Current Thoracic Surgery

To cite this article: Konuk Balcı BM, Kocaman G, Sakallı MA, Yenigün BM, Gürsoy Çoruh A, Kırmızı BA, Heper A, Enön S. Giant pediatric intrathoracic ganglioneuroma: a case report and review of the literature. Curr Thorac Surg 2022 May; 7(2): 92-96. doi: 10.26663/cts.2022.015. CTSID: 725. Online ahead of print.

Case Report

Giant pediatric intrathoracic ganglioneuroma: a case report and review of the literature

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ABSTRACT

Ganglioneuroma is an extremely rare (incidence 1/1,000,000) well-differentiated neurogenic tumor that mostly originates from neural crest cells. The average age of diagnosis is 7 years old and it is mostly located in the posterior mediastinum (37.5%). Here we present a 10-year-old girl with a giant thoracic ganglioneuroma. The tumor was resected via posterolateral thoracotomy. With our case report included, there are 10 cases of surgically treated pediatric giant intrathoracic ganglioneuroma.

Key words: giant, ganglioneuroma, pediatric, thoracic surgery, posterior mediastinal tumors

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Doi: 10.26663/cts.2022.015
Received 19.09.2021 accepted 22.12.2021

Introduction

Ganglioneuroma is an extremely rare (incidence 1/1,000,000) well-differentiated neurogenic tumor that mostly originates from neural crest cells. It is mostly seen in infants and children and the M/F ratio is 2/3. It can be detected incidentally or it can be diagnosed by symptoms as a result of compression on surrounding tissues or catecholamine release. It is generally located in the posterior mediastinum (37.5%) and retroperitoneally. The general treatment approach is total resection of the tumor, and postoperative local recurrence is rare [1]. In this article, we present the 10th case of surgical resected giant pediatric intrathoracic ganglioneuroma in the literature.

Case Report

A 10-year-old girl was admitted to the hospital with shortness of breath and chest tightness. A uniform large opacity was detected in the upper zone of the left hemithorax at the PA chest x-ray (Figure 1a). Thorax computed tomography (CT) of the patient revealed a well-circumscribed homogenous hypodense 15x8x7 cm (CCxTXAP) mass with millimetric calcifications located in left posterior mediastinum which causes minimal scoliosis in the T5 and T6 vertebral bodies, and minimal erosion in the left 6th rib (Figure 1b). Thoracic MRI showed a T1W hypointense, T2W hyperintense mass that did not show invasion to the mediastinum, lungs, and adjacent thoracic vertebrae (Figure 1c). In PET-CT SUVmax 2.8 involvement was observed in the mass.

Adamkiewicz artery has been shown to arise from the left T9 level in Thorax CT angiography (Figure 2).

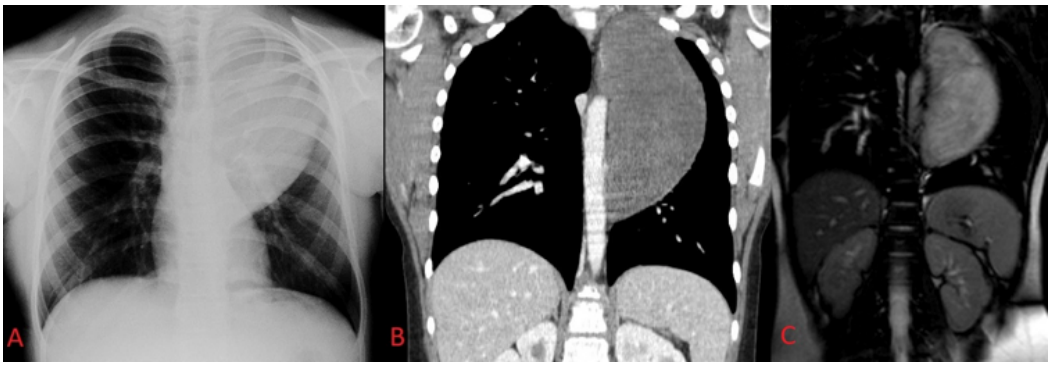

Figure 1. Posteroanterior chest x-ray showing a radioopacity at the upper zone of the left hemithorax **(A)**, coronal reformatted image of enhanced thorax CT demonstrates a well-circumscribed homogeneous hypodense mass lesion **(B)**, coronal T2-weighted MRI reveals the homogenous hyperintense left paravertebral mass lesion **(C)**.

