AMERICAN ACADEMY OF OTOLARYNGOLOGY–HEAD AND NECK SURGERY FOUNDATION

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DOI: 10.1177/2473974X18792488
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Esthesioneuroblastoma Isolated to the Maxillary Sinus Antrum Presenting as SIADH: A Case Report

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No sponsorships or competing interests have been disclosed for this article.

Keywords

esthesioneuroblastoma, olfactory neuroepithelium, sinonasal tumor, SIADH, paraneoplastic syndrome, endoscopic sinus surgery

Received February 16, 2018; revised May 9, 2018; accepted July 13, 2018.

Esthesioneuroblastoma is a rare neoplasm typically arising from olfactory neuroepithelium of the superior nasal cavity.[1,2] Esthesioneuroblastoma demonstrates bimodal age distribution, with peaks in the second to third, as well as sixth to seventh, decades of life.[3] Patients often complain of nonspecific sinonasal symptoms, including unilateral nasal obstruction, headache, and epistaxis.[2] Esthesioneuroblastoma represents 3% of all sinonasal tumors; however, its prevalence is likely underestimated due to delayed recognition.[2,3] Atypical clinical features arise in rare cases of ectopic disease loci or secondary to paraneoplastic syndromes.[1] We present a unique case of syndrome of inappropriate antidiuretic hormone (SIADH) secondary to esthesioneuroblastoma arising from, and completely isolated to, the maxillary sinus antrum.

Case Report

A 28-year-old woman presented to our rhinology clinic in Vancouver, Canada. She had a 6-month history of nasal congestion, postnasal discharge, and headache, as well as a history of persistent hyponatremia refractory to oral sodium chloride tablets and water restriction. She denied epistaxis or hyposmia. She had significant fatigue and weakness requiring 2 hospital admissions for electrolyte correction. Laboratory investigations revealed a plasma sodium concentration of 118 mmol/L and urine osmolality of 810 mOsm/kg. Thyroid-stimulating hormone, morning cortisol, and serum protein electrophoresis all returned within normal limits. She underwent full-body computed tomography (CT), head CT, and magnetic resonance imaging of the head, which demonstrated a homogenously enhancing mass within the right maxillary sinus (**Figure 1**). There was no pituitary abnormality identified. Positron emission tomography (PET)–CT demonstrated a 2-cm mixed-attenuated lesion isolated to the right maxillary sinus. There was no evidence of metastatic disease. The patient subsequently underwent endoscopic image-guided sinus surgery and complete excision of the mass originating from the right maxillary sinus and involving the uncinate process.

Histopathology confirmed a diagnosis of Hyams grade 1 esthesioneuroblastoma, with clear margins and no lymphovascular invasion (**Figure 2**). Normalization of the patient's hyponatremia occurred within 24 hours postoperatively and was sustained at the 6-month assessment, with considerable improvement in energy levels. Sinonasal symptoms resolved within 3 months postoperatively.

The patient underwent a 4-week course of postoperative adjuvant radiotherapy. She received 6000 cGy delivered over 25 fractions, followed by 5000 cGy delivered over 25 fractions. She has remained asymptomatic from both a sinonasal and a metabolic perspective and regularly undergoes endoscopic surveillance for tumor recurrence.

Discussion

SIADH is a common cause of euvolemic hyponatremia characterized by inappropriately elevated levels of arginine

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This article was presented at the annual meeting of the American Rhinologic Society; September 16-17, 2016; San Diego, California.

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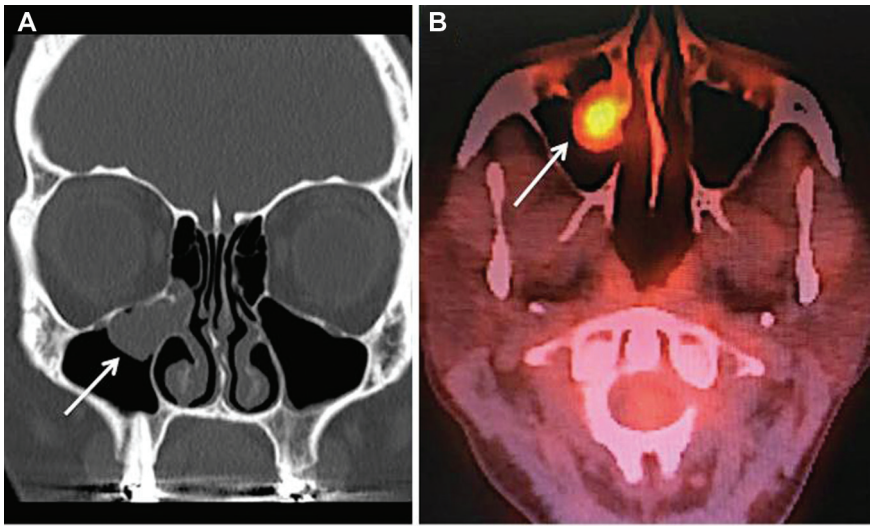

Figure 1. (A) Preoperative coronal computed tomography paranasal sinuses and (B) axial positron emission tomography–computed tomography demonstrating a single enhancing mass isolated to the right maxillary sinus antrum (arrows) with no radiologically identifiable extension into skull base.

