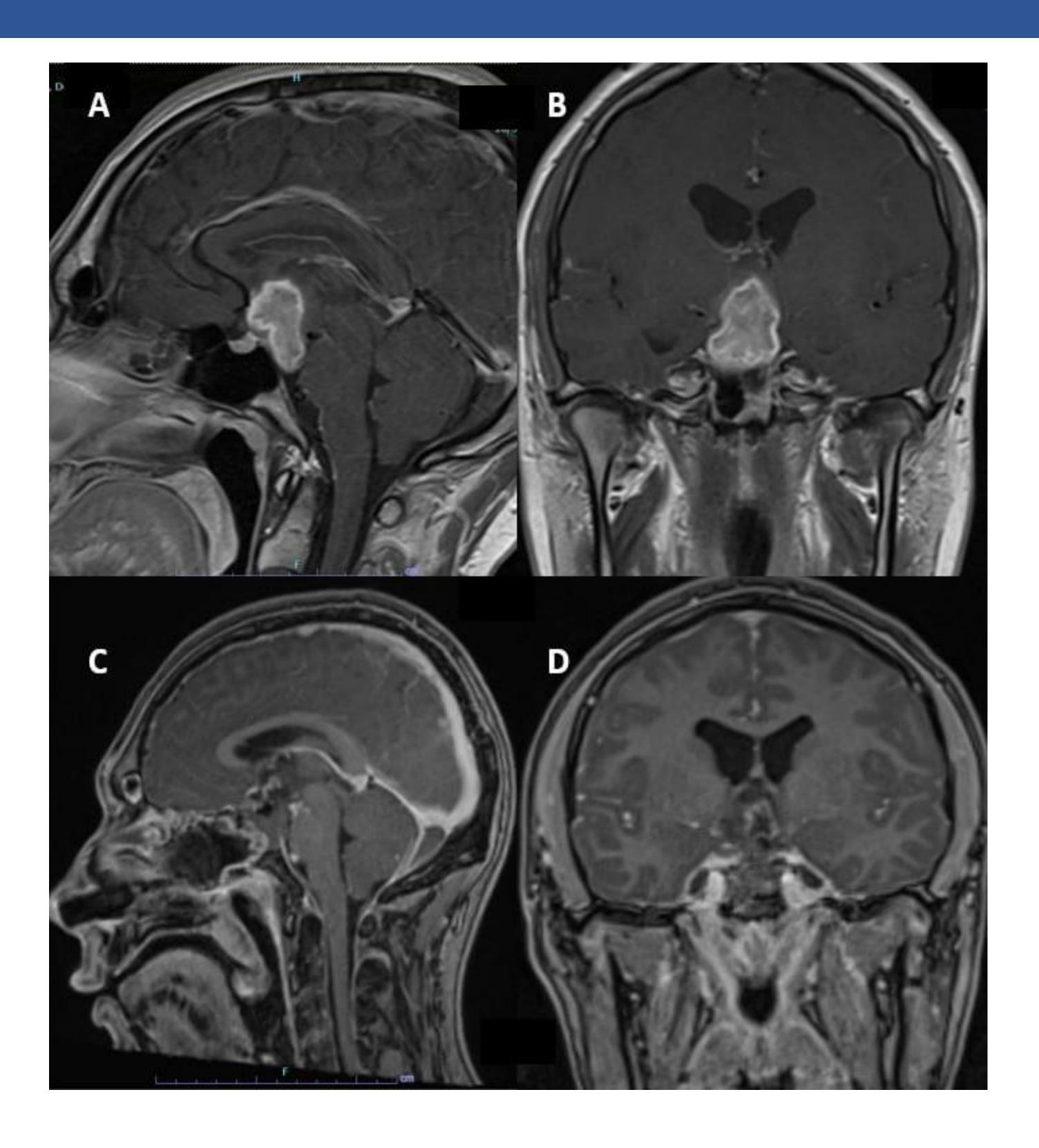
Ectopic Pituitary Neuro-endocrine Tumors: A case report and Literature Review

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Background

Ectopic pituitary neuro-endocrine tumor (EPNT) are extremely rare pituitary neuroendocrine tumors located outside the sella turcica without any direct connection to the intra-sellar pituitary gland. We present a case and provide a literature review on this rare condition.

Methods and Materials

A twenty-one-year-old male presented with complaints of headaches. MRI revealed a heterogeneously enhancing mass in the suprasellar region posterior to the pituitary gland extending along the prepontine cistern with calcifications and blood products with mild ventriculomegaly (Figure 1 A and B). There were no endocrine or visual abnormalities. The tumor was favored to be a craniopharyngioma.

We performed an endoscopic endonasal trans-sphenoidal, trans-tuberculum and trans-clival approach for resection of the tumor (Figure 1 C and D). The patient recovered uneventfully from surgery with improvement in headaches. The final pathology was null cell.

Literature Review

EPNTs, more commonly referred to as ectopic pituitary adenoma in the literature, are believed to arise from a disorder of the migration of the anterior lobe cells of the pituitary resulting in the final location outside the sella turcica, but the exact pathology is unknown. Our literature reviewed yielded a total of 180 cases described in the literature. Such tumors are more commonly located in the sphenoid sinus, followed by supra-sellar region, clivus and the cavernous sinus. Most EPNTs are hormonally active, with ACTH being the most common secreted hormone.

EPNTs are also more likely to be greater than 10 mm in size. The most commonly reported presenting symptoms are from hormonal over secretion, particularly ACTH over secretion. Headaches are also commonly reported. Based on tumor location, visual disturbances, oculomotor dysfunction and hearing loss are also possible. Nasal congestion, epistaxis and rhinorrhea, especially from a sphenoid location, have also been reported.

Conclusions

EPNT is a rare but important differential for a mass along the migration path of anterior pituitary lobe cells and warrants high clinical suspicion for effective diagnosis and treatment.

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