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Case Report

Case of nasopharyngeal metastases presenting with multiple atypical cranial nerve deficits

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ABSTRACT

Jugular foramen syndrome is an uncommon condition with a wide range of possible causes. We present a rare case of Villaret syndrome as an example of jugular foramen syndrome and illustrate a clinical-radiological approach for diagnosing jugular foramen syndrome. The morphological similarities between mesenchymal tumors such as rhabdomyosarcoma and phyllodes seen in our patient make immuno-histochemical (IHC) characterization essential for a definitive diagnosis. Jugular foramen syndromes, including Vernet syndrome, Collet Sicard, Villaret, Tapia, Jackson, and Schmidt, can manifest with multiple cranial nerve deficits in contiguous areas. In our case, the patient exhibited palate paralysis, pharyngeal anesthesia, and Horner's syndrome, thereby highlighting the differential diagnoses of Villaret syndrome and Tapia syndrome.

Keywords: Jugular foramen syndrome, Vernet syndrome, Multiple cranial nerve palsy

INTRODUCTION

Jugular foramen syndrome is a rare entity and can have a diverse etiology.[1] Presentation includes multiple but variable cranial nerve deficits, including loss of taste, loss of sensation in the posterior third of the tongue, paralysis of the vocal cords and palate, sensory loss of the pharynx and larynx, weakness of the trapezius and sternocleidomastoid, tongue paralysis and atrophy and Horner's syndrome. Cranial nerve deficits can be variable and hard to interpret in jugular foramen syndromes. Determining the underlying cause of the deficits through laboratory and neuroimaging can be challenging. We illustrate a case of Villaret syndrome and provide a clinical-radiological algorithm for diagnosing jugular foramen syndromes.[1-3]

CASE REPORT

A 48-year-old Nepali female presented with a unilateral throbbing headache for three months, peaking to an intensity of VAS 7/10 within five hours from its onset. She had pain on the left side of the face with decreased sensation to light touch for the past two and a half months. She also complained of a decreased sense of smell not localized to any nostril, blurring of vision in her left eye, and restricted outward right eye movement for two and a half months. She also complained of an inability to move her tongue and decreased hearing from the left ear. Other complaints included a soft speech and difficulty swallowing liquids for one month. There was no history of head injury, neurosurgery, loss of consciousness, limb weakness, facial deviation, sensory loss, alteration in bowel and bladder functions, cognitive disturbances, or recent history of COVID-19.

She underwent resection of a breast lump diagnosed with a phyllodes tumor two years prior, which was characterized by focal stromal cellularity, mitotic activity of five per high power field (out of 10), and stromal overgrowth with the closest surgical margin at 0.5 cm. IHC markers were unknown. Axillary nodes, NAC, and remaining margins were free of tumors.

On physical examination, the patient was conscious and cooperative with the Glasgow coma scale 15. A cranial nerve examination of cranial nerve I with tape revealed a complete loss of sensation of smell. Visual acuity, visual field, and color vision were normal. The left pupil was 2.5 mm in size and reactive, the right pupil was 4 mm in size and reactive, and the anisocoria was more in bright ambient light. Left eye lateral palsy was noted on extraocular muscle examination. The corneal reflex was present on the right side and absent on the left side. No facial deviation was noted. The tongue and uvula deviated to the left, and the gag reflex was absent. Weber's test was non-lateralizing; bone conduction was more than air conduction bilaterally. No gait abnormalities were noted. No sensory or motor abnormalities were recorded, and deep tendon reflexes were +2 in all four limbs.

Differential diagnoses were neuroborreliosis, cranial multineuritis/neuropathy, cranial nerve malignancy, encephalomyeloneuritis with immune checkpoint inhibitor chemotherapy, neuro-vascular compression syndromes such as internal carotid artery (ICA) pseudoaneurysm, dissection, fibromuscular dysplasia of ICA, skull base tumor/metastases, neuralgic amyotrophy, radiation-induced neuropathy, neurosarcoidosis, rhombencephalitis, Vernet syndrome (jugular foramen syndrome), collet sicard syndrome (condyle-jugular syndrome/Vernet syndrome with XII cranial nerve involvement), Villaret syndrome (collet sicard syndrome associated with Horner's syndrome), eagle's syndrome, parapharyngeal abscess/tumors, cerebellopontine angle tumor, brainstem stroke, viral illness, jugular phlebitis and connective tissue diseases such as polyarteritis nodosa, Ehlers–Danlos syndrome, and Marfan's syndrome.