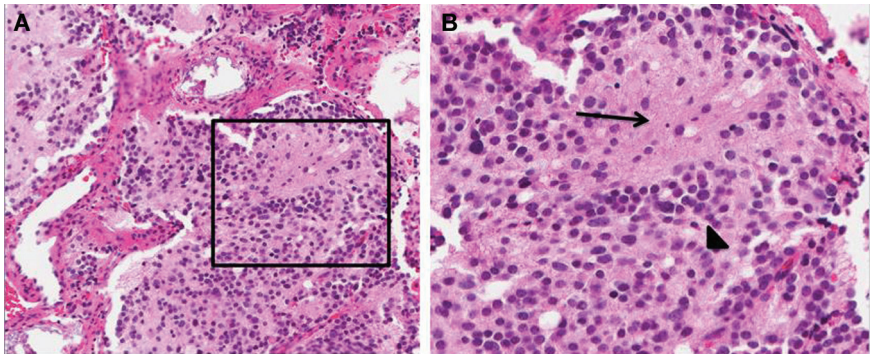

Figure 2. (A) Histopathology slide of right maxillary sinus mass with square inlay enlarged (B, right). Features characteristic of esthesioneuroblastoma are seen, including prominent neuropil production (arrow) and rare Homer-Wright rosettes (arrowhead).

vasopressin.[4] The diagnostic criteria for the diagnosis of SIADH include the presence of hyponatremia and hypoosmolality in an euvolemic patient, inappropriate urinary concentration of sodium (urine osmolality >100 mOsm/kg), elevated urine sodium excretion with normal intake of salt and water, and exclusion of other potential causes of hyponatremia, including adrenal failure, hypothyroidism, and recent diuretic use.[4] SIADH occurs in 3% of head and neck cancers.[5]

Esthesioneuroblastoma is thought to originate from the olfactory neuroepithelium of the superior turbinate, the cribriform plate, and along the superior septum.[2] There is a 2% prevalence of esthesioneuroblastoma-associated SIADH in the literature, with most patients initially presenting with signs secondary to hyponatremia.[1] The literature does report cases of SIADH-associated esthesioneuroblastoma arising from the maxillary sinus, osteomeatal unit, and middle turbinate; however, these cases were all associated with secondary extension into the nasal cavity, skull base, or orbit. Our case is unique, as the esthesioneuroblastoma was completely isolated to the maxillary sinus and demonstrated no visible extension into the skull base endoscopically or radiologically. Gabbay et al suggested significant resolution of SIADH posttreatment of esthesioneuroblastoma, with consideration of incomplete tumor resection in cases of failed return to normal osmoregulation within days posttreatment.[1]

Surgical management is considered mainstay for treatment of esthesioneuroblastoma.[5] Kane et al performed a systematic review analysis of 700 patients, which suggested no additional survival benefit with adjuvant radiotherapy.[5] The radiographic Kadish staging system and the Hyams histopathologic grading system have been used independently or in concert to evaluate patients' prognoses. Kane et al determined that Hyams grade 3 and 4 tumors independently predicted poor survival among patients with esthesioneuroblastoma (35% vs 86% grade 1 and 2).[5]

Conclusion

Given the potential consequences and morbidity associated with delayed recognition or inappropriate management, SIADH-associated esthesioneuroblastoma should be an early consideration for

patients presenting with hyponatremia of undefined etiology, and suspected patients should be questioned regarding concurrent sinonasal symptoms.

Author Contributions

Alysha Rasool, interpretation, manuscript draft editing; **Anali Dadgostar**, interpretation, manuscript draft editing; **Jamil Manji**, interpretation, manuscript draft editing; **Fahad Al-Asousi**, interpretation, manuscript draft editing; **Amin Javer**, interpretation, manuscript draft editing.

Disclosures

Competing interests: None.

Sponsorships: None.

Funding source: None.

References

1. Gabbay U, Leider-Trejo L, Marshak G, et al. A case and a series of published of esthesioneuroblastoma in which long-standing paraneoplastic SIADH had preceded ENB diagnosis. *Ear Nose Throat J*. 2013;92:E6.
2. Senchack A, Freeman J, Ruhl D, et al. Low-grade esthesioneuroblastoma presenting as SIADH: a review of atypical manifestations. *Case Rep Otolaryngol*. 2012;2012:1-6.
3. Lund VJ, Howard D, Wei W, et al. Olfactory neuroblastoma: past, present, and future? *Laryngoscope*. 2003;113:502-507.
4. Hannon MJ, Thompson CJ. The syndrome of inappropriate antidiuretic hormone: prevalence, causes and consequences. *Eur J Endocrinol*. 2010;162:S5-S12.
5. Kane AJ, Sughrue ME, Rutkowski MJ, et al. Posttreatment prognosis of patients with esthesioneuroblastoma. *J Neurosurg*. 2010:113:340-351.