the diagnostic pathology journal DIAGNOSTIC PATHOLOGY
Sundeep Mani, Aarchana Lakshmanan, Annapurneswari Subramanyan diagnostic pathology 2022, 7:284 ISSN 2364-4893
DOI: http://dx.doi.org/10.17629/www.diagnosticpathology.eu-2022-7:284

Case Report

Pituicytoma- Case Report of a Rare Sellar/ Suprasellar Mass

Sundeep Mani [1)] Aarchana Lakshmanan [2)] Annapurneswari Subramanyan [2]

1) Apollo Hospitals
2) Apollo cancer centre, Chennai

Abstract

Background: Pituicytoma is a rare primary tumor of the sellar and suprasellar region, arising from the pituicytes which are specialized glial cells in the neurohyphophysis and infundibulum.

Objective: Radiological features being non- specific, histopathological examination and immunohistochemistry will help us in arriving at the correct diagnosis.

Material and methods: We report a case of pituicytoma in a 48 year old woman arising in the sellar/suprasellar region.

Results: Histopathological examination of the lesion showed elongate bipolar spindle cells arranged in fascicular and storiform pattern. The neoplastic cells were diffusely positive for TTF-1, S-100, vimentin, variably positive for GFAP, negative for synaptophysin and Ki-67 index was less than one percentage.

Conclusion: Pituicytomas are rare tumors of the sellar and suprasellar region and its diagnosis is typically based on histopathological examination with a varied immunohistochemical profiles but with consistent TTF-1 positivity.

Key Words: suprasellar, pituicytoma, TTF-1, headache, visual disturbance

Key message: Pituicytoma is a rare primary tumor of the sellar and suprasellar region, arising from the pituicytes and its diagnosis primarily depends of histopathological examination and immunohistochemistry showing consistent TTF-1 positivity.

Sundeep Mani, Aarchana Lakshmanan, Annapurneswari Subramanyan diagnostic pathology 2022, 7:284 ISSN 2364-4893
DOI: http://dx.doi.org/10.17629/www.diagnosticpathology.eu-2022-7:284

Introduction

Pituicytomas are WHO grade I tumors defined by Brat et al. in 2000[3]. These are rare low grade glial neoplasms originating in neurohypophysis or infundibulum and approximately over 80 cases have been described in the literature[1]. The clinical features are due to slowly expansive non- hormonally active lesion which causes compressive effects on optic chiasma, infundibulum and/ or anterior pituitary gland. Radiologically these tumors are solid and circumscribed lesions and few cases reported with a cystic component[9]. These tumors usually occur in adults with a mean age at diagnosis being 50 years. Here we report a case of sellar/ suprasellar mass in a 48 year old woman.

Case History

A 48 year old female, known case of systemic hypertension and hypothyroidism on regular medications came with complaints of headache and visual disturbances. She also had history of tactile hallucinations with generalized tiredness and weakness. There was no history of abnormal weight gain/ loss, sleep disturbances, increased thirst or urination and tightness of limbs or weakness. MRI brain showed a well defined extra axial cystic lesion in the sellar region extending into the suprasellar area causing compression on the optic chiasma. She underwent endoscopic trans- nasal, trans- sphenoidal excision of sellar/ suprasellar lesion.

Histopathological examination of the lesion showed elongate bipolar spindle cells arranged in fascicular and storiform pattern. The tumor cells had eosinophilic cytoplasm, indistinct cell borders with moderately sized oval to elongated nuclei. Mitotic figures were hard to find. A small portion of unremarkable anterior pituitary was also noted.

By immunohistochemistry, the neoplastic cells were diffusely positive for S100 protein, vimentin and showed nuclear staining for TTF-1. Neoplastic cells also showed variable staining for GFAP, BCL-2 and CD-56. They were negative for EMA, cytokeratin and neuroendocrine markers like synaptophysin and chromogranin. Ki-67 proliferation index was less than 1%. Based on morphologic findings and immunohistochemistry, a diagnosis of pituicytoma was rendered.

Sundeep Mani, Aarchana Lakshmanan, Annapurneswari Subramanyan diagnostic pathology 2022, 7:284 ISSN 2364-4893
DOI: http://dx.doi.org/10.17629/www.diagnosticpathology.eu-2022-7:284

Discussion

Pituicytomas are classified as WHO grade I tumors and are usually clinically indolent. These rare glial tumors presumably arise from pituicytes and are found in neurohypophysis and infundibulum of the pituitary gland[2]. Pituicytomas are recognised as a separate lesion distinct from other lesions like granular cell tumor of the pituitary and pilocytic astrocytoma of neurohypophysis [3][7]. Peak incidence of these tumors is in adults but can occur in a wide age range (seven years to eighty three years). There is a slight male predominance. Clinical features are due to endocrine dysfunction (amenorrhea, infertility, diabetes insipidus, panhypopituitarism) or due to optic chiasma compression. Some cases can be asymptomatic and found incidentally[6][8]. Our patient presented with features of visual disturbances due to optic chiasma compression.