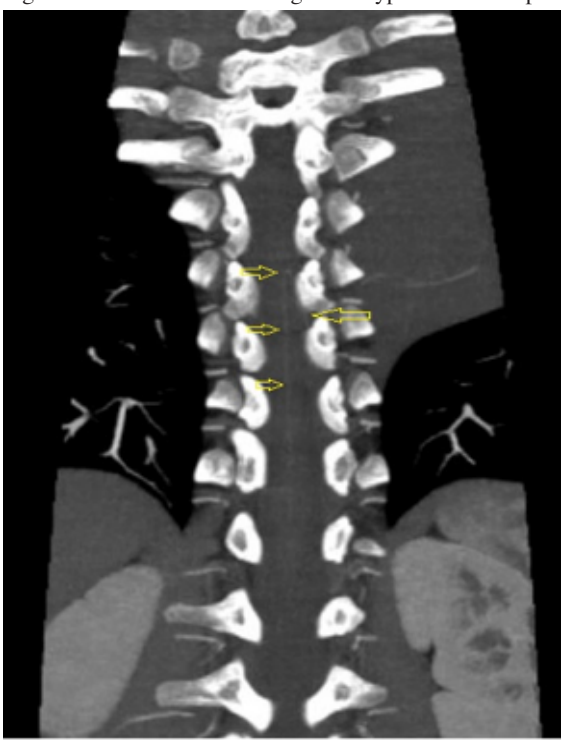

Figure 2. Longitudinal course of the Adamkiewicz artery (arrows) can be seen on maximum intensity projection (MIP) reformatted coronal thorax CT angiography.

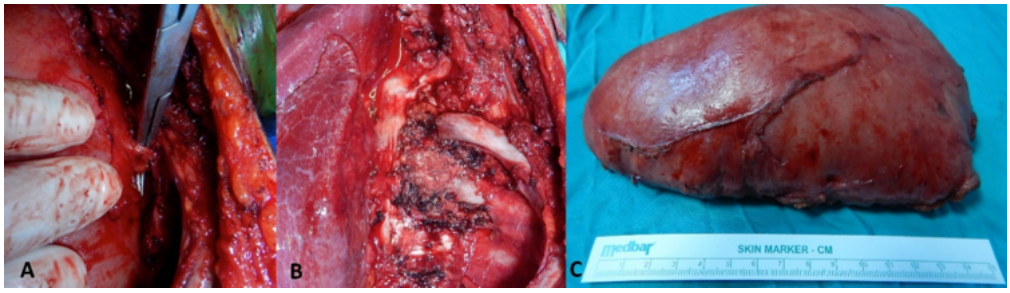

Figure 3. Intraoperative view of the mass and dissection of the intercostal nerve bundles going into the mass **(A)** , postoperative view of the thoracic cavity **(B)** and lateral view of the specimen **(C).**

Normetanephrine and homovalinic acid levels were found to be elevated in the 24-hour urine analysis (1106.7 µg/day and 9.2 µg/day, respectively). When the radiological and clinical results were evaluated together, the patient was thought to have a neurogenic tumor, and resection via posterolateral thoracotomy was planned. Making sure that the Adamkiewicz artery was below the dissection site, the posterior mediastinal mass was excised with partial resection of the left 6th rib (Figures 3a-c). The histopathological analysis revealed mature ganglion cells with large eosinophilic cytoplasm dispersed in the schwanian stroma and the pathology result was reported as mature ganglioneuroma with negative surgical margins (Figure 4). She was discharged on postoperative 5th day. No additional treatment was planned for the patient. Patient is asymptomatic during 96-months follow-up period. Written informed consent was obtained from the parents for publication of her data.

