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Case Report

Large posterior mediastinal ganglioneuroma with intradural cervical spine extension: A rare case report and review of literature

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ABSTRACT

Introduction: Ganglioneuroma (GN) is a rare benign tumor of ganglion cell origin and can occur anywhere along the sympathetic chain. These tumors can grow to a significant size without any symptoms unless they exert a mass effect on the region they grow and start showing symptoms. Spinal extensions are rare and they may produce neurological symptoms warranting further investigation.

Case presentation: We described a case of posterior mediastinal ganglioneuroma in a 4-year-old boy with cervical extension who presented with quadriparesis. The radiological scan revealed large ganglioneuroma having an intradural extramedullary extension with a large posterior mediastinal component compressing and displacing the surrounding structures. He underwent consecutive surgeries for complete excision of the tumor following which he regained his power in his upper and lower limbs over the period of 2 months.

Conclusion: Para spinal mass with consistent radiological features suggests ganglioneuroma but confirmation should be done with biopsy. Complete excision of the tumor is the treatment of choice with close follow-up for clinical improvement and recurrence.

1. Introduction

Ganglioneuroma (GN) is a rare neurogenic tumor that arises from the neural crest cells of the sympathetic ganglia or, even more rarely, the adrenal medulla; yet, it is benign in nature [1]. Histologically, they are benign tumors made up of mature schwannian stroma and ganglion cells [2]. They can develop anywhere along the sympathetic chain from the skull base to the pelvis, although the most usually afflicted areas are the posterior mediastinum, retroperitoneum, and adrenal gland [3]. These tumors are typically asymptomatic and develop to a considerable size; however, depending on the location of the tumor, symptoms may occur from loco-regional compression [4]. Although there have been reports of ganglioneuromas with spinal extension, intradural extramedullary ganglioneuromas are exceedingly rare [5]. Here, we describe a 4-year-old boy who had a large ganglioneuroma having an intradural extramedullary extension with a large posterior mediastinal component compressing and displacing the surrounding structures. A multidisciplinary team was consulted, and surgery was scheduled in two parts. The neurosurgery team conducted the first operation to remove the tumor's spinal component, and the cardiothoracic vascular surgeon team did the second surgery to remove the tumor's mediastinal component. This case report has been reported in line with the SCARE criteria [6].

2. Case presentation

A 4-year-old boy presented with bilateral upper and lower limb weakness that had developed gradually over the previous 3 months. His frailty had advanced dramatically by the time he presented, and he could no longer walk or stand unassisted. Further neurological evaluations indicated that his upper and lower limbs had diminished power (3/5 and 2/5); nonetheless, the remainder of his examination was normal, and his bowel and urine habits were normal. Following that, a magnetic

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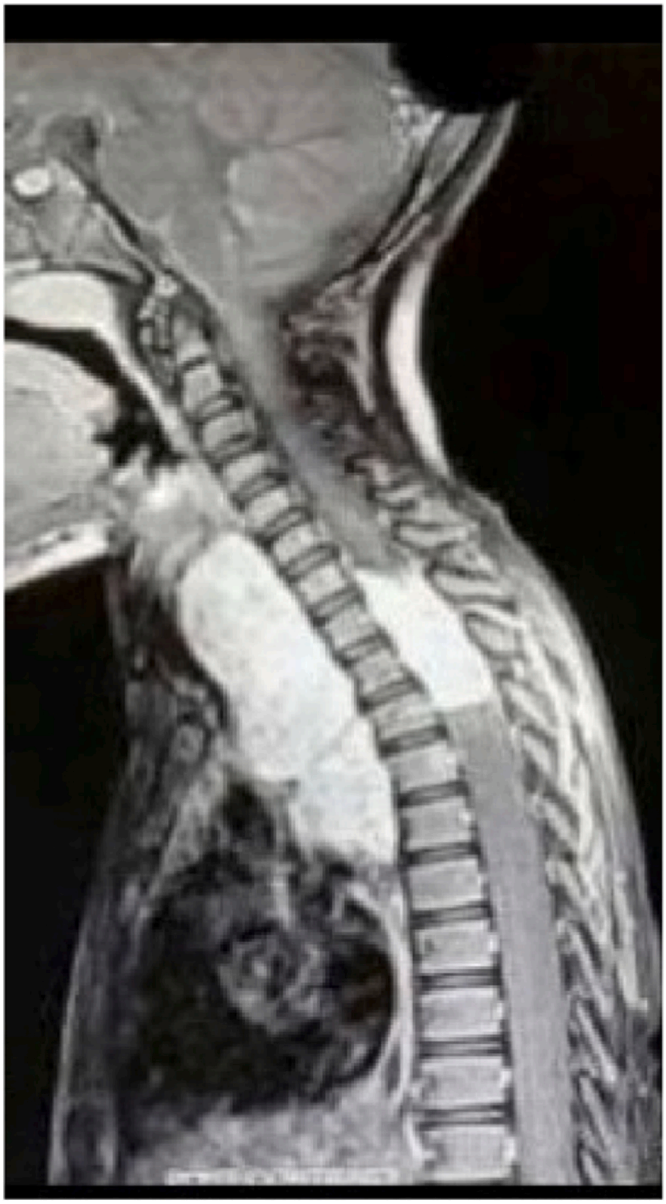