Radiographic features of pituicytomas on CT are homogenously enhancing mass either within the pituitary fossa or in the suprasellar region with variable size ranging from few millimetres to a few centimetres. Most of the reported cases are isointense on T1 weighted images and hyperintense on T2 weighted images, with marked homogenous enhancement after gadolinium enhancement. Heterogenous enhancement, calcification and cystic changes are rarely seen[4][10]. Our case showed an extra axial cystic lesion compressing the optic chiasma.

Histologically these tumors are composed of round to spindle shaped cells arranged in a fascicular or storiform pattern. The cells have an abundant eosinophilic cytoplasm with round to oval nuclei, without evident atypia or mitotic figures and rich capillary network is evident in many cases. TTF-1 is most reliable immunostain for pituicytoma diagnosis with variability in staining for other markers like GFAP, EMA, S-100, SSTR2A and synaptophysin[1].

Pituicytoma, spindle cell oncocytoma and granular cell tumor represent rare but important differential diagnostic considerations of sellar and suprasellar lesions. These tumors have overlapping clinical and radiographic findings but with varying morphologic features[2]. Granular cell tumors are composed of dense, polygonal cells with small nuclei and abundant granular, eosinophilic, PAS +ve and diastase resistant cytoplasm. Spindle cell oncocytomas are composed of fascicles of spindled or epithelioid cells with variably oncocytic cytoplasm. The positivity on EMA immunostain helps in distinguishing spindle cell oncocytoma from pituicytomas and granular cell tumor which are generally EMA negative. Nuclear expression of TTF-1 in all three entities suggest that spindle cell oncocytoma, granular cell tumor and pituicytoma probably constitute a spectrum of single nosological entity with varied morphology[6]. Absence of Rosenthal fibres and eosinophilic granular bodies helps to distinguish

Sundeep Mani, Aarchana Lakshmanan, Annapurneswari Subramanyan diagnostic pathology 2022, 7:284 ISSN 2364-4893
DOI: http://dx.doi.org/10.17629/www.diagnosticpathology.eu-2022-7:284

pituicytomas from pilocytic astrocytomas. Absence of spindle cells in Antoni A or B pattern and Verocay bodies differentiates pituicytomas from Schwannoma which are also S-100 positive[6][9].

There are no defined genetic signature for pituicytomas and few attempts on molecular testing have shown that these tumors are negative for IDH and BRAF mutations, which are the common changes seen in low grade brain tumors. One study has demonstrated tumors with loss of chromosome arms 1p, 14q, 22q and overrepresentation of chromosome arm 5p[1][5]. Viaene *et al*. have shown MAPK activation in pituicytomas using Next generation sequencing in their study[1].

Pituicytomas are slowly enlarging localized tumors which are amenable to complete surgical resection. Recurrences are rare, which are probably due to incomplete removal of tumour. No instances of malignant transformation or distant metastasis have been reported till date. Accurate assessment of prognosis needs follow up in a larger cohort[4].

Conclusion

Pituicytomas are rare tumors of the sellar and suprasellar region and they typically present with compression symptoms. Diagnosis is typically based on histopathological examination with a varied immunohistochemical profiles but with consistent TTF-1 positivity. The tumor is benign, slow growing and amenable to surgical resection.

Sundeep Mani, Aarchana Lakshmanan, Annapurneswari Subramanyan diagnostic pathology 2022, 7:284 ISSN 2364-4893
DOI: http://dx.doi.org/10.17629/www.diagnosticpathology.eu-2022-7:284

