

Silent stellate ganglion paraganglioma masquerading as schwannoma: A surgical nightmare

ABSTRACT

A 28-year-old normotensive female presented with Horner's syndrome and paresthesia over the left side of the chest. Imaging study showed a large heterogeneous enhancing lesion in short-T1 inversion recovery sequence with flow voids in T2W sequence of magnetic resonance imaging. The lesion was located in the left-sided D1 and D2 regions extending into the neural foramina and apical part of the lung. During surgery, even minimal dissection of the tumor resulted in marked fluctuation in hemodynamic parameters, requiring temporary suspension of the surgery multiple times until hemodynamic parameters were brought under control by the anesthesiologist with drugs. The massive fluctuation in hemodynamic parameters in an unprepared and unanticipated scenario was a challenge for the anesthetist and surgeon. The tumor was radically excised with improvement of paresthesia in the immediate postoperative period, but Horner's syndrome persisted. After 18-months of follow-up, she was relieved of all symptoms including Horner's syndrome. Histopathological examination confirmed our suspicion as paraganglioma.

Keywords: Horner's syndrome, paraganglioma, silently functional, stellate ganglion

INTRODUCTION

Stellate ganglion is called cervicothoracic sympathetic ganglion and is formed by coalescence of the inferior cervical ganglion with first thoracic ganglion or sometimes by second thoracic sympathetic ganglion. Stellate ganglion is related anteriorly to the vertebral artery, posteriorly to the transverse process of C7 and the head of the first rib, and medially to the prevertebral fascia.^[1] Lesion in stellate ganglion presents with a triad of symptoms of mydriasis and partial ptosis with or without anhidrosis. The symptom complex is called Horner's syndrome. We report a case of stellate ganglion paraganglioma with classical features of Horner's syndrome with intraspinal extension, which was managed successfully. Our literature survey could not isolate any such similar case.

CASE REPORT

A 28-year-old normotensive female presented with a history of paresthesia in the left side of the chest for 2 years. For the last 8 months, she noticed partial left-sided ptosis

and anhidrosis over the left half of the face. Neurological examination revealed the left-sided pupil to be dilated and nonreacting to light. Her symptom complex fitted into the classically described Horner's syndrome [Figure 1]. Sensory examination revealed hyperesthesia in D2 and D3 dermatome in the left side. There was no other positive neurological finding. Magnetic resonance imaging (MRI)


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of the cervico-dorsal spine showed a large paravertebral and extrapulmonary heterogeneously enhancing lesion in short-T1 inversion recovery sequence of MRI at the D1–D2 level on the left side that extended into the neural foramen and compressed the thecal sac. T2W imaging showed multiple flow voids with heterogeneous intensity [Figure 2].

Operative technique

The patient was operated in prone position. D1 and D2 hemilaminectomy was performed. The tumor extended into the spinal canal through the neural foramen. Major portion of the tumor was anterolateral to the transverse process and vertebral body. The tumor was reddish in color, was firm in consistency, and was highly vascular. It was adherent to the pleura. During the process of tumor resection, there were large fluctuations of pulse rate and blood pressure. The patient had tachycardia with heart rate ranging between 120 and 200/min. The range of systolic blood pressure fluctuated between 70 and 200 mmHg. Due to this unprecedented condition, part of the tumor in proximity to the pleura was left behind and rest of the tumor was resected rapidly. In the immediate postoperative period, the patient was relieved of paresthesia over the chest wall, but Horner's syndrome persisted. At a follow-up after 18 months, she was completely relieved of all symptoms.

Histopathology

Histopathology revealed a vascular neoplasm composed of small and large sinusoidal vessels along with focal anastomosing small vessels separated by poorly cohesive tumor cells focally showing ill-formed “Zellballen” pattern. The cells were ovoid to polyhedral with granular cytoplasm with pleomorphic nuclei. Some vessels showed hyalinized wall. Mitotic activity was not seen. The findings were consistent with the diagnosis of paraganglioma [Figure 3].

DISCUSSION

Paragangliomas are slow-growing vascular tumors that arise from neural crest cells. Head and neck is the most common site (62.7%) followed by jugular bulb/tympanic plexus (26.7%) and vagus nerve. The prevalence of other unusual sites of occurrence is reported to be 2%.^[2,3] These tumors are usually benign and locally aggressive in nature. Symptoms rarely occur until the tumor enlarges and exerts pressure on the surrounding structures. Carotid body and vagal paraganglioma present with a mass in the neck, and tinnitus and hearing loss is the common presentation in jugulo-tympanic region tumors. There are very few reports of cervical sympathetic paraganglioma reported in literature.^[2-5] All have presented with painless mass in the neck and rarely with Horner's syndrome. There are two case reports of



Figure 1: Left eye showing decreased lid aperture (partial ptosis)