Fig. 1. Preoperative MRI showing the cervical extension of the tumor with (1.1*0.9*2.7 cm) mass in the extramedullary intradural space.

resonance imaging (MRI) scan of the spine was performed, which revealed a 1.1*0.9*2.7 cm intradural extramedullary (Fig. 1) altered signal intensity lesion extending cranio-caudally from the D1 to D3 vertebra that was extending through the left exiting neural foramen at multiple levels (D1-D2, D2-D3, D3-D4 levels) expanding it, with a large extraspinal component involving the paravertebral and visceral compartment of mediastinum measuring 5.9*7.2 cm; compressing the surrounding structures and displacing the esophagus and trachea to the right side and great vessels (brachiocephalic trunk, left common carotid artery and left subclavian artery) anteriorly. The extraspinal component extended cranio-caudally from C5 to D5 vertebral level.

2.1. Surgical approach

The tumor infiltration into the cervical spine was initially resected surgically. A midline incision was used for laminectomy and exposure of the D1-D3 posterior spinal canal, and a durotomy was performed to expose the tumor's intradural component (Fig. 2). The compression and displacement of the cord were visually confirmed. Extradural and

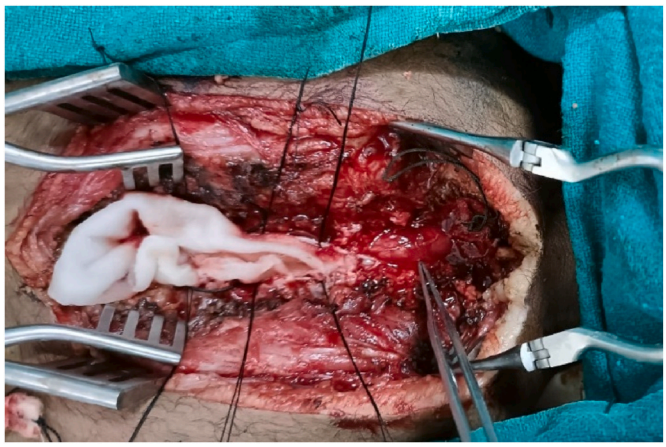

Fig. 2. Intra-operative view of the tumor in the cervical canal extending to the dura and durotomy beneath.

intradural tumor components, including nerve roots, were resected, and nerve roots and the spinal cord were freed from compression. Mass was sent for a histopathological examination, which confirmed the ganglioneuroma diagnosis. His weakness had significantly improved on regular follow-up, and he was able to stand and walk unaided again after 2 months postoperatively. The second stage of surgery was planned to remove the tumor's posterior mediastinal component. To remove the remaining tumor, a left thoracotomy with left neck exploration was performed. Mass was excised in fragments, releasing surrounding structures that had been compressed by mass.

2.2. Pathological examination

The tumor had a smooth and well-encapsulated macroscopic appearance, ranging from grey white to grey brown. The tumor was homogeneous when sliced, with no areas of hemorrhage, necrosis, or degeneration. The tumor was comprised of Schwann stromal cells organized in short fascicles and whorls with a fibrillary matrix under the microscope. Ganglion cells were scattered slightly and organized in sheets and cords. These cells have circular nuclei that are eccentrically placed, vesicular chromatin, prominent nucleoli, and rich eosinophilic cytoplasm. Nerve bundles, blood vessels, and mild inflammatory cell infiltrate of lymphocytes and mast cells were mixed in. The overall histomorphological findings were suggestive of ganglioneuroma.

2.3. Post-operative management

A post-operative MRI scan revealed the tumor's near complete excision and the re-alignment of the compressed tissues. The enlarged neural foramen and freed spinal cord were also confirmed. Neurological examinations and any signs of recurrence were closely monitored. Within two months, the patient's neurological function had recovered and he had full strength in his upper and lower limbs, and he has been under regular follow-ups to this day.

3. Discussion

GN are slow-growing, well-differentiated autonomic nerve system tumors that are usually asymptomatic. Clinical symptoms include local symptoms of obstruction, however, some individuals may also have diarrhea, hypertension, virilization, and myasthenia gravis [7]. The clinical characteristic that led to the diagnosis, in this case, was spinal cord compression, which resulted in bilateral (b/l) upper and lower limb paralysis. Ganglioneuromas usually afflict young children and are typically singular in nature [8]. Similarly, our patient was 4 years old at the time of presentation.

