Unusual presentation of a posterior mediastinal teratoma

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A B S T R A C T

We report a case of a five-year-old Caucasian girl with a posterior mediastinal teratoma. She presented with scoliosis and back pain. The teratoma was fixed to several surrounding structures, such as the aorta, the left lung and the chest wall, which complicated surgical excision. All these aspects are discussed and possible explanations for the development of mediastinal teratoma’s are reviewed.

The mediastinum is a common localization of extra-gonadal teratomas in children. It can present at any age and most are located in the anterior mediastinum with a close relationship to the thymus [1–3]. Only 3%–8% are located posteriorly [3,4]. Although mediastinal teratomas are mostly benign and asymptomatic, some patients present with respiratory symptoms.

Teratomas contain elements of all three germ layers ectoderm, endoderm, and mesoderm. Mediastinal teratomas may originate from primitive germ cells misplaced in the mediastinum during caudal migration in early embryogenesis or from native pluripotent mediastinal cells.

Therapy typically consists of complete surgical excision, without the need for chemotherapy.

We present a five-year-old Caucasian girl with a mature mediastinal posterior teratoma, growing into an intervertebral foramen, fixed to the aorta, lung and chest wall, and protruding into the posterior musculature of the back.

1. Case report

A five-year-old girl presented at our outpatient department with back pain over the last two years and a slight curvature to the right without relevant medical or family history. Physical examination revealed a mild spinal curve to the right with a left sided gibbus atypical for an idiopathic scoliosis. She did not show any neurological signs.

Spine and chest X-ray examinations showed a mass in the left hemithorax with a spinal curve convex to the right without vertebral torsion. Computed tomography (CT) scan of the chest showed a well-defined mass in the left paravertebral region from T6 to T11, measuring 10 × 8.8 × 9 cm. Macro calcifications were seen in the hypodense center of this mass with Hounsfield units resembling fatty tissue. A thin calcified layer, resembling an eggshell, largely surrounded the outer wall of the mass. Fig. 1A. The intervertebral foramina T8–T9 and T9–T10 were enlarged, suggesting extension of the tumor into the spinal canal. Fig. 1B. Vertebral anomalies suggestive of notochordal anomalies were not present. A hypodense structure was seen in the extra thoracic paravertebral back musculature. Fig. 1C. Ultrasound scan suggested a relationship between the intrathoracic mass and this lesion in the muscles.

Tumor markers for a neuroblastoma or a malignant germ cell tumor (GCT) (neuron specific enolase, urinary homovanillic acid and vanillylmandelic acid, αFP and β-HCG) were normal.

With a tentative diagnosis of teratoma a thoracoscopy was performed. Visualization was difficult because of the large mass. The approach was converted, therefore a posterolateral thoracotomy was chosen.

The mass was densely fixed to the lung, chest wall and aorta. For this reason and because the diagnosis was still uncertain, an incisional biopsy was performed. Hair bulged out of the tumor, confirming the diagnosis of a benign teratoma. Intracapsular resection
was decided upon because complete resection would be dangerous in the view of the dense adhesions to the aorta posterior to the large mass. Afterward the capsule was meticulously excised, confirming the adhesions to lung, aorta, and chest wall. The intercostal extension seen on the CT and ultrasound scans was transected at the level of the ribs. The extension into the vertebral foramina was transected at the level of the vertebral body.

The girl’s recovery was uneventful. The chest tube was removed on the 4th postoperative day. She was discharged home the next day. Two months later the lesion in the musculature of the back was removed through a separate longitudinal incision over the lesion, which was easily recognizable between the muscle fibers. A macroscopically complete resection was performed up to the level of the ribs. She was discharged the day after the operation.

Pathology showed a mature teratoma without immature or malignant aspects in both the intrathoracic tumor and the lesion in the back musculature, with respiratory, pancreatic and gastrointestinal epithelium next to multiple other types of tissue, classified as a type 1 GCT [3]. At follow up, one year after operation, she is doing well, the scoliosis recovered nearly completely and the back pain disappeared. MRI shows a small unchanged residual lesion in the intervertebral foramen.