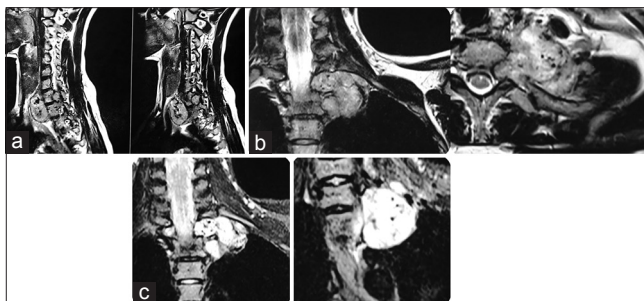


Figure 2: (a) T1W sagittal image showing left-sided isointense mass with flow voids at D1 and D2 level extending into the spinal canal. (b) D1 and D2 level with multiple flow voids compressing the thecal sac. (c) Short-T1 inversion recovery images showing intense contrast enhancement of the lesion with multiple flow voids

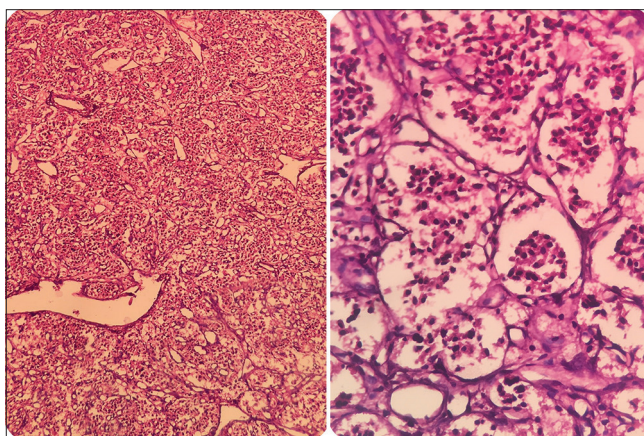


Figure 3: Histology revealing small and large sinusoidal vessels along with focal anastomosing small vessels separated by poorly cohesive tumor cells focally showing ill-formed “Zellballen” pattern

stellate ganglion schwannoma that presented with Horner's syndrome and with palpable mass in the neck.^[6,7] However, we could not find any case of paraganglioma affecting the stellate ganglion. During surgery, the patient developed extremely labile hemodynamics which made our surgery arduous. The labile hemodynamics were probably due to pouring of catecholamines into the blood during handling of the tumor. Adjuvant treatment was not given to the patient

for the residual tumor mass considering its benign nature. After 18 months of follow-up, the patient is asymptomatic.

Flow voids on T2W or T1W imaging can indicate the presence of paraganglioma. However, other vascular tumors in parapharyngeal space such as metastatic hypernephroma or thyroid carcinoma may mimic features of paraganglioma.^[4] A differential diagnosis of paraganglioma of neck can be made by pattern displacement of arteries and vein in the vicinity. Digital subtraction angiography can be considered in cases where preoperative embolization is deemed necessary to decrease the risk of bleeding during surgery.

Familial paragangliomas account for 7%–9% of those arising from the head and neck and show multicentricity in 25%–33% of cases as compared to sporadic ones (2%–10%).^[8,9] Malignant potential and metastasis of paragangliomas depends on the location, size, and germline mutation of succinate dehydrogenase (SDHB) gene mutation. Paragangliomas of size >5 cm and located in abdominal, para-aortic, and mediastinum have metastatic potential of up to 40%–70%.^[10] In this group of patients, functional imaging studies such as ¹²³I-meta-iodo-benzyl-guanidine or ¹⁸F-fluorodeoxyglucose positron emission tomography scans are recommended to detect metastases that cannot be detected by computed tomography or MRI.

In our literature survey, we found nine cases of cervical sympathetic paragangliomas. Four of these cases were catecholamine secreting. In five cases, there was ipsilateral Horner's syndrome.^[3,4] Our patient had a unique clinical presentation with ipsilateral Horner's syndrome and paraesthesia without any history of hypertension or mass in the neck. Although all paragangliomas are theoretically capable of producing catecholamines and other neuro-endocrine hormones, in fewer than 2% of head-and-neck tumors is this clinically detectable and they are often referred as extra-adrenal pheochromocytomas.^[9] In the present case, paraganglioma was silent preoperatively without any symptoms of functionality. We identified two case reports of silently functioning cervical sympathetic paraganglioma (not stellate ganglion) that presented with painless cervical mass and Horner's syndrome without any history of hypertension. One patient had mortality due to cardiac arrest in the immediate postoperative period.^[3] Paragangliomas can be missed in diagnosis preoperatively and come up with unwanted surprises during and after operation. If we can anticipate prior to surgery, urinary and plasma metanephrine tests and appropriate adrenergic blocking agents can be used before and during surgery to prevent hypertensive crisis.

CONCLUSION

Stellate ganglion paraganglioma is extremely rare, and its silent functionality may be a concern sometimes. Ipsilateral Horner's syndrome clinically and multiple flow voids in imaging constitute diagnostic hallmarks of this entity. Paraganglioma should be suspected as an important differential diagnosis of schwannoma, especially in the cervico-thoracic region. Adequate preparedness before surgery is essential to avoid untoward events during and after the surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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