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CASE REPORT

Thoracic Inlet Tumor in a Child – A Rare Case Report

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Abstract:

Background: Thoracic inlet tumors are extremely uncommon in the pediatric age group because of the crowded arrangement of vital neurovascular structures in this region. Their diagnosis and surgical management are technically demanding, often requiring a multidisciplinary approach. Most such tumors are neurogenic in origin, and the literature describing pediatric cases is scarce.***Case Report:*** A 9-year-old female presented with a gradually enlarging swelling over the right upper chest, associated with breathing difficulty and recurrent cough. On examination, a firm mass was noted in the right supraclavicular region extending into the thoracic cavity, displacing the trachea to the left with reduced air entry on the right. CECT thorax revealed a well-defined, heterogeneously enhancing soft-tissue lesion in the right mediastinum extending from mid-C7 to mid-D5 vertebral levels. Tru-cut biopsy suggested Schwannoma. Video-assisted thoracoscopic surgery (VATS) was attempted but converted to open surgery due to difficult posterior and superior dissection. A small sternotomy was performed, and complete tumor excision was achieved using diathermy and blunt dissection. Postoperatively, the patient required a blood transfusion and developed a transient pleural effusion, which resolved by postoperative day 8. She was discharged on day 11. Histopathology confirmed Schwannoma.

Keywords: Thoracic inlet tumor, Schwannoma, Paediatric mediastinal mass, Sternotomy

INTRODUCTION

The thoracic inlet, also known as the superior thoracic aperture, represents the anatomical junction between the neck and the thorax. It is a narrow, crowded region bounded by the manubrium sterni anteriorly, the first thoracic vertebra posteriorly, and the first pair of ribs laterally (1). This space accommodates vital neurovascular and aerodigestive structures including the trachea, esophagus, subclavian vessels, carotid arteries, internal jugular veins, vagus and phrenic nerves, sympathetic chain, and the apex of the lungs. Because of this dense anatomical composition, any tumor in this region poses considerable diagnostic and therapeutic challenges (2).

Thoracic inlet tumors are rare, particularly in children. Most lesions encountered in this region are neurogenic in origin, arising from peripheral nerves, sympathetic chains, or intercostal nerves (3). Among these, Schwannomas and neurofibromas are the most frequently reported benign entities. In adults, thoracic inlet masses may also include lymphomas, thyroid goiters, Pancoast tumors, or vascular malformations (4). However, pediatric cases remain exceedingly uncommon, and literature describing their presentation, management, and outcomes is limited.

Children presenting with thoracic inlet tumors often show non-specific respiratory or compressive symptoms such as cough, dyspnea, chest discomfort, or swelling in the supraclavicular region. Because of the limited space and proximity to major vessels, such tumors can cause tracheal deviation, venous congestion, or brachial plexus compression (5). Imaging modalities like contrast-enhanced computed tomography (CECT) and magnetic resonance imaging (MRI) are essential for delineating the extent, vascular relationships, and possible intraspinal extensions of these masses. In many instances, histopathological evaluation through biopsy or surgical excision confirms the diagnosis (6).

The surgical management of thoracic inlet tumors presents multiple complexities. The choice of approach depends on the location, size, and relation of the lesion to surrounding vital structures. Various approaches have been described in literature, including supraclavicular, transcervical, thoracoscopic, and median sternotomy (7). Minimally invasive techniques such as video-assisted thoracoscopic surgery (VATS) are increasingly utilized; however, in large or posteriorly located lesions, open procedures remain safer and more effective. Conversion from VATS to open surgery is not uncommon, particularly when exposure is limited or dissection is difficult due to proximity to the subclavian vessels or brachial plexus (8).

In pediatric patients, the challenges multiply due to smaller anatomical dimensions, increased risk of blood loss, and proximity to developing structures. The decision-making process often requires intraoperative flexibility and collaboration between pediatric surgeons, thoracic surgeons, and cardiovascular thoracic surgery (CVTS) teams (9). Complete surgical excision remains

the treatment of choice for benign tumors such as Schwannomas, as it provides both diagnostic confirmation and curative outcome (10).