Table 1
Clinicopathological features of ganglioneuroma in the literature.

Author	Age (yrs)/ Gender	Level/ Laterality	Size	Intraspinal Extension	Clinical Presentation	Operation/Resection	Follow-up/ Recurrence
Harsh Deora et al. [18]	57yrs/ male	C6,C7/ Right	No Data	Intradural	Quadriparesis and pain in the nape of the neck	C6–C7 Hemilaminectomy and total resection	12yrs/no data
Akira Hioki et al. [10]	72yrs/ male	C1,C2/b/l	No data	intradural	Progressive incomplete paraplegia	C1–C2 posterior arch removal and durotomy/subtotal resection	2yrs/no recurrence
Ana-Maria Lonescu et al. [19]	53yrs/ female	C5–C7/Left	58*28mm	intradural	Triparesis and impaired walking	C5-c7 laminectomy and durotomy	1yr/no recurrence
Panduranga Seetatial-Maraj et al. [20]	42yrs/ female	C2–C4/b/l	No data	intradural	B/l upper limb numbness and headache for a week	C1–C2 decompression and C2 nerve root tumor excision	6 weeks/no recurrence
Obande J O et al. [5]	26yrs/ male	C1–C2/ Right	No data	intradural	Neck pain with weakness in upper and lower limbs	Right hemilaminectomy of C1–C2	1 month/no recurrence

The thoracic cavity (60%–80%, posterior mediastinum), the abdominal cavity (10%–15%, adrenal gland, retroperitoneum, pelvic, sacral, and coccygeal sympathetic ganglia, and the organ of Zuckerkandl), and the cervical area (5%) are the most common locations for GN [9]. The spinal canal contributes to approximately 5–8% of all GN [5]. On radiological imaging, a paraspinal ganglioneuroma can also extend through the neural foramen into the spinal canal, producing a dumbbell shape [10]. Although intradural tumor extension into the spinal canal has been recorded, it is exceedingly rare [11].

Radiology is crucial in the early workup. A recent analysis of cases reported aided in defining the radiographic characteristics that identify GNs. GNs present as homogenous or slightly heterogeneous hypodense lesions on CT imaging. GNs can develop calcifications. GN histology describes the MRI results. On CT, the mucous matrix looks hypodense, but on T2 MRI, it appears hyperintense. The nerve cell bodies and fibers are responsible for the higher density areas seen on CT as well as the heterogeneous hypointense areas shown on T2 MRI. T1 MRI may show amplification of the GN edge owing to the capsule. In the delay phase, MRI will also show gradual amplification [12].

Excisional biopsy, core needle biopsy, or fine-needle aspiration are all methods used to diagnose ganglioneuroma [13]. Ganglioneuromas are the most highly differentiated and have the best prognosis among the neuroblastic tumors [14]. Mature ganglion and Schwann cells are expected histological findings in ganglioneuroma [15]. Pathological examinations of our patient revealed Schwann stromal cells arranged in small fascicles and whorls with a fibrillary matrix.

While ganglioneuromas are benign, their involvement with surrounding tissues may create functional difficulties, prompting surgical surgery to remove the tumor [16]. In our patient, however, the patient had a large ganglioneuroma with an intradural extramedullary extension and a large posterior mediastinal component compressing the surrounding structures and displacing the esophagus and trachea to the right side, as well as great vessels (brachiocephalic trunk, left common carotid artery, and left subclavian artery) anteriorly.

The therapy for GNS involves complete excision of the tumor, which should be planned depending on the location, size, and intricacy of individual cases, either in one-stage surgery or several phased surgeries. Because GNS has a favorable prognosis because of its limited metastatic potential and moderate growth, thick adhesions to nerve roots or other critical structures should be avoided to limit permanent harm [17]. Our patient's operation was divided into two parts. The first operation was performed by the neurosurgery team to remove the tumor's spinal component, and the second operation was performed by the cardiothoracic vascular surgeon team to remove the tumor's mediastinal component.

We have reviewed some of the cases of the cervical spine extension of ganglioneuroma that has been published and compared some parameters for our better understanding of its presentation, management, and outcomes (Table 1).

4. Conclusion

The case presented in this study is a rare example of cervical spine ganglioneuroma with intradural extension. Because GNs can be asymptomatic, any paraspinal mass should be evaluated for a differential diagnosis. A multidisciplinary team approach should be used to plan the tumor's complete excision. Resection of the tumor is curative since these tumors are seldom malignant and have a low recurrence rate.

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Conflicts of interest

None.

Registration of research studies

No new surgical techniques or new equipment/technology was used.

Guarantor

Alok Dahal.

Consent

Written informed consent was obtained from the patient party for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Declaration of competing interest

None.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.104833.

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