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Asymptomatic Giant Thoracic Ganglioneuroma: A Rare Case Report and Literature Review

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ABSTRACT

Background: Ganglioneuroma (GN) is a rare, benign neuroblastic tumor arising from neural crest cells along the sympathetic chain. It is well-circumscribed, slow-growing, and commonly occurs in the posterior mediastinum (41%), retroperitoneum, adrenal gland, and neck. Despite its benign nature, GN can grow significantly without symptoms and is often detected incidentally.

Case Description: We report a 22-year-old female with an asymptomatic thoracic ganglioneuroma, incidentally identified during a pre-employment medical check-up. Thoracic MRI revealed a well-defined posterior mediastinal mass (11.7 × 10.2 × 14 cm) occupying the left hemithorax. The tumour was successfully resected via a posterolateral thoracotomy, and histopathology confirmed benign mature ganglioneuroma.

Conclusions: Thoracic ganglioneuromas are rare and often asymptomatic, making early detection challenging. Imaging plays a crucial role in diagnosis, and surgical excision remains the preferred treatment for large tumours to prevent complications.

Keywords: *Ganglioneuroma, thoracic, lobular, skeletal erosion, literature review.*

1. INTRODUCTION

Ganglioneuroma (GN) is a benign neuroplastic tumour that originates from neuroectodermal cells derived from the neural crest [1]. t is a well-defined, slow-growing tumour classified by the International Neuroblastoma Pathology Committee (INPC) as a Schwannian stroma-dominant tumour [2, 3]. Although ganglioneuromas are benign, they can grow to a considerable size, and the standard treatment approach involves subtotal resection of the tumour mass [4]. The most affected anatomical sites include the posterior mediastinum (41%), retroperitoneum (37%), adrenal gland (21%), and neck (8%) [5]. Thoracic ganglioneuromas are often asymptomatic, rarely reported, and frequently identified as incidental findings. Consequently, perioperative misdiagnosis is not uncommon, particularly in cases of intrathoracic pathology [6]. In this report, we present a rare case of thoracic ganglioneuroma in a 22-year-old asymptomatic female with no prior comorbidities, incidentally, discovered during a routine pre-employment medical examination. Given the typically indolent nature and nonspecific clinical presentation of such tumours, we complement this case with a systematic literature review of thoracic ganglioneuromas published from 1990 to the present. Our review focuses on reported symptomatology and demographic patterns among patients.

Case presentation

A 22-year-old asymptomatic female patient, with no known medical or surgical history, was admitted in October 2020 following the incidental discovery of a posterior mediastinal opacity on a chest X-ray during a pre-employment investigation. She was reassured at the time and was not on any medications. Her physical examination was unremarkable.

Imaging and diagnosis:

Thoracic magnetic resonant imagining (MRI) (Figure 1) revealed a well-defined posterior mediastinal mass measuring 11.7 x 10.2 x 14 cm, occupying the left hemithorax and extending to the level of T8. At the T3-T5 level, the mass extended through the left neural exit foramina into the epidural space, displacing the thecal sac to the right. The mass had a heterogeneously cystic component with foci of T1 signal hyperintensity suggestive of microhaemorrhages, along with curvilinear T1 and T2 hypointense bands, giving it a characteristic whorled appearance.

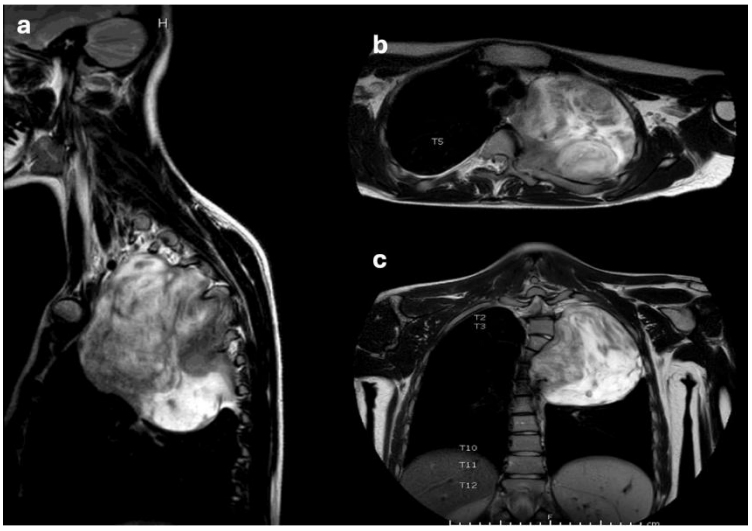

Figure 1: sagittal, axial, coronal T2-weighted MRI images (a, b, c respectively) showing left upper lung lobe ,mixed intensity lesion measuring 11.7x10.2x14 cm extending down to T8 vertebra.