Figures

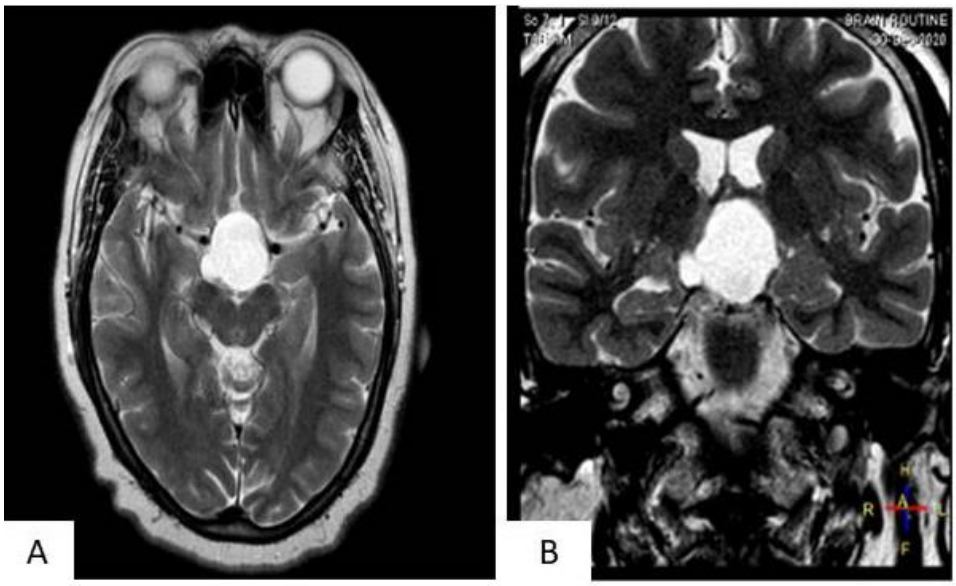

Fig 1- A and B shows MRI of the brain with a cystic lesion in sella extending to suprasellar cistern with mass effect over the optic chiasma and third ventricle

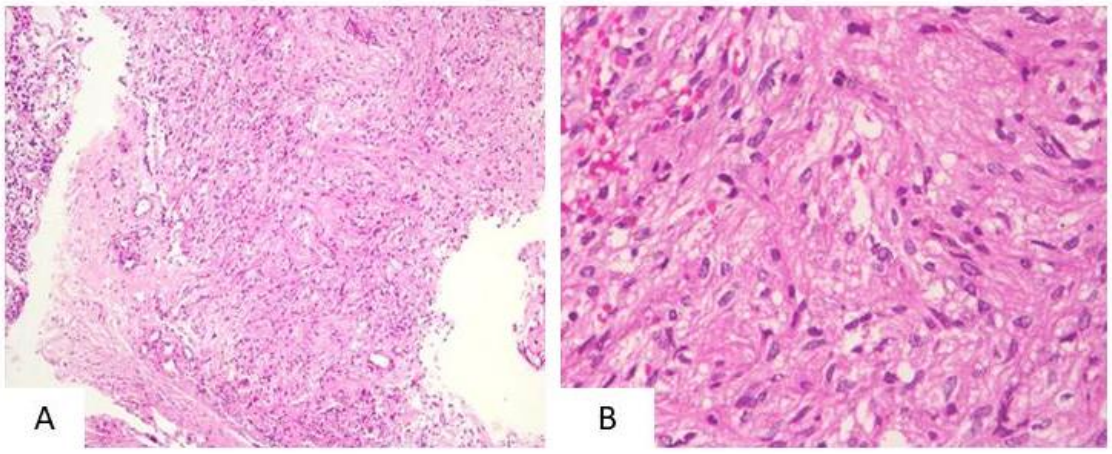

Fig 2- A (H&E- 100X) and B (H&E- 400X) shows spindle cells arranged as fascicles

Sundeep Mani, Aarchana Lakshmanan, Annapurneswari Subramanyan diagnostic pathology 2022, 7:284 ISSN 2364-4893
DOI: http://dx.doi.org/10.17629/www.diagnosticpathology.eu-2022-7:284