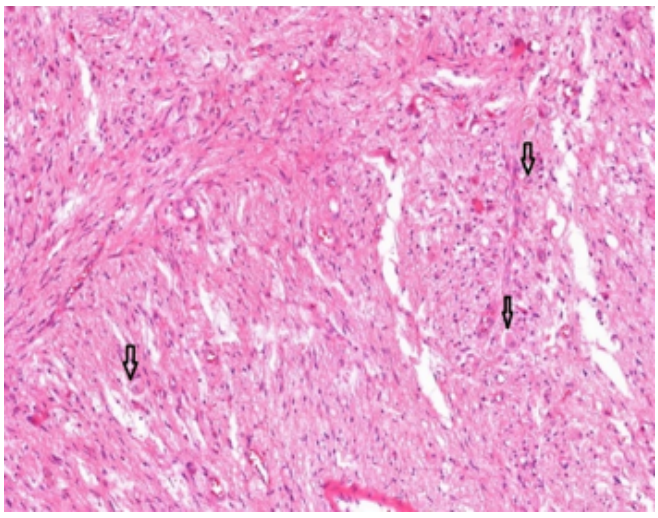

Figure 4. The histopathological analysis revealed mature ganglion cells with large eosinophilic cytoplasm (marked by arrowheads) dispersed in the schwanian stroma (HEx400).

Discussion

Ganglioneuroma is a benign neurogenic tumor that mostly originates from the sympathetic ganglion consisting of ganglion cells and stroma containing Schwann cells [2]. The average age of diagnosis is 7 years old, but it is likely to occur in older ages. Patients are usually asymptomatic at admission and tumoral tissue is detected incidentally [2]. However, cough, chest pain, shortness of breath due to compression of lungs may be present as in our case [3]. Diarrhea, sweating, and hypertension may occur due to the release of catecholamine and metabolites from the tumor. The rate of diagnosing ganglioneuroma under the age of 20 is 60%. It is mostly located in the posterior mediastinum but it can be seen in different parts of the body, especially retroperitoneum, adrenal glands or head and neck region and gastrointestinal tract. Mediastinal ganglioneuroma on imaging should be differentiated from cystic teratoma, neurilemmoma, lymphangioma cysticum or bronchocele [1]. When the tumor location is paraspinal, it can cause long-term compression- related scoliosis [2]. Although the etiology is not fully known, genetic diseases such as neurofibromatosis type 1-2 and multiple endocrine neoplasia (MEN) type 2 may be present in patients who have a family history. There are 10 cases of operated pediatric intrathoracic ganglioneuroma larger than 10 cm with the present case. The mean age of the patients is 9.7 years (5-17 years). Seven (64%) of the patients were female. The average follow-up time was 19.6 months, and none of them had recurrence. Five of all patients had left sided tumors and accompanying scoliosis needed correction surgery [1-6]. Only one patient had subtotal resection and only one patient [2] had serious bleeding as an intraoperative complication [6]. The list of publications operated for pediatric intrathoracic ganglioneuromas larger than 10 cm in diameter are summarized in table 1.

Table 1. List of publications presenting operated pediatric intrathoracic ganglioneuromas larger than 10 cm in diameter.