A thorough history and physical examination were conducted to rule out positional dyspnoea and potential airway obstructive symptoms. Chest radiography was also reviewed to exclude any airway compromise, deviation, or anomalies. And her weight was recorded as 65 kg. Preoperatively, the patient's vital signs were as follows: blood pressure 116/75 mmHg, pulse rate 130 bpm, and oxygen saturation 98%. The cardiology team was consulted regarding the elevated pulse rate. They recommended intravenous cardio selective beta-blockers if tachycardia persisted after anaesthesia induction.

Surgical Procedure:

The surgery was performed by a multidisciplinary team comprising thoracic surgeons, neurosurgeons, pulmonologists, and cardiologists. A one-stage resection was planned. The tumour was initially dissected from its lateral attachment to the third and fourth ribs following resection of the fifth rib via a posterolateral thoracotomy incision. The tumour was found infiltrating the T3-T5 thoracic vertebrae and was carefully dissected (Figure 2).

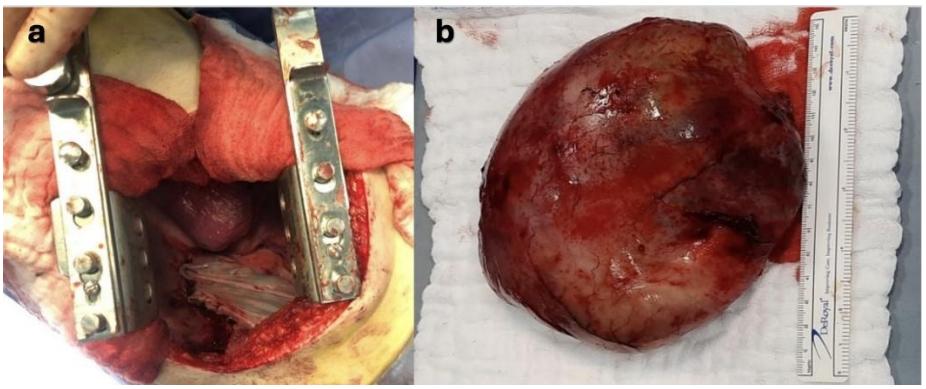

Figure 2: (a) Intraoperative view through a left thoracotomy showing the tumour's location and involvement; en-bloc excision was performed. (b) Gross specimen of the excised tumour placed next to a ruler for size reference the tumour bed.

The feeding artery was ligated using an Endo clip, and complete resection of the posterior mediastinal tumour was achieved. Haemostasis was secured, the collapsed lung was re-expanded using positive pressure, and a chest tube was inserted.

Pathology Examination:

On gross examination, the tumour appeared smooth and well-encapsulated. Formalin-fixed tissue specimens were sent for histopathological analysis. Microscopically, the tumour consisted of clusters of mature ganglion cells with abundant eosinophilic cytoplasm, distinct cell borders, a single eccentric nucleus, and a prominent nucleolus. These ganglion cells were interspersed with Schwan Nian stroma arranged in small intersecting fascicles. Findings were consistent with a benign mature ganglioneuroma, without evidence of neuroplastic elements. The post-operative course of patient stay was unremarkable and devoid of complications; she was followed up with chest x-rays during her stay. Discussion GN is a benign neuroplastic tumour that arises from neuroectodermal cells originating from the neural crest [1]. Thoracic GN is typically asymptomatic; however, in rare instances, it may present with general symptoms such as chest tightness, wheezing, and coughing. If located near the thoracic vertebrae, it can contribute to scoliosis. Additionally, when the tumour extends into the intervertebral foramina and compresses the spinal cord, neurological symptoms may develop [7, 8]. To better understand the clinical presentation of intrathoracic GNs, we conducted a comprehensive literature review spanning from 1990 to the present. Our aim was to identify common symptoms of this rare entity. The inclusion criteria were studies specifically addressing primary intrathoracic GN. Exclusion criteria included cases with incomplete or missing critical patient information, reports of secondary intrathoracic GN, cases involving cross-regional GNs such as those extending into the cervical spine or abdomen, and GNs occurring outside the thoracic cavity. This review provides insight into the clinical manifestations and diagnostic challenges associated with intrathoracic GNs, contributing to a better understanding of their management and outcomes. A total of 27 articles and 54 cases including the present case, were analysed using Jamovi 2.6. The clinical characteristics of the included patients are presented in Table (1). A total of 54 cases were analysed, with a mean age of 20.7 years (SD: 19.5) and an age range from 2 to 74 years. Regarding gender distribution, most cases were female (75.9%), while 22.2% were male, and 1.9% were unspecified. The tumor was located on the left side in 44.4% of cases, the right side in 38.9%, and was bilateral in 1.9%.

