosinophilic cytoplasm, plump to elongated nuclei with vescicular chromatin, conspicuous nucleoli. Numerous vascular channels were seen throughout. No mitosis or necrosis seen. The immunohistochemistry showed positivity for vimentin, TTF-1 and S-100 and showed focal positivity for GFAP, CD 34 and EMA. The tumour was non immunoreactive for syanptophysin. Ki 67 was 3-4%. Given the histology, location and immunohistology staining, pathologic diagnosis of pituicytoma was made.

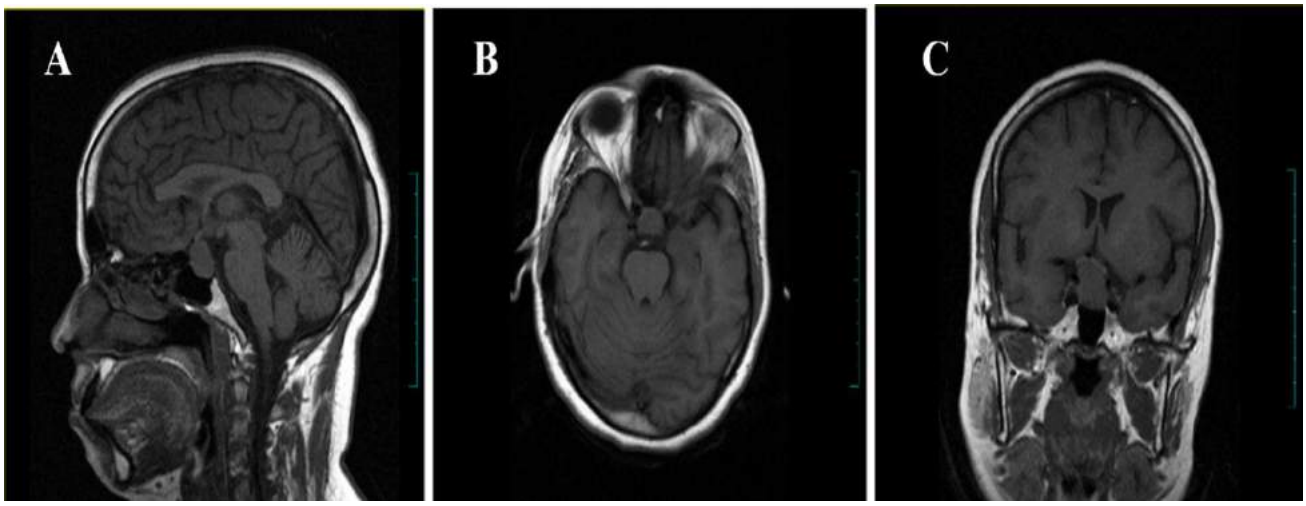

Fig 1: MRI BRAIN - T1 Images: A - Sagital View, B - Axial View and C - Coronal View showing well defined isointense lesion in the pituitary fossa