Hemogram, complete metabolic profile, lipid profile, cerebrospinal fluid analysis, serum ANA and ACE levels were within normal limits. Brain magnetic resonance imaging (MRI revealed a large osteolytic destructive lesion involving the left petrous temporal bone, predominantly the apex, and significant adjacent soft tissue swelling with central necrosis and peripheral rim enhancement (Figures 1 and 2). Intracranially, a mass was seen extending to the left of the sela turcica with effacement of the left cavernous sinus reaching posteriorly to the prepontine cistern. There was encasement of the left petrous and cavernous ICA with luminal narrowing. PET CT revealed a large heterogeneously enhancing soft tissue mass lesion in the nasopharynx with involvement of the bone and upper deep cervical nodes and no signs of recurrence at the chest wall or mastectomy site. Chest HRCT revealed two discrete nodules bilaterally in the lower lung lobes. Laryngoscopy was nonrevealing of any pathology. Histo-pathological examination of the lesion revealed a high-grade mesenchymal tumor suggestive of rhabdomyosarcoma. IHC markers were not placed on the tumor.

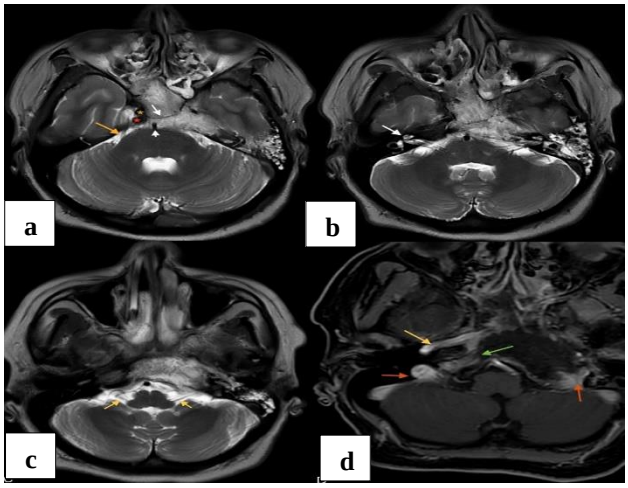

Figure 1: (a) This is a T2-weighted MR venogram of the brain in axial section. The white arrow shows tumor infiltrating the sela tercica and the yellow arrow shows normal right trigeminal nerve. The red and yellow arrows depict right ICA and ECA respectively, compare all structures to the left side. White arrow head shows patent basilar artery; (b) the white arrow shows right vestibulofacial facial nerve, compare with left side where tumor has infiltrated the nerve. The abducens nerves are not visualized because of tumor infiltration, (c) the glossopharyngeal and vagus nerve shown exiting the brainstem on both sides, and (d) the red arrow indicated the sigmoid sinus on the right and left side. The yellow arrow is the ICA. The green arrow is the inferior petrosal sinus. Compare the course of glossopharyngeal and vagus nerves with respect to jugular bulb on right side. Tumor has completely distorted the jugular anatomy.

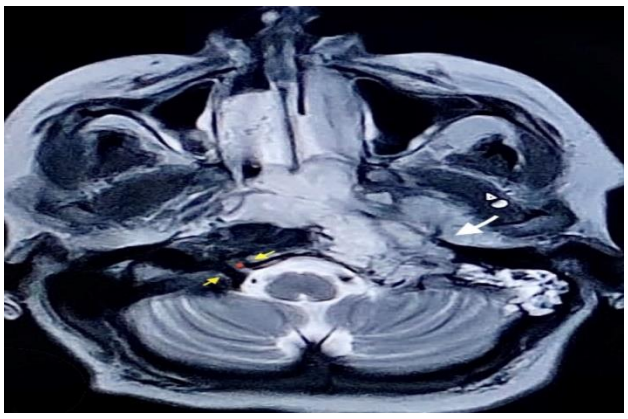

Figure 2: This is a T2-weighted MR venogram of the brain in axial section. The right hypoglossal leaving the hypoglossal foramen. Compare with left side.

She was diagnosed with skull base osseous metastases involving the left cranial nerves I, V, VI, IX, and XII and the sympathetic plexus around the ICA.

She received vincristine, actinomycin D, and cyclophosphamide (VAC regimen), i.e., the first chemotherapy cycle during her hospital stay. She was discharged in stable condition on medication advice of painkillers, anti-reflux, and antiemetics and to follow up with an oncology outpatient with serial hemograms. The need for a speech therapist and placement of a nasogastric tube for feeding was assessed at serial intervals, and the decision was based on the patient's own expected functional recovery.