Table 1: Review of Published Thoracic Ganglioneuroma cases. R: Right; L: Left; M: Male; F: Female; N/A: Not Available; yr: Years; SOB: Shortness of breath.

Study Name (Year)	Age (yr)	Gender	Side	Size (cm)	Asymptomatic	SOB	Cough	Haemoptysis	Chest Pain	Spine Pain	Scoliosis-Related Symptoms*	Additional Symptoms
Simpson 1991[10].	2	F	L	5×3×2	Yes	No	No	No	No	No	No	-
Simpson 1991	5	F	R	10×10	No	No	Yes	No	No	No	No	1.5 years of mild stridor, dry cough; no response to anti-asthma agents
Simpson 1991	5	M	R	4×3×1	No	No	Yes	No	No	No	No	One-week cough, anorexia, fever; chest X-ray showed RML pneumonia + posterior mediastinal mass
Simpson 1991	5.5	M	L	8×7×2	No	No	Yes	No	No	No	No	-
Simpson 1991	8	F	L	7×5×5.5	No	No	No	No	Yes	No	No	-
Simpson 1991	9	F	L	N/A	No	Yes	No	No	No	No	No	Long history of asthma; recent SOB; chest X-ray showed mass
Simpson 1991	10	F	L	10×7.5×7	No	Yes	No	No	Yes	No	No	Exertional dyspnea

Study Name (Year)	Age (yr)	Gender	Side	Size (cm)	Asymptomatic	SOB	Cough	Haemoptysis	Chest Pain	Spine Pain	Scoliosis-Related Symptoms*	Additional Symptoms
Simpson 1991	11	F	L	9×5×3	Yes	No	No	No	No	No	No	Routine chest X-ray for clinical duodenal ulcer
Simpson 1991	12	F	R	12×8×2	No	No	No	No	No	No	No	Cold, dry, red right hand since birth; X-ray to exclude cervical rib showed mass
Sakai 1992[11].	17	F	L	N/A	Yes	No	No	No	No	No	No	-
Osterhouse 2002[12].	25	F	L	15×7×3	No	No	No	No	No	Yes	Yes	Upper thoracic/lower cervical pain, sleeping difficulties, headache; pre-existing scoliosis diagnosis
Duffy 2005[13].	27	F	R	N/A	No	No	No	No	No	No	No	Gastrointestinal symptoms; history of asthma; brisk lower extremity reflexes
Velyvis 2005 [14].	15	F	R	8×8×2	No	No	No	No	No	Yes	Yes	T4–T5 back pain for 2.5 years postmenarche; no neurological symptoms
Maruyama 2007 [15].	74	F	R	6.9×5.8×1.6	Yes	No	No	No	No	No	No	-
Ko 2007 [16].	53	F	R	9×4.5×10	Yes	No	No	No	No	No	No	Incidental chest radiograph abnormality
Zhang 2009[17].	3	F	L	5.8×4.5×4.5	No	No	No	No	No	No	No	Watery diarrhea, hypokalemia, weight loss
Kitagawa 2010[18].	4	F	R	N/A	No	No	No	No	No	No	No	Dyspnea, severe wheezing, fever (1-day history)
Kato 2012[19].	62	F	R	5×5×2	Yes	No	No	No	No	No	No	-
Kato 2012	45	M	L	6×3.5×2.5	Yes	No	No	No	No	No	No	-
Kato 2012	16	F	R	9.2×6×4	Yes	No	No	No	No	No	No	-
Kato 2012	54	F	L	11×6.5×3	Yes	No	No	No	No	No	No	-
Kato 2012	57	M	R	3×2×1.5	Yes	No	No	No	No	No	No	-

Study Name (Year)	Age (yr)	Gender	Side	Size (cm)	Asymptomatic	SOB	Cough	Haemoptysis	Chest Pain	Spine Pain	Scoliosis-Related Symptoms*	Additional Symptoms
Kato 2012	20	F	R	7×4×3	Yes	No	No	No	No	No	No	-
Kato 2012	57	M	L	3×2.5×2	Yes	No	No	No	No	No	No	-
Kato 2012	12	F	Bilateral	L 9.5×6.5×5. R 3.5×2.5×1.5.	Yes	No	No	No	No	No	No	-
Kato 2012	36	F	R	8.2×6.4×3	Yes	No	No	No	No	No	No	-
Kato 2012	7	M	L	11×7×6	Yes	No	No	No	No	No	No	-
Kato 2012	15	F	R	8.3×6.6×3.5	Yes	No	No	No	No	No	No	-
Kato 2012	6	F	L	8.4×6.5×4.9	Yes	No	No	No	No	No	No	-
Kato 2012	62	F	L	10.7×4.1×2.9	Yes	No	No	No	No	No	No	-
Sánchez-Galán 2014[20].	3	F	N/A	N/A	Yes	No	No	No	No	No	No	-
Sánchez-Galán 2014	4	M	N/A	N/A	Yes	No	No	No	No	No	No	-
Sánchez-Galán 2014	4	F	N/A	N/A	Yes	No	No	No	No	No	No	-
Sánchez-Galán 2014	4	F	N/A	N/A	Yes	No	No	No	No	No	No	-
Sánchez-Galán 2014	5	F	N/A	N/A	Yes	No	No	No	No	No	No	-
Sánchez-Galán 2014	9	M	N/A	N/A	Yes	No	No	No	No	No	No	-
Sánchez-Galán 2014	11	F	N/A	N/A	Yes	No	No	No	No	No	No	-