2. Discussion

In contrast to the ovarian and sacrococcygeal region, the mediastinum is a rare location for a teratoma in childhood. Schneider et al. describes two German studies including 1193 patients with testicular and non-testicular GCT’s, 4.3% of these patients had a mediastinal teratoma [5]. Only 3–8% of these are located posteriorly [4].

Benign mediastinal teratomas are often asymptomatic and not diagnosed until a chest X-ray or CT scan is made for other indications [4,6]. If symptoms occur, they are usually respiratory problems: respiratory distress, dyspnea, pneumonia, cough, wheezing, and chest pain. The mass, might result in a chest wall deformity or signs of spinal cord compression [7].

![Fig. 1.](image1.png)

**Fig. 1.** A: CT scan showing macrocalcifications in the center of the tumor and a thin “egg-shell like” border (coronal reconstruction). B: Enlarged intervertebral foramen (arrow) with ingrowth of the tumor toward the spinal cord (coronal reconstruction). C: Extension of the tumor in the back musculature (sagittal reconstruction, between arrows).

![Fig. 2.](image2.png)

**Fig. 2.** Histology of the teratoma, with mature differentiated tissue of various origin e.g., pancreas (A), respiratory epithelium (B), colon (C) and bone marrow (D) (magnification ×200).
To our knowledge, the atypical scoliosis associated with lower back pain in our patient has not yet been described. A gibbus in scoliosis patients is normally seen on the side of the convexity: in this case, however the gibbus was on the left, whereas the convexity was to the right.

By the classification of GCTs into five types, proposed by Oosterhuis and Looijenga, all posterior mediastinal teratomas are type I (teratomas and yolk sac tumors, mostly extra-gonadal). Anterior mediastinal tumors can be type I or type II (seminoma or non-seminoma, mostly gonadal but also from the anterior mediastinum and the pineal region) [8].

The midline distribution of GCTs is still best explained by the route of embryonic germ cells from the yolk sac to the genital ridge. During migration they might become misplaced and/or dys functioning. However, alternative theories have been proposed, including progenitor cells within the thymus.

The theory that germ cells lie at the basis of these teratomas, which is in contrast to theories proposing stem cells as cells of origin, would require reprogramming because germ cells must be activated to pluripotency. The resultant type of GCT, i.e., type I or type II, to develop is likely determined by the interplay between the germ cell maturation status and the microenvironment.

Intramedullar teratomas have been reported, but they are rare and often associated with spinal disraphic lesions, such as myelomeningoceles [9,10]. Our search of the literature failed to find a report of an intramedullar extension of an otherwise posterior mediastinal teratoma. The absence of congenital vertebral anomalies or connection to the foraget made us conclude that this is not a case of a notochordal anomaly such as a neurenteric cyst.

Several articles describe dense adhesions of the mediastinal teratomas to surrounding structures such as the lung, aorta, and the heart; pulmonary resection is necessary in the case of bronchial fistulization [7,11,12]. The easiest explanation for these adhesions is rupture. The incidence of rupture of mediastinal teratomas (up to 36%) is higher than that of teratomas in other locations [13]. Possible explanations vary from necrosis/ischemia to autolytic effects by pancreatic secretions to infection. Pancreatic tissue is described more often in mediastinal teratomas [13–15]. The reason for this increased pancreatic differentiation is not clear. These adhesions are often mistaken for malignancy. In our case the dense pleural adhesions to lung and thoracic wall, as well as the extension in the vertebral foramina and into the muscles of the back, suggest leakage of autolytic enzymes, facilitating muscle ingrowth. Both pancreatic and intestinal tissue was present histologically.

3. Conclusion

Posterior mediastinal teratoma may present as a fixed lesion with unusual pattern of growth due to a higher incidence of rupture, possibly due to greater abundance of pancreatic tissue.

Although this type of teratoma is often asymptomatic, atypical symptoms such as thoracic deformity or atypical scoliosis are worth further investigation.

Informed consent

Written informed consent was obtained from the patient 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.

Conflicts of interest statement

For all authors none were declared.

References