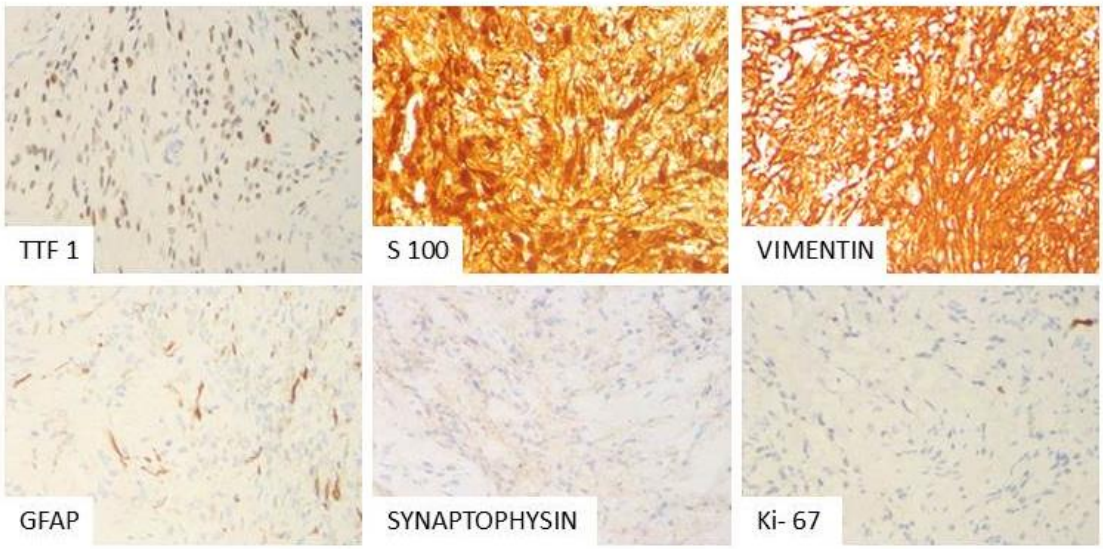

Fig 3- The neoplastic cells diffusely positive for TTF 1, S 100, vimentin, variably positive for GFAP, negative for synaptophysin and Ki-67 was <1%.

Reference

1. Viaene, Angela N., et al. "Histologic, Immunohistochemical, and Molecular Features of Pituicytomas and Atypical Pituicytomas." *Acta Neuropathologica Communications*, vol. 7, no. 1, Dec. 2019, p. 69. *DOI.org (Crossref)*, https://doi.org/10.1186/s40478-019-0722-6.
2. Ulm, Arthur J., et al. "Pituicytoma: Report of Two Cases and Clues Regarding Histogenesis." *Neurosurgery*, vol. 54, no. 3, Mar. 2004, pp. 753–58. *DOI.org (Crossref)*, https://doi.org/10.1227/01.NEU.0000108983.50966.B7.
3. Brat, Daniel J., et al. "Pituicytoma: A Distinctive Low-Grade Glioma of the Neurohypophysis." *The American Journal of Surgical Pathology*, vol. 24, no. 3, Mar. 2000, pp. 362–68. *DOI.org (Crossref)*, https://doi.org/10.1097/00000478-200003000-00004.
4. Yang, Xiaoyu, et al. "Pituicytoma: A Report of Three Cases and Literature Review." *Oncology Letters*, vol. 12, no. 5, Nov. 2016, pp. 3417–22. *DOI.org (Crossref)*, https://doi.org/10.3892/ol.2016.5119.
5. Chakraborti, S., et al. "Pituicytoma: Report of Three Cases with Review of Literature." *Pathology - Research and Practice*, vol. 209, no. 1, Jan. 2013, pp. 52–58. *DOI.org (Crossref)*, https://doi.org/10.1016/j.prp.2012.10.006.

Sundeep Mani, Aarchana Lakshmanan, Annapurneswari Subramanyan diagnostic pathology 2022, 7:284 ISSN 2364-4893
DOI: http://dx.doi.org/10.17629/www.diagnosticpathology.eu-2022-7:284

6. Figarella-Branger, D., et al. “Pituicytomas, a Mis-Diagnosed Benign Tumor of the Neurohypophysis: Report of Three Cases.” *Acta Neuropathologica*, vol. 104, no. 3, Sept. 2002, pp. 313–19. *DOI.org (Crossref)*, https://doi.org/10.1007/s00401-002-0557-1.
7. Mao, Zhigang, et al. “Pituicytoma: Report of Two Cases.” *Oncology Letters*, vol. 2, no. 1, Jan. 2011, pp. 37–41. *DOI.org (Crossref)*, https://doi.org/10.3892/ol.2010.209.
8. Hammoud, Dima A., et al. “Magnetic Resonance Imaging Features of Pituicytomas: Analysis of 10 Cases.” *Journal of Computer Assisted Tomography*, vol. 34, no. 5, Sept. 2010, pp. 757–61. *DOI.org (Crossref)*, https://doi.org/10.1097/RCT.0b013e3181e289c0.
9. Nakasu, Yoko, et al. “Pituicytoma-Two Case Reports-: —Two Case Reports—.” *Neurologia Medico-Chirurgica*, vol. 46, no. 3, 2006, pp. 152–56. *DOI.org (Crossref)*, https://doi.org/10.2176/nmc.46.152.
10. Uesaka, M. Miyazono, S. Nishio, T., T. “Astrocytoma of the Pituitary Gland (Pituicytoma): Case Report.” *Neuroradiology*, vol. 44, no. 2, Feb. 2002, pp. 123–25. *DOI.org (Crossref)*, https://doi.org/10.1007/s002340100654.