Study Name (Year)	Age (yr)	Gender	Side	Size (cm)	Asymptomatic	SOB	Cough	Haemoptysis	Chest Pain	Spine Pain	Scoliosis-Related Symptoms*	Additional Symptoms
Sánchez-Galán 2014	13	M	N/A	N/A	Yes	No	No	No	No	No	No	-
Huang 2017[21].	12	F	L	12×12×12	No	No	No	No	Yes	No	No	Gradual lower extremity weakness (unable to stand/walk without support)
Jeon 2017[22].	6	M	R	4×3.5×2	No	Yes	Yes	No	No	No	No	Dyspnea, barking cough (3 days); Harlequin syndrome post-resection
Lambdin 2018[23].	42	F	L	23×10×10	No	No	No	No	Yes	No	No	History of hypothyroidism; decreased left breath sounds
Algazwi 2020[24].	18	F	R	N/A	No	No	No	No	No	No	Yes	Scoliosis diagnosed at 13; tumor found at 18; MRI mimicked idiopathic scoliosis
Elnady 2020[25].	17	F	L	N/A	No	Yes	No	No	No	Yes	Yes	-
Brock 2020[26].	12	F	R	10×9.1×9.5	No	Yes	Yes	No	Yes	Yes	No	Nausea, right shoulder pain; obesity; decreased right lung sounds
Aljuboori 2021[27].	30	N/A	R	N/A	No	No	No	No	No	No	No	Chronic right upper quadrant pain, nausea, bloating, constipation
Tiwari 2022[28].	4	F	L	3.8×2.5×2.3	No	No	No	No	No	No	No	Paraneoplastic cerebellitis and ROHHAD-NET syndrome
Nemoto 2022[29].	15	F	L	N/A	No	No	No	No	No	Yes	No	-
Chen 2022[30].	57	M	L	6.5×5×2	Yes	No	No	No	No	No	No	Lipomatous ganglioneuroma
Zhuang 2023[31].	15	F	R	14.5×12×5	Yes	No	No	No	No	No	No	Hashimoto’s thyroiditis
Chauhan 2024[32].	7	F	L	11.8×13.1×14.7	No	No	No	No	No	No	No	Weight loss

Study Name (Year)	Age (yr)	Gender	Side	Size (cm)	Asymptomatic	SOB	Cough	Haemoptysis	Chest Pain	Spine Pain	Scoliosis-Related Symptoms*	Additional Symptoms
Song 2024[33].	48	F	L	18.3×15.2×11	No	No	Yes	No	No	No	No	Hot flashes, night sweats, poor appetite, weight loss (2.5 kg/week); left lung hypopnea
Dinh 2024[34].	10	F	R	9×7.5×3	No	No	No	No	No	No	Yes	-
Dinh 2024	13	M	R	9×6.6×2.4	No	No	No	No	No	No	Yes	Pre-existing scoliosis with progression
Current Study	22	F	L	11.7×10.2×14	Yes	No	No	No	No	No	No	-

Clinical presentation:

More than half of the cases (54%) were asymptomatic, while 46% exhibited symptoms. Among symptomatic patients, the most reported symptom was cough (11%), followed by chest pain (9.3%), shortness of breath (9.3%), and spine pain (9.3%). Haemoptysis was not observed in any case (0%). Additionally, 7.4% of cases were initially identified due to scoliosis, and 3.7% of patients had pre-existing scoliosis We excluded Guan's paper [9] from our analysis because it lacked specific symptom data for each patient. However, the study reported a total of 22 cases, including 15 males and 7 females, aged between 4 and 57 years, with a median age of 21 years. Of these, 14 patients were under 20 years old, and 6 were younger than 10. In terms of case detection, 20 patients were identified through chest radiographs, with 16 being asymptomatic cases found during health exams and 4 symptomatic cases presenting with symptoms like cough, influenza, and fever. Additionally, two cases were discovered through CT scans—one involving cough and shortness of breath during sleep, and the other hospitalized due to latent chest and back pain.

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Conflicts of interest

The authors declare no conflicts of interest.

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