CASE REPORT

Positional hoarseness: an unusual symptom in jugular foramen mass

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SUMMARY

We came across a case of jugular foramen mass causing positional hoarseness on turning the head left and disappearing on returning the head to a straight position. Hoarseness of voice due to vagus nerve involvement is seen in jugular foramen mass but positional hoarseness has never been seen before. We report this rarest presentation and discuss the pathophysiology behind it.

BACKGROUND

Glomus jugulare tumour may present with various clinical symptoms from lower cranial nerves palsy and pulsatile tinnitus to hearing loss, depending on the extent of growth. Hoarseness of voice due to vagus nerve involvement is seen in jugular foramen mass but positional hoarseness has never previously been seen.

CASE PRESENTATION

A 35-year-old man presented with insidious onset, gradually progressive dysphagia, dysarthria and unilateral wasting of tongue muscles. One additional interesting and unusual presenting symptom was positional hoarseness of voice, only on turning his head towards the left. There was no history of long tract or cerebellar involvement, trauma, neck mass or pulsatile tinnitus. On examination, there was strictly unilateral paralysis of the soft palate and uvula, diminished pharyngeal laryngeal sensations and vocal cord palsy on the left. Atrophy and fasciculations of tongue muscles on the left were seen (figure 1) along with ipsilateral loss of gag reflex, weakness of contralateral head turning and ipsilateral shoulder elevation. Positional hoarseness was observed only on turning the head towards the left;

the voice quality in other positions was normal. Direct laryngoscopy confirmed partial left vocal cord palsy (figure 2). In view of the strictly unilateral involvement of lower cranial nerves 9–12, and absence of long tract/cerebellar signs and Horner's syndrome, a diagnosis of Collet-Sicard syndrome was considered.

INVESTIGATIONS

MRI of the brain with special emphasis on the skull base was carried out, which revealed a salt-and-pepper appearance of the left jugular foramen region mass on both T1-weighted and T2-weighted sequences (figure 3); CT of the head to access the bony margins of the tumour was typically irregularly eroded, with a moth-eaten pattern.

Figure 1 Left side tongue atrophy.

Figure 2 Direct laryngoscopy (partial left vocal cord palsy).

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Figure 3 Salt-and-pepper appearance on MRI of the left jugular foramen region mass.

DIFFERENTIAL DIAGNOSIS

None

TREATMENT

The patient was operated on using a transcondylar approach after preoperative embolisation. Biopsy showed a glomus tumour.

OUTCOME AND FOLLOW-UP

The patient was neurologically in the same status postoperatively as well as in follow-up.

DISCUSSION

The jugular foramen paraganglioma is a locally invasive, benign, encapsulated, hypervascular tumour that arises from the jugular foramen of temporal bone, growing slowly and causing various symptoms such as pulsatile tinnitus and low cranial nerve palsy.¹ Tumours of the jugular foramen are uncommon, constituting ~0.3% of all intracranial tumours. Glomus jugulare tumours account for 60–80% of all such cases. Jugular foramen paragangliomas are known to occur predominantly in the 50–60-year age group and the female–male ratio is reported to be 5:1.^{1 2}

The clinical course of jugular foramen paragangliomas reflects their slow growth and paucity of symptoms, and often results in a significant delay in diagnosis.^{1 3} The most common presenting symptom is pulsatile tinnitus, followed by hearing loss.^{2 4} Lower cranial nerve dysfunction is relatively common with glomus jugulare tumours and includes dysphagia, hoarseness, aspiration, tongue paralysis, shoulder drop and voice weakness.^{3 4} Neurological examination of lower cranial nerve palsy is crucial since most patients have involvement of at least one lower cranial nerve.

Possible pathophysiology of positional hoarseness

The normal tone of the vocal cords is maintained by the geometric arrangement of the various laryngeal cartilages in addition to active muscle contraction. This geometric arrangement undergoes slight changes with neck rotation, causing the resultant change in the tone of the vocal cords. When the neck rotates to the right, the resting tone of the right vocal cord is

slightly decreased and that of the left vocal cord is increased. This is explained by the decrease in the distance between the thyroid cartilage and the right arytenoid cartilage, and the increase in the distance between the thyroid cartilage and left arytenoid cartilage. The converse happens when the neck rotates to the left. Normally, these variations do not produce a change in voice because when sound is produced the active muscular contraction equalises the vocal cord tension on both sides. This normal balance is disturbed when there is vocal cord paresis on one side (figure 4). We hypothesise that our patient had early paresis of the left vocal cord, which was evident as hoarseness was only on turning to the left, because in that position the disproportionate tension of the two vocal cords cannot be overcome by the weak contraction of the paretic muscles. It is to be stressed that hoarseness of voice in relation to the position of the neck will only be evident in early paresis of one of

Figure 4 Sketch diagram of pathophysiology of positional hoarseness: (A) vocal cords on straight head position having equal tension at rest, (B) on left side head turn position, partially weak left vocal fold is lax and causes hoarseness during phonation, (C) compensation by right vocal fold (arrows) during phonation while head is straight and (D) while head is turned to the right.

the vocal cords, because once full-blown palsy of one of the vocal cords sets in, hoarseness will be evident in all positions.

The initial symptom of voice change, caused by vocal cord paralysis, preceded the presenting symptoms of hearing loss and tinnitus by an average of 2.5 years. The concept of early cranial nerve involvement by glomus vagale tumours was supported in the literature review. The evaluation of 'idiopathic' vocal cord paralysis must include radiographic inspection of the skull base, as was carried out in our case, whereas an accurate, temporal account of presenting symptoms may help distinguish vagal body tumour from other vascular neoplasms of this region.⁵ Since the tumour is hypervascular in nature, angiography is

crucial to identify the main feeding vessels and to embolise them prior to surgery, which decreases the chance of massive bleeding intraoperatively.⁶ Complete surgical resection is the ideal management of most jugular foramen paragangliomas.^{2 4 7}

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Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- Symptoms of positional hoarseness and direct laryngoscopy assisted confirmation of positional vocal cord paresis can be an early feature of ipsilateral extrinsic vagal nerve compression.
- A jugular foramen mass should be suspected in a case of positional hoarseness with or without involvement of other cranial nerves.

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