Given the rarity of such tumors in children and the limited data available on optimal surgical approaches, each case contributes valuable insights to existing literature. The present report describes a rare case of a thoracic inlet Schwannoma in a 9-year-old female. It highlights the clinical presentation, diagnostic work-up, intraoperative challenges, and postoperative recovery, while emphasizing the importance of multidisciplinary planning and tailored surgical strategies for safe and complete tumor excision in pediatric thoracic inlet tumors.

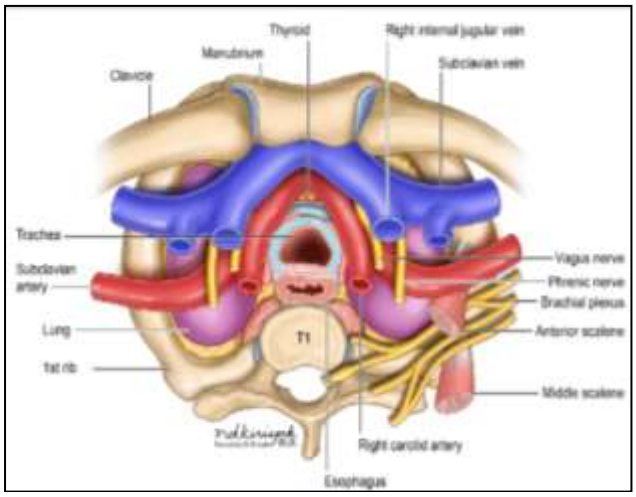

Fig 1- Thoracic Inlet (Superior Thoracic Aperture)

CASE REPORT

Patient Information: A 9-year-old female child presented to the outpatient department with a progressively increasing swelling over the right upper chest for the past six months. The swelling was insidious in onset and gradually increased in size. The child also complained of intermittent cough and mild respiratory difficulty, particularly on exertion. There was no history of fever, weight loss, dysphagia, or neurological symptoms such as limb weakness or paresthesia. Her developmental milestones were normal, and there was no significant family or medical history.

Clinical Findings: On physical examination, a swelling measuring approximately 8 × 6 cm was seen in the right supraclavicular region, extending inferiorly into the upper thoracic cavity. The mass was firm to hard in consistency, non-tender, and immobile. The overlying skin was normal. The swelling caused anterior displacement of the clavicle. Tracheal deviation to the left was evident, and air entry on the right side of the chest was reduced. No regional lymphadenopathy or upper limb neurological deficits were detected.

Diagnostic Assessment: A chest radiograph revealed a right paratracheal opacity extending into the superior mediastinum. Contrast-enhanced computed tomography (CECT) of the thorax demonstrated a well-defined, heterogeneously enhancing soft tissue lesion in the right mediastinum extending from the mid-C7 to mid-D5 vertebral levels, measuring 8.0 × 5.6 × 8.2 cm. The mass was abutting the subclavian vessels and the apex of the lung, with no intraspinal extension. A CT-guided tru-cut biopsy was performed, and histopathological analysis suggested a benign nerve sheath tumor, likely Schwannoma. Routine blood investigations and cardiac evaluation were within normal limits.

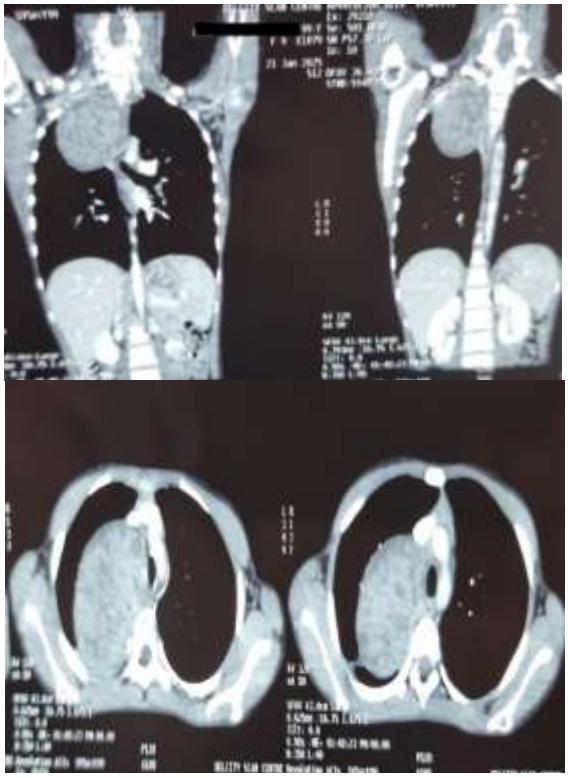

Fig 2: CECT Thorax suggestive of well-defined heterogeneously enhancing soft tissue lesion in right mediastinum extending from mid C7 to mid D5 levels measuring 8.0 X 5.6 X 8.2 cms.