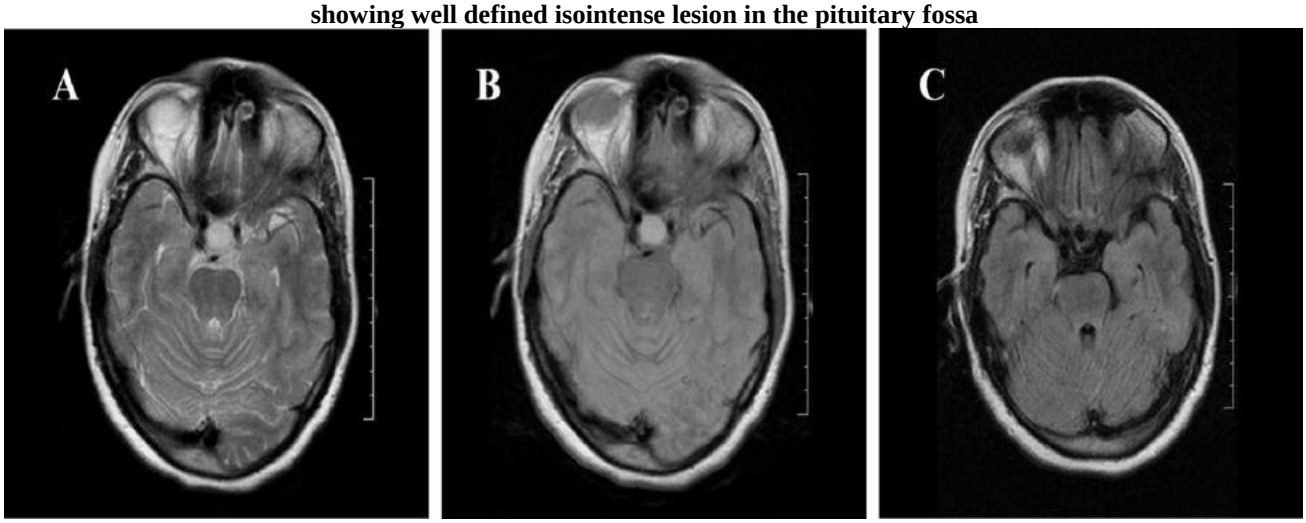

Fig 2: MRI BRAIN - Images: A - T2 Axial View and B - FLAIR - Axial View showing well defined hyperintense lesion in the pituitary fossa and C - Post Operative FLARE - Axial View showing complete excision of the lesion on follow up scan

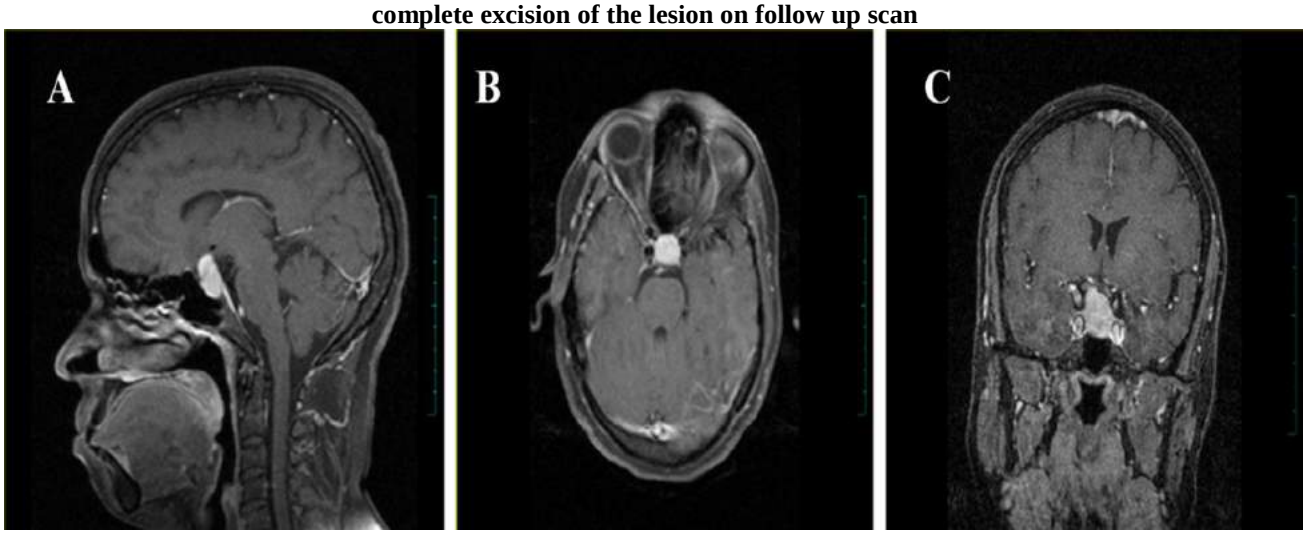

Fig 3: MRI BRAIN - Post Contrast Images: A - Sagital View, B - Axial View and C - Coronal View showing homogenous post contrast enhancing lesion compressing the adjacent optic chiasma

