Acta Scientific Otolaryngology

Volume 3 Issue 7 July 2021

Case Report

How Can an Attention-Grabbing and Thought-Provoking Case Keep the Mind Ticking for a Challenging and Perplexing Diagnosis???

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Received: May 28, 2021

Published: June 12, 2021

Abstract

When a patient complaints with various symptomatology, a "syndrome" like picture often runs through the mind. Here, is one such female patient who presented with an unusual and interesting case with multiple cranial nerve involvement in the areas of ear, nose and throat with its symptomatology ranging from tinnitus to trismus along with facial numbness to paranasal sinus tenderness. When all these symptoms amalgamate, the mind wanders and raises queries to plenty of differentials. Hence, approach to one such diagnosis is demanding.

Keywords: Syndrome; Cranial Nerve Palsy; Extrapulmonary Koch's; Cervical Lymphadenopathy

Introduction

Neurological presentation with isolated multiple cranial nerve palsy is common. It has a varied group of etio-pathogenesis which can be mainly classified into inflammatory, infectious and/or neoplastic origin. Dysfunction of the cranial nerves can occur due to the lesion all along their course from intrinsic brainstem to peripheral sequences [2]. Here is one such case mentioned below which presented with multiple cranial neuropathies.

Case Report

23 year old married female patient comes to ENT outpatient department with multiple ENT complaints since 15 days (about which is discussed later in this case report). Her history goes back to nearly 1 year prior to the present consultation. During her 5th month of gestation, she developed a left lateral neck swelling. This swelling was evaluated which turned out to be Extrapulmonary Koch's on cervical node biopsy examination. She completed 6 months of Anti-tubercular therapy and the swelling resolved after treatment. During this process, patient delivered comfortably and had been lactating during her course of anti-tubercular treatment. Patient was asymptomatic for 1 month following completion of anti-tubercular treatment and tested negative for Extrapulmonary Koch's.

After 1 month, patient further complaints of diffuse headache, fever on and off, facial numbness, painful trismus, dysphagia, odynophagia, vomiting, low-pitched voice, significant loss of weight (10 kg from the time of complaints), loss of appetite, loss of taste, tinnitus and decreased hearing in both ears, giddiness, nasal obstruction, cough with minimal expectoration, double vision, ptosis of right eye, difficulty in shrugging shoulders, difficulty in tongue protrusion and re-developed swelling in the left lateral side of the neck since > 4 weeks, which have begun one by one and initially mild in nature but has now aggravated and become worse at the

time of presentation. At this point, she visited me for consultation. Patient consulted at various places with no improvement in her condition. Patient by then had weaned off lactation. The baby is fine and doing well.

Clinical examination findings are as follows:

- General physical examination:
- Patient is anorexic, poorly built and nourished.
- Vitals (Pulse rate/Blood pressure/Respiratory rate/Saturation/Temperature): Within normal limits, Weight: 35 kgs at present.
- Pallor is present. No Icterus, Cyanosis, Clubbing, Generalised lymphadenopathy, Edema.
- Systemic examination:
- Cardiovascular system/Respiratory system/Per abdomen are all within normal limits. Central nervous system examination findings are mentioned below.
- Central nervous system:
- Cranial nerve examination: Multiple cranial nerve involved with palsy of [III, IV, V, VI, VIII, IX, X, XI, XII]. whose features as mentioned below:
- Examination of the face: Facial numbness present. Reduced sensation over the face especially fine touch but can appreciate painful stimuli.
- Eye examination: Black floaters present. Visual acuity: Double vision/Diplopia present. Horner's syndrome present. Right abductor palsy + Right ophthalmoparesis.
- Ear examination: Bilateral tympanic membrane intact. Tuning fork test with 512Hz suggestive of severe sensorineural hearing loss. Normal vestibular system. No signs of facial nerve palsy.
- Nose examination: DNS to left and caudal dislocation to right. Right inferior turbinate hypertrophy present. Hence, unable to negotiate the 0^0 endoscope upto the choanae. Tenderness present in all paranasal sinuses.
- Throat examination:
- Oral cavity/Oropharynx: Grade III-IV trismus, unable to protrude her tongue and restricted tongue movements.
- Larynx: Unable to visualise the laryngeal structures due to trismus.
- Neck examination: Swelling was round, nearly 3X3 cm noted in the left posterior triangle of the neck (Level-V) which was firm in consistency, regular margins and surface, skin pinchable over the swelling, mobile in both directions, non-tender most likely cervical lymph node. Laryngeal crepitus: present. Patient was unable to shrug her shoulders.

Radiological investigations were done earlier. MRI Brain was within normal limits. HRCT temporal showed soft tissue in epitympanum, mesotympanum encasing ossicles and extending into aditus ad antrum and in mastoid air cell system in both ears. Rest was normal. Incidental soft tissue noted in both sphenoid sinus.

Patient had been treated conservatively for these multiple complaints. On first and only follow up by the patient after this treatment, patient seemed better with treatment but not upto expected satisfaction. As the situation was little dicey, dubious and doubtful; this needed further evaluation. But the patient was not willing for any further evaluation and wanted to get better only on conservative line of treatment. She did not follow up after this probably tired of multiple hospital hopping.

Discussion

Isolated multiple cranial neuropathies are commonly encountered as a clinical entity. The evaluation of patients is prodigious due to a wide range of aetiologies as well as potential for distressing, devastating and disturbing neurologic outcomes. Both afferent and efferent connections of the cranial nerves traverse through meninges, subarachnoid space, bony structures of the skull and superficial soft tissues. As the cranial nerve nuclei are embedded within the brain stem, intra-axial pathologic process may present initially with only cranial nerve dysfunction [5,8].