Therapeutic Intervention: The patient was scheduled for surgical excision. Under general anesthesia, a right-sided video-assisted thoracoscopic surgery (VATS) approach was initially attempted. Anterior dissection was possible; however, due to difficulty in separating the posterior and superior margins from adjacent vital structures, the procedure was converted to open surgery. A small upper sternotomy was performed in consultation with the cardiothoracic and vascular surgery (CVTS) team. The tumor was carefully dissected and excised en bloc using diathermy and blunt dissection techniques. Intraoperatively, there was moderate blood loss requiring transfusion of three packed red cell units.

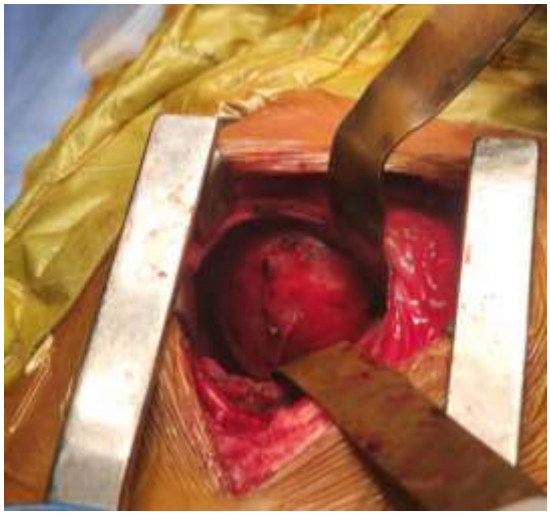

Fig 3: Intraoperative View of Thoracic Inlet Tumor Excision

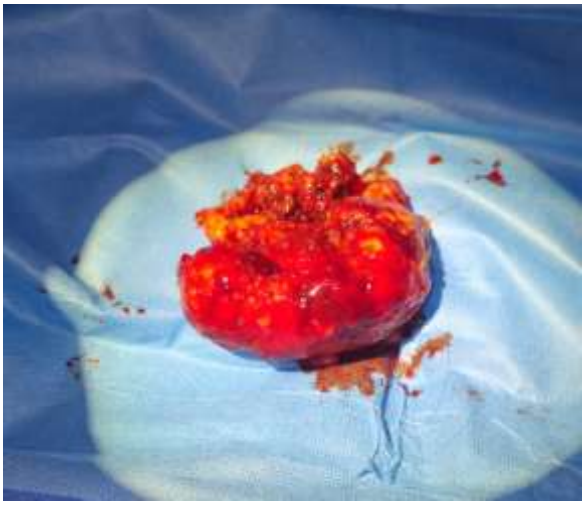

Fig 4: Excised Thoracic Inlet Tumor (Schwannoma)

Postoperative Course: The immediate postoperative period was uneventful except for the development of a mild right-sided pleural effusion on postoperative day 2, which was managed conservatively with an intercostal drain. The effusion resolved by postoperative day 8, and the drain was removed. The patient was mobilized early and discharged on postoperative day 11 in stable condition. Final histopathological examination confirmed the diagnosis of benign Schwannoma, showing characteristic Antoni A and Antoni B cellular patterns without atypia or mitotic activity.

Follow-Up and Outcomes: At the one-month follow-up, the patient was asymptomatic with complete wound healing and no signs of recurrence. Chest radiograph and ultrasound of the operative site revealed no residual mass or effusion. Long-term follow-up at six months demonstrated normal respiratory function and satisfactory cosmetic results.

DISCUSSION

Thoracic inlet tumors represent a distinct surgical challenge because of their deep anatomical location and close relationship with vital neurovascular structures. The thoracic inlet, bounded by the first rib, manubrium sterni, and T1 vertebra, contains essential structures such as the subclavian vessels, brachial plexus, trachea, esophagus, and sympathetic chains (11). Surgical access to this region is technically demanding, and tumors in this location are infrequent, particularly in children. Among the few pediatric cases reported, neurogenic tumors especially Schwannomas are the most common histological types (12).