DISCUSSION

Pituicytoma is a benign, slow-growing and special neoplasm of the neurohypophysis and it is confused with more common neoplasms, such as pituitary macroadenoma, meningioma and craniopharyngioma. Pituicytoma is a rare neoplasm in adults. It corresponds to a low-grade astrocytoma of the neurohypophysis that presumably arises from pituicytes of the stalk and posterior lobe of the pituitary gland. This tumor is included in the 2007 World Health Organization (WHO) classification of tumors of the central nervous system as

a WHO grade 1 tumor.[15,16] Several authors have also used the term 'pituicytoma' to refer to other tumors in the neurohypophysis, especially pilocytic astrocytoma and granular cell tumor. The 2007 WHO classification of tumors of the central nervous system defines pituicytoma as a low-grade glial neoplasm of the neurohypophysis or infundibulum that originates from pituicytes Although the imaging characteristics of pituicytoma are not pathognomonic, these lesions are highly vascular, and MRI after gadolinium injection discloses a strong enhancement of the solid tumour. The diagnosis is usually with histological features.

The neuroimaging characteristics of pituicytomas are nonspecific. Pituicytomas have been described to be located within the sellar, the suprasellar region or both. Although there were not many reports on the CT findings of pituicytomas, in the case reported by Wolfe et al., the tumor showed sellar enlargement and bony remodeling on CT scan[4]. MRI shows an isointense mass on T1-weighted images, hyperintensity on T2-weighted images, and homogeneous enhancement with gadolinium[17]. Therefore, the radiological differential diagnosis should include other sellar or suprasellar tumors including meningioma, craniopharyngioma, hemangiopericytoma, granular cell tumor, and pilocytic astrocytoma. It is often difficult to identify these tumors in preoperative neuroradiological findings, especially for nonfunctioning pituitary adenoma in our case. However, the histologic appearance and immunohistochemical findings are distinctive. Pituicytomas contain elongated spindle cells that are arranged in a bundle or storiform pattern with no granular component. Staining is positive for S-100, focally positive for GFAP and EMA, and negative for PAS.

The pituicytoma is a rare neoplasm whose histogenesis is debated partly because of the diversity of tissue types present in the sellar region. In this article we illustrate the characteristic histologic, immunohistologic, and ultrastructural features of this unique neoplasm. Tumors were composed of cells arranged in fascicles and bundles. The cells are plump to elongated to spindly with moderate eosinophilic cytoplasm, plump to elongated nuclei with vescicular chromatin, conspicuous nucleoli. Numerous vascular channels were seen throughout. No mitosis or necrosis seen. The immunohistochemistry showed postivity for vimentin, TTF-1 and S-100 and showed focal positivity for GFAP, CD 34 and EMA. The tumour was non immunoreactive for syanptophysin. Ki 67 was 3-4%. In summary, our data suggest that pituicytomas are a unique subset of tumors of the sellar region.

The pituicytoma, or infundibuloma, is a rare tumor of the sellar region that displays a spindle cell morphology, fascicular arrangement of tumor cells, and variable glial fibrillary acidic protein (GFAP) immunoreactivity. GFAP is a 54kDa, type III intermediate filament protein that is the major constituent of the glial filaments in astrocytes[18]. GFAP is considered to be a general marker for astrocytes in the central nervous system[19]. The accurate diagnosis of brain invasion is therefore critical and in borderline cases, an immunohistochemical stain for GFAP aids in the delineation of entrapped glial elements within the substance of a brain invasive meningioma[20]. The transmembrane glycoprotein of EMA is encoded by MUC1 gene located on chromosome 1 in the 1q21-24 region. EMA is present in numerous epithelial cells. EMA, an integral mucin complex, is representative of the antigen involved in the secretory process of glandular epithelia, which change quantitatively during specialization, differentiation and neoplastic transformation[18]. Despite the characteristic histologic features of pituicytomas, they can be confused with other more common sellar or suprasellar neoplasms on the basis of their location and their solid, uniformly contrast-enhancing appearance on neuroimaging. By their morphology and immunostaining pattern, pituicytomas are thought to arise from pituicytes, specialized glial cells of the neurohypophysis. Recent data, however, suggest that pituicytomas may also share some features with the folliculostellate cell. Folliculostellate cells are nonendocrine, spindle-shaped cells of the adenohypophysis that express S100 and Bcl-2. The clinicopathologic features of this interesting neoplasm have recently been reviewed, but subsequent reports have invigorated the debate regarding the tumor's histogenesis. In this study, we present the histologic and immunohistologic features of a cases of pituicytoma. These findings clearly illustrate the unique features of these neoplasms.

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