Year	A	S	Symptoms	Dm (cm)	Site	Treatment	Comp	Relapse	Follow-up (m)
2005[5]	12	F	Scoliosis	10x9, Spinal canal extension	R	R TR and correction surgery (Two stage surgery)	None	None	24
2007[4]	9 14	M F	Scoliosis Scoliosis	10x9 13.5x9, 11x7 Spinal canal extension	L R	L TR and correction surgery (Two stage surgery) R TR and correction surgery (Two stage surgery)	None None	None None	12 30
2017[2]	12	F	Chest tightness and lower extremity weakness	12x12 Spinal canal extension	L	LM and L TR (Subtotal resection)	None	No progression	12
2017[1]	5	M	Chest and upper abdomen pain	16x14	R	R TR	None	None	1
2017[9]	12	F	Shortness of breath	18x10	R	R TR	None	None	24
2019[8]	8	M	Upper respiratory tract infection	10x8	L	L TR	None	None	1
2020[6]	8	F	Chest and upper abdominal pain	23x20	R	R TR	Bleeding (intraoperative embolisation)	Unknown	10 days
2020[3]	17	F	Scoliosis	20x15, Spinal canal extension	L	Correction surgery and L TR (One stage surgery)	None	None	24
2021 (present case)	10	F	Dyspnea and chest pain	15x8	L	L TR	None	None	48

Abbrev.: A: age, S: sex, Dm: diameter, F: female, M: male, TR: thoracotomy, LM: laminectomy, Comp: complication, m:month, R: right, L: left

Giant ganglioneuroma is observed in cross-sectional images as a mass that compresses the surrounding tissues or surrounds them but does not invade them. It is usually observed as a homogeneous, and more rarely heterogeneous hypodense mass on CT examinations. It may contain calcifications. It shows weak to moderate or no enhancement in the early arterial phase and late venous phases [7]. In MRI, it is predominantly hypointense in T1W sequences, and mostly hyperintense in T2W sequences [1]. Low FDG uptake is expected to be seen in PET-CT [7] and biopsy is usually not required for diagnosis as in our case. Another important role of preoperative radiological imaging in thoracic ganglioneuroma cases is to reveal the origin and course of the Adamkiewicz artery. The Adamkiewicz artery occurs on the left side in 80% of the cases, between vertebrae T7 and L4. However, the anatomy frequently differs. If this large anterior radiculomedullary artery is damaged during surgery, neurological deficits may occur due to spinal ischemia-infarction [8]. In the present case, the origin of the Adamkiewicz artery was determined by CT angiography and it was reported to be far from the tumor origin.

Ganglioneuroma originates from the neural crest and is known to be hormonally inactive, although it

has neuroendocrine properties. It has been reported that catecholamine and metabolite release rate is 39% in ganglioneuroma, and an elevation of metanephrine, catecholamine, vasoactive intestinal peptide, dopamine, cortisol, homovanilic acid, or vanillylmandelic acid can be detected in urine or blood in those especially with adrenal gland localization [2]. The gold standard for hormonal activity in catecholamine secreting tumors is an increase in the level of metanephrine in the blood. In the present case, the levels of homovalenic acid and metanephrine in the urine were high but the catecholamine and metabolite values in the blood were in a normal range [6]. Thoracotomy or minimally invasive surgical approach can be applied in cases of intrathoracic ganglioneuroma, considering the size and location of the tumor. However, it has been reported in the literature that some ganglioneuroma cases have extension into the spinal canal and laminectomy with a posterior approach was also performed for complete resection. Scoliosis surgery can be performed to fix the thoracic vertebra angle that deteriorates with mass compression [4-9]. In the thoracotomy approach, the relation of the tumor with the surrounding tissues is better explored and intraoperative complications can be managed more safely [1]. Thoracoscopic resection can also be performed in smaller tumors. Malignant transformation of ganglioneuroma is rare. The recurrence rate is low and no additional postoperative treatment is required after complete resection [5].

In conclusion, ganglioneuroma should be considered in the differential diagnosis of giant posterior mediastinal tumors with smooth margins and low SUVmax values in pediatric patients. These tumors can be successfully resected with thoracotomy and can be cured without any additional treatment.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support.

Authors' contributions

BMKB, GK, MAS, BMY, AGÇ, BAK, AH, SE: conceived and designed the current case report, co-wrote the paper, collected the clinical data. The authors discussed the case under the literature data together and constituted the final manuscript.

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