Schwannomas are benign, slow-growing tumors arising from the Schwann cells of peripheral nerve sheaths. They can occur anywhere along the course of peripheral, cranial, or autonomic nerves . In the thoracic cavity, they typically arise from intercostal nerves or sympathetic chains. Involvement of the thoracic inlet, however, is rare, and only isolated pediatric cases have been described in literature. These tumors often remain asymptomatic until they reach a considerable size, when compressive symptoms or cosmetic deformity prompt clinical attention (13).

The clinical presentation depends on the site and size of the lesion. In our patient, the presenting symptoms of cough, dyspnea, and swelling over the right upper chest were due to compression of the trachea and displacement of the clavicle. The tracheal deviation and reduced air entry on the right side further reflected the mass effect of the tumor. Because these features can mimic other mediastinal or apical chest pathologies such as Pancoast tumors, tuberculosis, or lymphomas, detailed imaging is indispensable.

Contrast-enhanced computed tomography (CECT) and magnetic resonance imaging (MRI) remain the gold standards for diagnosis and preoperative planning. CECT helps delineate the tumor's size, extent, and vascular relationships, while MRI is superior for evaluating neural involvement and intraspinal extension. In this case, CECT showed a well-circumscribed, heterogeneously enhancing lesion with typical features of a neurogenic tumor. Tru-cut biopsy confirmed Schwannoma, allowing for definitive surgical planning.

The mainstay of treatment for thoracic inlet Schwannomas is complete surgical excision. Since these tumors are encapsulated, total resection is usually curative with negligible recurrence. However, the choice of surgical approach is crucial and must be tailored to the tumor's location and relation to adjacent structures (14). Minimally invasive methods such as video-assisted thoracoscopic surgery (VATS) are preferred for small, anteriorly located lesions because they offer reduced postoperative pain and quicker recovery (15). Nevertheless, large or posteriorly situated masses, especially those closely associated with major vessels or

the brachial plexus, often require conversion to open approaches for adequate exposure and safe dissection (16).

In our patient, thoracoscopic excision was initially attempted but was converted to an open approach due to difficulty in dissecting the superior and posterior margins of the mass. A limited upper sternotomy provided excellent exposure and facilitated complete excision while minimizing morbidity. The intraoperative decision to seek cardiovascular and thoracic surgery (CVTS) assistance exemplifies the importance of multidisciplinary collaboration in managing complex thoracic inlet lesions.

Postoperative complications primarily include bleeding, pneumothorax, pleural effusion, and transient nerve palsies. Our patient developed a mild pleural effusion, which resolved spontaneously by postoperative day eight. Histopathology confirmed the diagnosis of benign Schwannoma, characterized by Antoni A and Antoni B cellular patterns, Verocay bodies, and absence of atypia. Long-term prognosis following complete excision is excellent, with recurrence being exceedingly rare.

Comparatively, thoracic inlet tumors in adults are more frequently malignant, such as Pancoast tumors, and require multimodal therapy involving radiotherapy and chemotherapy in addition to surgery (17). In contrast, pediatric Schwannomas are benign and curative with surgery alone. The rarity of these tumors in children makes each reported case valuable in contributing to the understanding of their presentation and surgical management (18).

This case reports several important learning points. First, thoracic inlet Schwannomas, although rare, should be considered in the differential diagnosis of pediatric supraclavicular or apical chest swellings. Second, thorough preoperative imaging and multidisciplinary planning are vital for minimizing intraoperative complications. Third, while minimally invasive techniques are ideal, open approaches such as partial sternotomy or combined cervicothoracic incisions should not be hesitated when exposure is inadequate. Finally, complete excision remains the cornerstone of treatment, offering both diagnostic confirmation and long-term cure.

CONCLUSION

Thoracic inlet tumors in children are exceptionally rare, with Schwannomas being the most common benign variant. Their diagnosis and management demand careful imaging, surgical planning, and a multidisciplinary approach due to the complex anatomy of the region. Although minimally invasive surgery offers advantages, open approaches such as limited sternotomy may be necessary for safe and complete excision. Early recognition and total resection ensure excellent long-term outcomes with minimal recurrence risk. This case highlights that individualized surgical strategies and collaborative intraoperative decision-making are essential for successful management of pediatric thoracic inlet Schwannomas.

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