CLINICAL VIGNETTE

3rd Nerve Palsy: Rare Presentation Related to Secondary Hyperparathyroidism

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Case Report

A 37 y/o R handed woman with history of ESRD secondary to HTN/Preeclampsia s/p renal transplantation in 2001 with allograft failure in 2005 on Hemodialysis, with secondary hyperparathyroidism presents to the ER with headaches and vision changes for 3 weeks. The patient was seen by Neurology who noted horizontal diplopia and ptosis consistent with a complete left 3rd nerve palsy. Fundoscopic examination showed bilateral papilledema. No other abnormalities were noted on examination. On further history the patient denied fevers, chills, neck stiffness, nausea or vomiting. She had been on dialysis intermittently for a total of 16 years. She reported no current problems with hemodialysis but vaguely recalled being told there was "something wrong" with her parathyroid gland.

Labs and Imaging

A diffusion weighted MRI showed a sub acute stroke in the anterior limb of the left internal capsule along with superior ophthalmic vein engorgement on the left. The differential was some sort Cavernous sinus pathology which could include infectious, compressive, cavernous sinus thrombosis, cavernous sinus arteriovenous fistula or a vasculitic process. Additional testing is summarized in Table 1, which included an unremarkable lumbar puncture including a normal opening pressure. There was consideration for MRI with gadolinium to further assess the cavernous sinus anatomy but given the risk for nephrogenic systemic fibrosis this was deferred and an IR Cerebral Angiogram was obtained. The IR Cerebral Angiogram ruled out a cavernous venous thrombosis and fistula but was suggestive of hyperostosis leading to a partial outflow obstruction of the L Superior Ophthalmic Vein. The PTH which had been pending prior to the Angiogram returned > 1700 pg/ dl (range 11-51 pg/dl) suggesting that the hyperostosis is secondary to long standing and untreated secondary hyperparathyroidism leading to Renal Osteodystrophy.

Table 1

Labs within normal limits	Protein C Protein S Factor VIII HepC HepB Ab+ HepB An thyroperoxidase Ab RF ANA Crypto ab csf Thyroglobulin antibody ATIII Cardiolipin Beta 2 glycoprotein CSF labs: Final acid fast, bacteria and fungal cultures Histo, toxo, ebv, cmv, vzv, hsv Flow cytometry and cytology
Elevated/Abnormal labs	ESR PTH
Decreased	Vitamin D 25

Initial Treatment Course

Once renal osteodystrophy became the primary diagnosis the patient was started on high dose methylprednisolone in hopes of reducing Cavernous sinus pressures and improve CN III function. Because the patient had cranial nerve palsy for three weeks, steroids were started rather than surgical compressive therapy as the risks likely outweighed the possible benefit.

Given the untreated secondary hyperparathyroidism, renal osteodystrophy and cranial hyperostosis, parathyroidectomy was recommended. Endocrine, Endocrine surgery, Neurology and the Nephrology service concurred to fully replace Vitamin D 25, monitor for improvements in the PTH, and have the

patient return for parathyroidectomy to minimize risk for Hungry Bone syndrome post parathyroidectomy. There was little improvement in the CN III palsy on steroids.

Discussion

Renal osteodystrophy is a broad term related to the consequences of the abnormal bone and mineral metabolism that progressively worsens as CKD progresses to ESRD. The term Chronic Kidney Disease-Mineral and Bone Disease (CKD-MBD) has been coined by KDIGO as an all-encompassing term that refers to abnormalities of calcium, phosphorus, vitamin D, PTH, bone turnover, mineralization's and extraskeletal calcification. Secondary hyperparathyroidism is an essential part of the pathogenesis of CKD-MBD as the secretion of PTH becomes pathologically elevated secondary to phosphorus retention, decreased free ionized calcium concentration, decreased calcitriol (vitamin d 1, 25) production, and FGF-23 abnormalities all of which interact to perpetuate secondary hyperparathyroidism[1].

The theoretical risk of cranial nerve palsies secondary to Renal osteodystrophy have been described in both animal studies and case reports[2-4]. We describe a case of severe secondary hyperparathyroidism left untreated leading to calvarial/petrosal hypertrophy with a CN III palsy as a consequence. A previous case report described successful surgical decompression after the diagnosis of CN III palsy from hyperostosis in a patient who received prompt surgical decompression[4].

Conclusion

Secondary hyperparathyroidism is a complex and prevalent disorder in CKD which if left untreated can lead to a wide variety of pathologies including increased fractures and abnormal extraskeletal calcification. Hyperostosis/Renal Osteodystrophy should remain in the differential of cranial nerve palsies especially in the ESRD population. When this condition is recognized, prompt treatment with medical or surgical decompression is recommended.

REFERENCES

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