There may be involvement of homologous nerves on both sides or different nerves on either the same or the contralateral side. In some instances, a group of nerves are involved in distinct anatomical region constituting discrete syndrome. Most of the literature constitutes only case reports or case series of etiology of multiple cranial neuropathies [6,9]. Therefore, I have come across 2 such studies which give a broad outlook over the various scenarios of

multiple cranial neuropathies. The study by Keane [3] among 979 cases over 34 years is one such study who presented with multiple cranial nerve palsy which is by far the largest reported retrospective study. Second, is the observational study by Mehta., *et al*. [1] among 54 patients over 3 years.

Extensive and progressive involvement of cranial nerves indicates possible diagnosis of malignant infiltration of meninges with confirmation either by biopsy or before autopsy [4,10]. In India, cranial nerve palsy occur in 1/3rd of cases as a result of tuberculous meningitis. The incidence and occurrence of cranial neuropathy is usually associated with poor outcome [1,3].

Multiple cranial nerve palsy is defined as involvement of 2 or more non-homologous cranial nerves [1,3]. The anatomical syndrome is classified based on clinical and imaging findings such as: Cavernous sinus, Orbital apex, Jugular foramen, Cerebello- pontine angle, Base of skull and Discrete cranial polyneuropathy syndromes.

The onset of illness is acute, sub-acute and chronic. Acute onset is the onset of symptoms within less than 7 days. Sub-acute onset is the onset of symptoms ranging between 8-30 days. Chronic onset is when onset of symptoms is more than 30 days [2].

The common syndromes involved with multiple cranial nerve involvement as per observational study by Mehta., *et al*. [1] infers as follows: Cavernous sinus syndrome (CSS) is defined as involvement of two or more cranial nerves: III, IV, V (V1, V2) or VI. Orbital apex syndrome (OAS) is defined as involvement of cranial nerves: III, IV, V (V1), VI in combination of II dysfunction. Jugular foramen syndrome (JFS) is defined as involvement of cranial nerves: IX/X along with XI or XII or both. Base of skull syndrome is defined as involvement of multiple cranial nerves that exit from the skull and meninges. Cerebello-pontine angle syndrome (CPAS) is defined as involvement of cranial nerves: V, VII, VIII. Remaining combinations of cranial nerve involvement is categorized as Discrete multiple cranial nerve neuropathy (DMCNN).

According to the study by Mehta., *et al*. [1] etiology for cranial nerve involvement are as follows: Infections, Neoplastic, Idiopathic, Tolosa-Hunt syndrome and Vascular cause with no Traumatic or Iatrogenic cause. Among infections, most common is Bacterial (Tuberculous) and then Fungal (*Mucor* and *Aspergillus*). Diabetes Mellitus is the risk factor with respect to fungal etiology [3]. Infections form the most common etiology as the prevalence is higher in developing countries like India than any other developed country. Tuberculous meningitis is a common cause of multiple cranial nerve palsy in India. In Western countries, Neoplastic cause (Tumor) forms the most common etiology. Among neoplastic origin, spread of the tumors are usually primary, secondary and locally. Among neoplastic tumors, most common tumors belong to Intra-axial group (Glioma) and the rest belongs to extra-axial group which includes (Schwannoma, Meningioma, Chordoma, Lymphoma, Nerve sheath tumor, Epidermoid cyst). In metastasis, there are 3 groups: primary, multiple and unknown primary. Both CSS and OAS cause acute illness and are due to Mucor and Tuberculosis with risk factor as DM of 15% each. JFS causes chronic illness. Imaging in fungal patients shows Pansinusitis with multiple areas of focal destruction, heterogenous signal intensity, contrast enhancement of orbit/PNS/Cavernous sinus. THS: cause for painful ophthalmoplegia with unknown etiology. On MRI, THS shows gadolinium enhancement of cavernous sinus wall with or without focal narrowing of ICA within cavernous sinus [4].

In the study by Mehta., *et al*. [1,7] most common cranial nerves involved are Abducent (6th), Occulomotor (3rd) and Trigeminal (5th) while most common combination of lower cranial nerve involvement is 9th, 10th, 11th. The cause for lower cranial nerve involvement is vascular, traumatic, neoplastic, iatrogenic and less likely infective. The most common anatomical site of involvement is Cavernous sinus and then is Orbital apex. The most common cause for infections are Tuberculous, Fungal and in tumors: Metastasis, Schwannoma leading to cranial neuropathy. The study shows a very close resemblance to this case report in terms of presentation.

As per Keane' series [5]: The most common cranial nerves to be involved are VI, VII, V. Among the combination of two cranial nerve involvement- (III, VI)/(V, VI)and lastly (V, VII) while combination of 3 cranial nerves (III, IV, VI)/(V, VI, VII) and lastly (II, III, VI). The most common anatomical site to involve is Cavernous sinus, Brainstem and Nerve. The most common cause is Tumor (Schwannoma, Metastasis, Meningioma), Vascular disease and lastly Traumatic etiology.

In this study:

- This young female patient had a chronic course of illness.
- She had a history of Extrapulmonary Koch's for which she was treated with probable history of recurrence.
- There were no risk factors except for the fact that she was in her 7th month of gestation when she contracted this infection plus she even lactated during the course of her illness and also while on Anti-tubercular therapy.
- She had multiple cranial nerve involvement with palsy probably due to tuberculous etiology as per the signs and symptomatology though there was nothing specific on radiological investigations which most likely seemed to be Cavernous sinus syndrome.

Conclusion

Multiple cranial neuropathy is a huge entity on its own involving assorted factors such as source of origin, symptoms and signs during presentation, site occupancy to involve the cranial nerves which all in all rolls up together to lead to a plausible, provisional and prompt diagnosis.

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Volume 5 Issue 7 July 2021