DISCUSSION

The prevalence of brain metastasis in breast cancer ranges between 20-40% (BCBM). In some small studies, the incidence of metastases in phyllodes tumors ranges from 10.8-12.2%. The rate of brain metastases in phyllodes tumors is meager. There is one case in the literature similar to ours regarding BCBM in phyllodes tumors of the breast.[1-4] Our patient, in contrast, is a probable case of dual malignancy. Rhabdomyosarcoma and phyllodes are mesenchymal tumors that are morphologically similar and, for definitive diagnosis, need IHC characterization, which was not available in our patient.

Jugular foramen syndromes such as Vernet syndrome, Collet Sicard, Villaret, Tapia, Jackson, and Schmidt can present with multiple contiguous cranial nerve deficits. In our case, the patient presented with paralysis of the palate and anesthesia of the pharynx along with Horner's syndrome, which makes Villaret syndrome and Tapia syndrome different.[5-8] The hypoglossal and glossopharyngeal nerves pass through the space between the transverse process of the atlas and the styloid process, which is populated by tumor cells, as seen on MRI. Involvement of the occipital condyle, jugular foramen, and hypoglossal foramen by inflammatory exudates, tumors, and local tissue trauma can lead to similar presentations. Due to Horner's syndrome, patients with Collet Sicard syndrome and Vernet syndrome were excluded.[9-11] Neurological recovery is slow, and residual deficits may persist for a long time.

CONCLUSION

In this report, we describe a unique occurrence of Villaret syndrome in a patient with dual malignancy as an illustration of jugular foramen syndrome. We also outline a clinical-radiological strategy for diagnosing jugular foramen syndromes. It is important to consider jugular foramen syndrome as a potential diagnosis in all cases presenting with multiple cranial nerve deficits, regardless of the contiguous pattern of involvement, and conducting an MRI scan as an initial screening tool is recommended for all such cases.

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REFERENCES

1. Magistrelli L, Varrasi C, Stecco A, Carriero A, Cantello R. Villaret syndrome as clinical presentation of occult metastatic breast cancer. Neurol Sci. 2018;39(1):177-8.
2. Sellami M, Kallel S, Ben Ayed M, Mellouli M, Boudawara TS, Mnejja M, et al. Nasopharyngeal Metastasis from Breast Carcinoma: A Case Report and a Review of the Literature. Ear, Nose Throat J. 2022;01455613221119047.
3. Mituś JW, Blecharz P, Walasek T, Reinfuss M, Jakubowicz J, Kulpa J. Treatment of Patients with Distant Metastases from Phyllodes Tumor of the Breast. World J Surg. 2016;40(2):323-8.
4. Riebensahm C, Joosse SA, Mohme M, Hanssen A, Matschke J, Goy Y, et al. Clonality of circulating tumor cells in breast cancer brain metastasis patients. Breast Cancer Res. 2019;21(1):101.
5. Li Y, Song Y, Lang R, Shi L, Gao S, Liu H, et al. Retrospective study of malignant phyllodes tumors of the breast: Younger age, prior fibroadenoma surgery, malignant heterologous elements and surgical margins may predict recurrence. Breast. 2021;57:62-70.
6. Baek TW, Kang Y, Lee HJ. Improved Lesion Conspicuity with Contrast-Enhanced 3D T1 TSE Black-Blood Imaging in Cranial Neuritis: A Comparative Study of Contrast-Enhanced 3D T1 TSE, 3D T1 Fast-Spoiled Gradient Echo, and 3D T2 FLAIR. Am J Neuroradiol. 2021;42(5):945-50.
7. De Gennaro R, Gastaldo E, Tamborino C, Baraldo M, Casula N, Pedrali M, et al. Selective cranial multineuritis in severe COVID-19 pneumonia: two cases and literature review. Neurol Sci. 2021;42:1643-8.
8. Halperin JJ. Neuroborreliosis. Am J Med. 1995;98(4):52S-9S.
9. Sheng S, Sharma R, Samant R, Yuan J, Nalleballe K, Kapoor N. Isolated primary neurolymphomatosis with cranial multineuritis: a case presentation. Neurol Sci. 2021;42:1223-6.
10. Beer FD, Post B. Teaching NeuroImages: Villaret syndrome. Neurology. 2010;75(9).
11. Garrett D Jr, Ansell LV, Story JL. Villaret's syndrome: a report of two cases. Surg Neurol. 1993;39(4):282-5.

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